Anatomy

Question 8: Anatomy - Peripheral Nervous System

Discussion:

The suprascapular nerve innervates the supraspinatus and infraspinatus muscles. The supraspinatus muscle is responsible for the first 15 degrees of humeral abduction, and the infraspinatus muscle externally rotates the arm.

References:


Question 10: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The circumventricular organs, which do not have a blood-brain barrier, are the area postrema, subfornical organ, organum vasculosum, neurohypophysis, median eminence, pineal gland, and subcommissural organ. The area postrema has been implicated as a chemoreceptor trigger zone for vomiting.

References:


Question 27: Anatomy - Peripheral Nervous System

Discussion:

Some of the muscles with contributions from the L5 myotome include the tibialis anterior, tibialis posterior, peroneus longus, tensor fascia latae (TFL), gluteus medius, semitendinosus and semimembranosus, and the biceps femoris (long and short head). The sciatic nerve innervates all of the hamstring muscles and all muscles below the knee via the peroneal and tibial nerve branches. Of the choices, only the tensor fascia latae could be weak with an L5 but spared with a sciatic mononeuropathy.

References:


Question 32: Anatomy - Brainstem/Cerebellum

Discussion:

Golgi cells are found in the granule cell layer of the cerebellum and provide feedback inhibition of the granule cells. Basket cells, stellate cells, and parallel fibers are found in the molecular layer along with Purkinje cell dendrites.

References:
Question 43: Anatomy - Peripheral Nervous System

Discussion:

The sweat glands are innervated by postganglionic sympathetic cholinergic fibers. Cholinergic inputs stimulate sweat production via M3 type muscarinic receptors.

References:


Question 50: Anatomy - Cranial Nerves

Discussion:

The baroreceptor reflex helps to regulate blood pressure by detecting changes via baroreceptors mostly located in the carotid sinus and aortic arch. Information from the carotid sinus travels via the glossopharyngeal nerve while that from the aortic arch is carried by the vagus. All baroreceptor afferents terminate in the nucleus tractus solitarius, which then projects to the ventrolateral medulla. A decrease in blood pressure results in an increase in sympathetic tone and decrease in parasympathetic tone, with an increase in blood pressure producing the opposite effect.

References:


Question 60: Anatomy - Embryology

Discussion:

Lissencephaly, which is characterized by a reduction in the number of gyri and sulci, is one of the disorders of neuronal migration. Other neuronal migration disorders include polymicrogyria, schizencephaly, and focal cortical dysplasia.

References:


Question 71: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The central retinal artery is a branch of the ophthalmic artery and makes only a minimal contribution to the vascular supply of the optic disc. The short posterior ciliary arteries are branches that also arise from the ophthalmic artery and supply the optic disc and the retro-orbital optic nerve.

References:

Question 78: Anatomy - Brainstem/Cerebellum

Discussion:

The three openings in the 4th ventricle are the two paired lateral apertures (foramina of Luschka) and the median aperture (foramen of Magendie). CSF flows from the 4th ventricle, via these foramina, to reach the subarachnoid space of the cisterna magna.

References:


Question 86: Anatomy - Cranial Nerves

Discussion:

Efferent fibers from the subnuclei of cranial nerve III for the medial rectus, inferior rectus, and inferior oblique proceed ipsilaterally. Fibers from the subnucleus for the superior rectus decussate. The central caudal nucleus is a midline cell group of III that gives rise to both crossed and uncrossed fibers that innervate the levator palpebrae muscle.

References:


Question 88: Anatomy - Brainstem/Cerebellum

Discussion:

The majority of axons of cerebellar Purkinje cells synapse with neurons in the deep cerebellar nuclei. A few from the vermis and flocculonodular lobule project directly to the vestibular nuclei.

References:


Question 120: Anatomy - Peripheral Nervous System

Discussion:

Preganglionic sympathetic fibers leave the spinal cord via the ventral roots of T1 and T2 and then join the paravertebral sympathetic chain and synapse in the superior cervical ganglion. Postganglionic fibers follow the carotid plexus, eventually reaching the pupillo-dilator muscle.

References:


Question 127: Anatomy - Cortex and Connections

Discussion:
The insula and the lateral aspect of the postcentral gyrus are primarily involved in the initial cortical processing of taste.

**References:**


**Question 137: Anatomy - Cranial Nerves**

**Discussion:**

The nervus intermedius is the portion of the facial nerve carrying all the general visceral efferent, general somatic afferent, and special afferent information. The branchial motor fibers going to the muscles of facial expression travel in a separate bundle. Of the potential answers given, an injury to the nervus intermedius would therefore impair taste from the ipsilateral anterior tongue. Facial muscles would not be affected. The parotid gland is innervated by the glosopharyngeal nerve while the striated muscles of the pharynx are innervated by the vagus nerve.

**References:**


**Question 144: Anatomy - Basal Ganglia and Thalamus**

**Discussion:**

Fibers from the anterior nucleus of the thalamus project to the cingulate gyrus. The anterior nucleus receives projections from the mammillary bodies. This is part of the classic Papez circuit that formed the basis of the concept of the limbic system.

**References:**


**Question 151: Anatomy - Cortex and Connections**

**Discussion:**

The dentate gyrus (DG) is contiguous with the hippocampal formation (HF); both are three-layered cortex (archipallium). The cortical zones from the DG and HF, through the prosubiculum, subiculum, presubiculum, and parahippocampal gyrus show a gradual transition from three-layer to six-layer cortex. The entorhinal region (area 28) is six-layered cortex.

**References:**


**Question 163: Anatomy - Brainstem/Cerebellum**

**Discussion:**

Parinaud syndrome is due to lesion in the dorsal midbrain, involving the quadrigeminal plate, the pretectum and periaqueductal gray matter. Clinical findings include impaired upgaze, convergence retraction nystagmus, eyelid retraction and light-near dissociation.
**Question 167: Anatomy - Cranial Nerves**

**Discussion:**

A unilateral lesion of the vagus nerve produces ipsilateral paralysis of the soft palate, pharynx and larynx. The glossopharyngeal nerve does supply the stylopharyngeus muscle which elevates the pharynx during talking, swallowing, and gagging. A lesion of IX, however, would not be expected to cause complete unilateral vocal cord paresis. The other nerves do not innervate the palate or laryngeal muscles. The nucleus tractus solitarius processes visceral afferent information carried by cranial nerves.

**References:**


**Question 168: Anatomy - Blood Supply of Brain/Spinal Cord**

**Discussion:**

The medial medullary syndrome is most commonly associated with infarction in the anterior spinal artery distribution at the level of the medulla. An occlusion at this level may result in ipsilateral CNXII paralysis (fascicle of CNXII), contralateral hemiparesis (pyramid), and contralateral loss of position and vibratory sensation (medial lemniscus). A bilateral lesion in this vascular territory will result in quadriparesis, bilateral loss of proprioception and vibration, and complete paralysis of the tongue.

**References:**

Discussion:

Proprioceptive and vibratory loss in the lower extremities, due to a spinal cord lesion, involves the fasciculus gracilis which serves these functions below the level of T6.

A central cord lesion which produces pain and temperature dysfunction in a bilateral "shawl" or "cape" distribution is due to involvement of crossing fibers for these modalities in the anterior (a.k.a. ventral) white commissure.

In high cervical cord lesions, ipsilateral diminished pain and temperature sensation in the preauricular area of the face is due to involvement of cells in the substantia gelatinosa which is the distal continuation of the descending trigeminal nucleus and tract.

The intermediolateral cell column contains preganglionic sympathetic neurons. Involvement of this area in the upper thoracic cord may result in an ipsilateral Horner syndrome.

Extramedullary impingement of the lateral cervical cord may cause weakness and upper motor neuron signs in the ipsilateral lower extremity because of the somatotopic organization of sacral and lumbar fibers being most lateral within the lateral corticospinal tract.

References:


Question 222: Anatomy - Cortex and Connections

Discussion:

The arrow is pointing to the external capsule at the level of the union of the putamen and caudate. The external capsule carries cholinergic fibers from the basal forebrain and is bounded by the putamen and the claustrum.

References:


Question 225: Anatomy - Cortex and Connections

Discussion:

The arrow is indicating the hippocampus which is the structure first affected in the neurodegenerative pathophysiology of Alzheimer disease. Multisystem atrophy involves striatonigral and olivopontocerebellar regions. Parkinson’s disease involves dopaminergic systems. Huntington Disease involves the caudate and putamen.
Question 248: Anatomy - Cranial Nerves

Discussion:

The arrow is pointing to the third cranial nerve - which emerges ventrally from the midbrain in the interpeduncular fossa at the level of the superior colliculus. The only other cranial nerve which arises from the midbrain is the fourth and it emerges from the dorsum. The third cranial nerve leaves the skull by passing through the superior orbital fissure (as does four, V1, and six). V2 exits via the foramen rotundum while V3 exits via the foramen ovale. The optic nerve exits by way of the optic canal. The foramen lacerum provides entry for the internal carotid artery.

References:


Question 263: Anatomy - Basal Ganglia and Thalamus

Discussion:

The arrow is pointing to the substantia nigra, which normally functions to facilitate voluntary motor activity originating in the prefrontal and motor cortex, ipsilateral to the substantia nigra. Damage to one substantia nigra results in hemiparkinsonism. Since the upper motor neurons of the motor strip are the motor output elements, and these cross at the medullary decussation, the hemiparkinsonism resulting from a unilateral substantia nigra lesion is contralateral to the lesion.

References:


Question 278: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The posterior cerebral artery supplies parts of the inferior temporal lobe, occipital lobe, splenium of the corpus callosum and superior parietal lobule.

References:


Question 337: Anatomy - Peripheral Nervous System

Discussion:

After exiting the tarsal tunnel, the tibial nerve will divide into the medial and lateral plantar nerves. These nerves relay sensation from the plantar skin of the foot. Another branch of the tibial nerve is the calcaneal nerve which innervates the skin over the heel.
Secondary auditory fibers from the cochlear nuclei form the dorsal, intermediate, and ventral acoustic striae. The dorsal and intermediate striae cross the midline and enter the lateral lemniscus. The fibers of the ventral stria terminate in the superior olivary nuclei and the nucleus of the trapezoid body. These nuclei give rise to tertiary fibers that enter the lateral lemnisci. The lateral lemniscus ascends to the midbrain where most of the fibers terminate in the inferior colliculi.

References:


Established projections to the globus pallidus arise from the striatum and subthalamic nuclei. The pallidum does not receive direct afferents from the cerebral cortex, dentate nucleus, or thalamus.

References:

The medial lemniscus ascends through the brainstem and terminates in the ventral posterolateral nucleus of the thalamus. The centromedial nucleus, one of the intralaminar nuclei, and the ventral anterior nucleus are involved in basal ganglia circuitry. The ventral lateral nucleus receives input from the deep cerebellar nuclei. The ventral posteromedial nucleus receives sensory input from the face via the trigeminothalamic tract.

References:

A lesion to the angular gyrus of the dominant parietal lobe can cause Gerstmann syndrome, which is characterized by the dyscalculia, finger agnosia, dysgraphia, and right-left confusion. The angular artery, a branch of the middle cerebral artery supplies this part of the cortex.
Question 361: Anatomy - Peripheral Nervous System

Discussion:

The common peroneal nerve divides into two branches, the deep and superficial peroneal nerves. The deep peroneal nerve innervates the tibialis anterior and extensor digitorum brevis. The peroneus longus and brevis are supplied by the superficial peroneal nerve. The soleus and tibialis posterior muscles are innervated by the tibial nerve.

References:

Question 380: Anatomy - Blood Supply of Brain/Spinal Cord

Discussion:

The internal auditory artery (IAA) typically arises from the anterior inferior cerebellar artery (AICA), but may also arise directly from basilar artery. The IAA will then divide into a cochlear and vestibular branch to supply the labyrinth. AICA ischemia can therefore result in unilateral deafness.

References:

Question 390: Anatomy - Cortex and Connections

Discussion:

Vertical saccades are initiated by activating the connection between the frontal eye fields and the rostral interstitial nucleus of the MLF (rMLF). Horizontal saccades are initiated by a signal from the frontal eye fields to the ipsilateral superior colliculus and the contralateral paramedian pontine reticular formation.

References:

Question 395: Anatomy - Peripheral Nervous System

Discussion:

The posterior cord of the brachial plexus gives off the thoracodorsal and subscapular nerves and terminates by splitting into the axillary and radial nerves. Any muscles innervated by these branches may be weakened with a lesion in the posterior cord of the plexus.

References:
Behavioral/Psychiatry

Question 6: Behavioral/Psychiatry - Language Disorders

Discussion:

A transcortical motor aphasia is a nonfluent aphasia with preserved naming and repetition, differentiating it from a Broca aphasia, which has impairment of spontaneous speech, naming, and repetition. The most common neurologic symptom is a right hemiparesis that predominately affects the leg. The lesion producing a transcortical motor aphasia lies in the left frontal lobe and is usually caused by a stroke of the anterior cerebral artery. Sensory loss is usually absent or restricted to the right leg and visual field deficits are usually absent.

References:


Question 7: Behavioral/Psychiatry - Developmental Disorders

Discussion:

Pimozide and haloperidol are approved and the most effective treatments of Tourette's disorder.

References:


Question 12: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Symptoms of herpes simplex encephalitis include anosmia, olfactory and gustatory hallucinations, and personality changes that can involve bizarre and psychotic behaviors. Symptoms of rabies encephalitis include agitation, restlessness and hydrophobia. Symptoms of neurosyphilis personality change, poor judgement, delusions of grandeur, poor self-care. Symptoms of cryptococcal meningitis include headache, memory impairment and confusion. Symptoms of Lyme disease include irritability, depression, poor concentration and memory impairment.

References:


Question 20: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

A patient with a 4- to 5-day history of headache, malaise, personality change, anterograde memory deficit, and fever should be presumed to have HSV-1 encephalitis until proven otherwise. The most affected areas of the brain are the medial temporal and orbitofrontal lobes, although the cingulate gyrus is less commonly affected as well. The treatment of choice is acyclovir given IV at 10mg/kg every 8 hours for 3 weeks.

References:

Question 22: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

The Alzheimer disease neuroimaging initiative (ADNI) was established to look for biomarkers to assist in the diagnosis and treatment of Alzheimer disease (AD) and mild cognitive impairment (MCI). Numerous studies have evaluated the cerebrospinal fluid profiles of normal controls, patients with MCI who progressed to dementia, stable MCI, and Alzheimer disease. The presence of increased phospho-tau levels, in conjunction with decreased AB42 levels, is most closely associated with conversion from MCI to AD. AB42 levels are also most sensitive to functional decline in patients with AD.

References:


Question 25: Behavioral/Psychiatry - Developmental Disorders

Discussion:

The described patient has Kleine-Levin syndrome. As with most other syndromes that affect sleep cycle and eating behavior, lesions affecting these behaviors have most commonly been found in the hypothalamus.

References:


Question 34: Behavioral/Psychiatry - Dementia

Discussion:

Alzheimer disease is characterized by amnestic memory loss, transcortical aphasia, and visuospatial deficits. Patients with dementia with Lewy bodies typically have Parkinsonism. Pick disease and frontotemporal dementia typically do well with calculations and constructions. Progressive posterior cortical atrophy has severe visuospatial deficits but not as significant memory problems.

References:


Question 35: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Dopamine agonist therapy has been associated with several impulse control and repetitive behaviors in Parkinson disease. These behaviors include new onset gambling, compulsive shopping and eating, hypersexuality, and punding. This appears to be a class effect with dopamine agonists. Risk factors for the development of these disorders has been associated with younger age, unmarried status, and family history of gambling disorders.

References:


**Question 38: Behavioral/Psychiatry - Anatomic syndromes**

**Discussion:**

This patient is presenting with the sudden onset of the inability to balance her checkbook suggesting she has trouble with calculations. Her exam is indicative of mild finger agnosia and dysgraphia. This constellation of symptoms is part of the Gerstmann syndrome that has been localized to left angular gyrus.

**References:**


**Question 40: Behavioral/Psychiatry - Language Disorders**

**Discussion:**

Patients with auditory receptive aphasia experience difficulty verbalizing their thoughts because of output errors at the level of words (semantic paraphasias) and parts of words (phonemic paraphasias).

**References:**


**Question 44: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam**

**Discussion:**

In the Stroop test, names of colors are spelled in different color ink and the patient is asked to name the ink color although the actual word spelled may be different than the ink color. For example, the word RED is spelled in blue ink and the patient needs to be able to suppress the actual printed word they see and say "blue."

**References:**


**Question 61: Behavioral/Psychiatry - Dementia**

**Discussion:**

According to Practice Parameter update: Evaluation and management of driving risk in dementia, the clinical features that indicate his driving is unsafe is the concern reported by the family and his aggressive personality change.

**References:**

Question 62: Behavioral/Psychiatry - Dementia

Discussion:

Atypical antipsychotics have been shown to have a small increased risk of death, compared to placebo, when used in patients with dementia.

References:


Question 67: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

This syndrome is known as Klüver-Bucy syndrome and occurs as a result of bilateral temporal lesions involving amygdalae. Hypermetamorphosis, a common feature, involves excessive fascination with objects in the patient's environment and the urge to touch them all.

References:


Question 74: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

Achromatopsia, the loss of the ability to recognize color, will cause total failure of number or pattern recognition on the Ishihara plates. Patients with color anomia still retain the ability to perceive the patterns on the Ishihara plates. Optic aphasia renders patients unable to name visually presented objects and would not affect Ishihara testing. Optic ataxia is the inability to move one's limb toward an object using visual cues.

References:


Question 77: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

Simultagnosia is the inability to integrate different aspects of a visual scene to perceive it as a whole. It is one of the characteristics of the Balint syndrome. The other features include ocular apraxia and optic ataxia. The disorder usually results from bilateral damage to the occipital and parietal visual association areas superiorly.

References:


Question 82: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:
Structural or functional lesions in the left anterior frontal lobe are more often associated with depression than other areas.

References:


Question 90: Behavioral/Psychiatry - Language Disorders

Discussion:

Defective repetition may be associated with Broca aphasia, Wernicke aphasia, conduction aphasia, and global aphasia. Intact repetition is seen in transcortical sensory aphasia.

References:


Question 94: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Depression is seen commonly in patients with multiple sclerosis. It has a higher incidence than most chronic illnesses suggesting cortical involvement of MS contributes to the mood disorder.

References:


Question 95: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:

PET using Pittsburgh compound-B is currently the most specific in vivo reflection of amyloid-beta protein load.

References:


Question 99: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:
The child has manifestations of juvenile Huntington disease (HD). The adult onset form of the disorder is associated with greater than 40 CAG repeats. The gene location is 4p16.3. Like many trinucleotide repeat disorders, earlier onset is often reported in subsequent generations (anticipation), as the repeat expands.

References:

Question 106: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:
Transactivating responsive sequence DNA-binding protein (TDP-43) is a protein encoded by the TARDBP gene on chromosome 1. Numerous studies have linked this protein to the syndromes of amyotrophic lateral sclerosis, frontotemporal dementia with motor neuron disease and frontotemporal dementia with ubiquitin inclusions. Microtubule associated protein tau and valosin-containing protein have also been found in some cases of frontotemporal dementia, however rarely in those also exhibiting motor neuron disease.

References:

Question 108: Behavioral/Psychiatry - Genes, Biomarkers, & Networks

Discussion:
Mutations in the progranulin gene can cause frontotemporal dementia. They are not associated with any of the other listed degenerative dementia conditions. Biomarkers for Alzheimer's disease involve markers for amyloid accumulation and markers for neurodegeneration. Parkinson's disease and dementia with Lewy bodies are associated with abnormal synuclein. Abnormality of the huntington gene on chromosome 4p16.3 results in Huntington disease.

References:

Question 111: Behavioral/Psychiatry - Psychopharmacology

Discussion:
Paroxetine (Paxil) is often used to reduce anxiety, and can often help tics as an additional effect. Paroxetine is most often prescribed when an anxiety disorder, such as obsessive-compulsive disorder, accompanies Tourette's Syndrome. Methylphenidate (Ritalin) is often used to treat associated behavioral disorders such as ADHD, but can sometimes have the side effect of increasing tics. Clonidine can help with the reduction of tics, and as a treatment for the effects of ADHD but is not particularly useful for OCD. Modafinil is a stimulant which can umask tics. Lorazepam would not be particularly helpful for ADHD or Tourette’s.

References:
Robertson, Mary. Tourette Syndrome, Associated Conditions and the Complexities of Treatment.. Brain 2000;123: 425-462.
Question 142: Behavioral/Psychiatry - Neurobehavioral/Neuropsychological Exam

Discussion:

The impairment in motor programming with insufficient rhythm tapping, set shifting, and go/no-go tasks is typically seen in injuries of the dorsolateral frontal regions.

References:


Question 148: Behavioral/Psychiatry - Dementia

Discussion:

Mild memory impairment or subtle changes in other cognitive functions that do not interfere with daily activities and for which no other underlying cause can be found constitute mild cognitive impairment (MCI). Age-associated memory impairment (AAMI) is normal for age but worse than young adults. In DSM 5, major neurocognitive disorder is the new name for dementia. In DSM 5, minor neurocognitive disorder is equivalent to mild cognitive impairment. This patient does not have dementia since there is not functional change.

References:


Question 160: Behavioral/Psychiatry - Language Disorders

Discussion:

Aphemia, also known as apraxia of speech, is characterized by occasional mutism, diminished fluency, and deficits in articulation. Despite appearing to have nonfluent aphasia, patients with aphemia have retained the ability to write.

References:


Question 171: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

Apathy and disinhibition are two of the most recognizable neuropsychiatric symptoms seen in frontal systems disorders. Visual hallucinations are seen more in conditions causing temporal-occipital regional dysfunction. Delusions are thought to be more temporal lobe in origin. Sleep disturbances and aberrant motor behaviors can be seen in a variety of neuropsychiatric conditions and are not specific to the frontal lobes.
References:


Questions 193 - 196: Behavioral/Psychiatry - General Psychiatry

Discussion:

A key feature in the diagnosis of somatization disorder is the onset of multiple physical complaints beginning before the age of 30. After appropriate evaluation symptoms can't be fully explained by a known medical condition. Diagnosis requires four pain symptoms, two gastrointestinal symptoms, one sexual symptoms and one pseudoneurologic symptom.

Conversion disorder is associated with symptoms suggesting a neurologic cause in which there is initiation or exacerbation of symptoms in association with psychological conflicts or stress. Psychological mechanisms include expression of psychological conflict that has been repressed and identification with a family member (often the source of conflict) who has the same symptoms.

Characteristics of dissociative disorders include disruption of normally integrated functions of consciousness, environmental perception, memory and identity. Patients with dissociative identity disorder, in particular, will manifest two or more distinct personalities and etiology is related to severe physical and/or sexual abuse in childhood.

Malingering is associated with the voluntary production of symptoms to accomplish a specific goal (such as obtaining drugs, getting insurance benefits or avoiding something perceived as unpleasant).

References:


Question 314: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

This patient is presenting with the clinical syndrome of Hashimoto encephalopathy. In this condition, TSH and T3/T4 levels are frequently normal; however, anti-microsomal antibodies are typically elevated. This condition should be in the differential of any patient with a rapidly progressive dementia, especially in the presence of seizures. Hashimoto encephalitis responds well to intravenous corticosteroid treatment and plasmapheresis.

References:


Question 339: Behavioral/Psychiatry - Anatomic syndromes

Discussion:

This patient presents with symptoms of a stroke that involves the left posterior cerebral artery affecting the temporo-occipital lobe with corpus callosal sparing. Unilateral left temporo-occipital lesions usually result in object agnosia or the inability to recognize individual objects but spares face recognition (prosopagnosia) which is usually caused by right hemisphere temporo-occipital involvement.

If the splenium of the corpus callosum was damaged, it can cause a pure alexia without agraphia due to the right occipital lobe's inability to send visual information to the language centers of the brain on the left. This patient is able to read and write.
Simultagnosia, or the inability to perceive more than one item or feature at a time, is usually caused by bilateral lesions to the occipitoparietal lobes. When combined with optic apraxia and ocular ataxia, it is more commonly known as Balint syndrome.

Another common finding that was not mentioned is some degree if not a complete right homonymous hemianopsia.

**References:**


**Question 352: Behavioral/Psychiatry - Psychopharmacology**

**Discussion:**

Quetiapine is an effective antipsychotic with very little dopaminergic blocking activity; hence, it is useful in Parkinson disease patients with hallucinations or delusions. It acts predominantly on serotonin receptors. Risperidone, perphenazine, and haloperidol all have more dopamine blocking properties than quetiapine. Chlorpromazine carries a risk for orthostatic hypotension and potential for falls. Olanzapine has more extrapyramidal and anticholinergic properties than quetiapine.

**References:**


**Question 356: Behavioral/Psychiatry - Dementia**

**Discussion:**

Rivastigmine is FDA approved to treat mild to moderate Parkinson and Alzheimer disease. Donepezil is FDA approved for treatment of mild to severe Alzheimer disease. Galantamine is FDA approved for mild to moderate Alzheimer disease.

**References:**


**Question 357: Behavioral/Psychiatry - Language Disorders**

**Discussion:**

This case is classical for a conduction aphasia. Typical features of conduction aphasia include frequent paraphasic errors, impaired repetition, spontaneous verbal output but fair to good comprehension and only mild impairment in fluency. Aphemia is due to subcortical frontal lobe damage resulting in initial muteness but then clearing to an effortful and slow articulation problem. Broca aphasia typically effects frontal cortex regions, results in poor repetition, dysfluent speech and reasonable comprehension abilities. Global aphasia results in both impaired fluency and comprehension and relates to large lesions in the left hemisphere. Transcortical sensory aphasia involves parietal regions and typically results in intact repetition and a paraphasic fluent aphasia.

**References:**

Question 363: Behavioral/Psychiatry - Dementia

Discussion:

Functional imaging such as PET provides a measure of how the brain is functioning. In patients with frontotemporal dementia (FTD), PET scans typically show frontal and anterior temporal regional hypometabolism. In patients with probable Alzheimer disease (AD), the PET pattern reveals bilateral posterior temporal and parietal hypometabolism. Each shows a distinct pattern on PET imaging. EEGs are more nonspecific in dementia and typically show just generalized slowing and frontal slowing in both FTD and AD. Routine CSF analysis and cerebral angiogram would show similar results in probable AD patients and FTD patients. Routine MRI could show differential frontal atrophy in the FTD patients and hippocampal atrophy in the probable AD patients, but these findings are subtle.

References:


Question 364: Behavioral/Psychiatry - Psychopharmacology

Discussion:

The serotonin syndrome results from concomitant administration of medications that enhance serotonin transmission via decreased breakdown, or increased production. Medication combinations to use cautiously include monoamine oxidase inhibitor agents with selective serotonin reuptake inhibitors, tricyclic antidepressants, or dextromethorphan. The serotonin syndrome can be differentiated from neuroleptic malignant syndrome by the presence of shivering and myoclonus in the former.

References:


Question 367: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:

Neurologic involvement can occur in 5% to 10% of patients with sarcoidosis. Manifestations can include motor and sensory deficits, progressive visual loss, and cranial nerve palsies. Cognitive impairments can range from mild memory loss to progressive dementia. Sarcoidosis appears to have a predilection for the hypothalamus and basal forebrain resulting in hypopituitarism, hyperphagia, and hypersomnolence.

References:


Question 375: Behavioral/Psychiatry - Psychopharmacology

Discussion:

Agranulocytosis, defined by an absolute neutrophil count of less than 500 per cubic millimeter, has been estimated to occur in association with clozapine use at a cumulative incidence at 1 year of about 1.3%. This is why all patients need to be registered and white blood counts followed regularly if this drug is prescribed. This amount of risk is not seen with any of the other antipsychotic medications listed here.
References:

Clozaril Package Insert.

Question 377: Behavioral/Psychiatry - Behavioral Complications of Systemic Disease

Discussion:
MS patients often develop pseudobulbar affect, associated with involuntary emotional displays (typically laughing or crying) that are often out of proportion to the patient's mood. While depression is common, bipolar illness is not. Obsessive-compulsive traits, visual hallucinations, and physical aggression are seen less often.

References:


Question 384: Behavioral/Psychiatry - Dementia

Discussion:
The dementia with Lewy bodies (DLB) consortium has revised criteria for the clinical and pathologic diagnosis of DLB, incorporating new information about the core clinical features and suggesting improved methods to assess them. REM sleep behavior disorder, severe neuroleptic sensitivity, and reduced striatal dopamine transporter activity on functional neuroimaging are given greater diagnostic weighting as features suggestive of a DLB diagnosis. When any of these features are present with one of the primary findings of visual hallucinations, parkinsonism, or fluctuating attention, then the diagnosis of probable DLB is supported.

References:


Question 396: Behavioral/Psychiatry - Psychopharmacology

Discussion:
Sexual side effects may occur in up to 80% of patients on a selective serotonin receptor antagonist (SSRI) class of medication. The most common symptoms are decreased libido and inhibited orgasm. These side effects are dose dependent, do not resolve over time and can be expected to continue as long as the patient takes the medication. Medications in the SSRI class include fluoxetine, paroxetine, sertraline, citalopram and escitalopram. Bupropion is an antidepressant that is not in the SSRI class and is not commonly associated with sexual dysfunction.

References:

Question 398: Behavioral/Psychiatry - Anatomic syndromes
Discussion:

Balint's syndrome is composed of the triad of optic ataxia (the inability to reach for an object under visual guidance), simultanagnosia (the inability to view multiple objects at once), and ocular apraxia (the inability to voluntarily direct gaze to a specific target). It usually results from bilateral damage to the parietal-occipital region secondary to separate infarctions. However, cases have been reported following head trauma as well as neoplastic disease.

References:


Clinical Adult

Question 2: Clinical Adult - Neuromuscular Disorders

Discussion:

Benign fasciculation syndromes presents with fasciculations in multiple muscles without associated cramps or weakness. Neurologic examination and electrodiagnostic testing are necessary to ensure that the patient does not have motor neuron disease or cramp-fasciculation syndrome. Patients can improve with withdrawal of caffeinated substances or treatment to allay anxiety.

References:


Question 5: Clinical Adult - Critical Care/Stroke

Discussion:

An epidural hemorrhage is most commonly caused by trauma and can involve the middle meningeal artery. It is characterized by a lucid period and a delay prior to symptomatic development of a blood collection. In this patient, extensive bleeding in the left hemisphere led to right-sided long tract signs and uncal herniation.

References:


Question 15: Clinical Adult - Neurogenetics

Discussion:

The history and examination are consistent with type 1 Charcot-Marie-Tooth disease (CMT1), the most common form of CMT disease. CMT1 is an inherited autosomal dominant sensory and motor peripheral neuropathy with hypertrophic demyelinating pathology. The most commonly present genetic alterations are either duplications or point mutations on the PMP22 gene. Nerve conduction slowing with velocities less than 30 m/s is characteristic. The other gene alterations listed are associated with CMT2, an axonal CMT variant typically manifesting nerve conduction velocities faster than 40 m/s.

References:

Question 29: Clinical Adult - Motor Neuron/Nerve

Discussion:

Painless asymmetric weakness is a classic presentation of motor neuron disease. In patients with no evidence of upper motor neuron involvement, a diagnosis of multifocal motor neuropathy (MMN) must be excluded. Partial motor conduction blocks are a diagnostic hallmark of MMN.

References:

Mendell JR, Kissell JT, Comblath DR. Diagnosis and Management of Peripheral Nerve Disorders. New York: Oxford University Press, 2001;592-638

Question 31: Clinical Adult - Neurology of Systemic Disease

Discussion:

A femoral neuropathy or high lumbar plexus lesion, associated with retroperitoneal pain in a patient with coagulopathy, suggests a retroperitoneal hemorrhage. Imaging is required to confirm the diagnosis.

References:


Question 42: Clinical Adult - Other Pain Syndromes

Discussion:

This patient most likely has herpes zoster infection affecting the trigeminal ganglion. The most appropriate therapy would be treatment with IV acyclovir.

References:


Question 48: Clinical Adult - Dementia

Discussion:

The APOE-4 gene located on chromosome 19 is associated with late-onset Alzheimer disease. Presenilin 1, presenilin 2 and amyloid precursor protein are associated with early-onset Alzheimer disease. The protein tau is encoded by MAPT and is implicated in the pathogenesis of frontotemporal dementia.

References:


Question 49: Clinical Adult - Neuromuscular Disorders

Discussion:
The patient has the hallmarks of an inflammatory myopathy. Muscle biopsy is the best way to confirm and characterize inflammatory myopathies. Although both EMG-nerve conduction studies and skeletal muscle MRI can help to support a diagnosis, neither is specific for any one type of inflammatory myopathy. Interstitial lung disease and dysphagia can be associated with inflammatory myopathies but are also not specific. Acetylcholine receptor binding antibodies are used to diagnose myasthenia gravis, which is unlikely given the examination and markedly elevated serum CK.

References:

Question 51: Clinical Adult - Demyelinating Disease
Discussion:
An area postrema syndrome, characterized by intractable hiccups, nausea, and vomiting, occurs in up to 43% of patients with Neuromyelitis Optica Spectrum Disorder.

References:

Question 63: Clinical Adult - Neurorehabilitation
Discussion:
The patient sustained 2 concussions in close temporal relationship resulting in an increased risk of prolonged postconcussion symptoms. The cornerstone of concussion management is physical and cognitive rest from activities exacerbating symptoms. After acute symptoms abate, a graded program of exertion can be implemented prior to clearance for return to play. Conventional structural neuroimaging is typically normal in concussive injury, but should be employed whenever suspicion of an intracerebral or structural lesion (eg, skull fracture) exists.

References:

Question 73: Clinical Adult - Dementia
Discussion:
Adult-onset polyglucosan body disease is a disorder typically presenting in the fifth to seventh decades of life. Patients present with gait difficulties with lower and upper motor neuron involvement and peripheral neuropathy, with sensory deficits predominantly in the lower extremities, neurogenic bladder, and dementia. The combination of lower and upper motor neuron signs with leukoencephalopathy should lead the clinician to suspect this entity.

References:

Question 81: Clinical Adult - Movement Disorders
Discussion:
Acute dystonic reactions are often observed after treatment with potent dopamine D2-receptor antagonists, including metoclopramide. Cervical and limb dystonia are most common. Laryngeal dystonia may be a life-threatening form of dystonic reaction in these patients. Anticholinergic medication, including diphenhydramine, is the most appropriate treatment.

References:


**Question 83: Clinical Adult - Spinal and Root Disorders**

Discussion:

Metastatic epidural spinal cord compression presents most commonly with pain and is associated with weakness, bowel and bladder incontinence, and sensory loss that can be localized to the spinal cord. Emergent treatment in patients with evidence of myelopathy should be instituted with high-dose dexamethasone in order to maintain and preserve ambulation.

References:


**Question 97: Clinical Adult - Cerebrovascular Disease**

Discussion:

Although the pattern of recovery following a stroke may be variable, the earliest evidence of a return of neurologic function is usually an increase in tone. This generally occurs before improvement in strength or a return of reflexes.

References:


**Question 102: Clinical Adult - Sleep**

Discussion:

This individual has REM behavior disorder (RBD). It is estimated to be a harbinger of an alpha-synucleinopathy in over 50% of individuals with RBD (especially for Parkinson disease, dementia with Lewy bodies and multiple systems atrophy). Alzheimer disease (AD) is associated with sleep-wake disturbances, but the latter is not a known risk factor for AD. ALS is associated with sleep related breathing disorders, but the latter are not known risk factors for ALS. Nocturnal seizures with complex behaviors can simulate RBD. They are generally focal seizures and arise out of NREM sleep. The prevalence of progression to generalized seizures is unknown. Obstructive sleep apnea is a well documented risk factor for stroke.

References:


**Question 105: Clinical Adult - Dementia**

Discussion:

Primary progressive aphasia (PPA) is a frontotemporal dementia (frontotemporal degeneration, FTD) that is characterized initially by word-finding trouble, difficulty with naming, and comprehension. These language features are typically a dominant component of the disease in the first 2 years. Patients can later develop dysfunction in other domains, and late stage PPA patients may not appear that different from patients with other late stage dementias.
References:


**Question 110: Clinical Adult - Infectious Disease**

**Discussion:**

This patient most likely has toxoplasma encephalitis based on history, multiple lesions on imaging, and serology. Fungal or bacterial abscesses, and primary CNS lymphoma, are statistically much less likely. Given the absence of the suggestion of meningitis and lymphoma LP is unlikely to be helpful, and may be frankly dangerous. TB can present with ring enhancing lesions but in this setting usually presents as a meningitis. PPD is positive in 50-80% and chest x-ray is positive in about 50% of adult HIV patients infected with TB. Because of the high likelihood of toxoplasmosis empiric antibiotic therapy with pyrimethamine and sulfadiazine is appropriate. If the patient improves after 2 weeks of therapy, the drugs should be continued for at least 6 weeks. If he fails to improve after 2 weeks, then brain biopsy should be considered to rule out other disorders.

References:


**Question 112: Clinical Adult - Demyelinating Disease**

**Discussion:**

The risk of MS relapses during pregnancy, and the postpartum period, is proportionate to the rate of relapses in the year prior to pregnancy. Since there were 2 relapses in the prior year she remains at higher risk for relapses if she conceives within the next year. This supports the recommendation to continue her MS modulating therapy. Beta-interferon, natalizumab and dimethylfumarate are classified as FDA category C for pregnancy. Each are associated with either embryolethality or teratogenicity in animal studies, although this has not been proven in humans. At this time glatiramer acetate is considered the safest agent to use during pregnancy and lactation. Ideally this individual should be transitioned to GA 1-2 months prior to conception.

References:

Fabian M. Pregnancy in the Setting of Multiple Sclerosis. Continuum Lifelong Learning in Neurol 2016;22(3):837-850

**Question 114: Clinical Adult - Neurology of Systemic Disease**

**Discussion:**

Although the patient's symptoms of pain into the buttock may suggest nerve root irritation as the cause of his problem, specifically pain on internal and external rotation of the hip and occurring only on weight bearing is highly suggestive of a primary problem (such as an impingement syndrome) in the hip. Pain into the groin unilaterally without back or thigh pain from spinal stenosis would be unusual; the vast majority of root compressions are in the L5-S1 distribution. EMG, in the absence of weakness, will have very low yield. Plain radiograph of the hip would be the next appropriate step.

References:


Question 116: Clinical Adult - Neuromuscular Disorders

Discussion:

Spinal bulbar muscular atrophy (Kennedy disease) is characterized by lower motor neuron findings, especially affecting cranial musculature associated with decreased libido and gynecomastia. Female carriers are asymptomatic. Muscle biopsy shows nondiagnostic neuropathic changes. Testicular biopsy shows diminished spermatozoids with abnormal motility and mild elevation of FSH and LH. Diagnosis is by DNA analysis which shows expanded CAG repeats translated into polyglutamine repeats.

References:


Question 119: Clinical Adult - Cerebrovascular Disease

Discussion:

In the absence of clear contraindications, anticoagulation with warfarin is the most appropriate agent for the prophylaxis against recurrent strokes in a patient with atrial fibrillation.

References:


Question 121: Clinical Adult - Epilepsy

Discussion:

The patient presents with neurologically unexplained symptoms. Neither the history, neurologic examination or testing conform to a known neurologic disorder or pattern of neurologic pathology. The diagnosis of a functional disorder is supported by the dissociative non-epileptic seizures that are triggered by panic attacks, and the giveaway weakness on neurologic examination. Neuropsychologic testing can help to document positive findings supporting the diagnosis of a functional disorder (conversion disorder in this case). Pursuing additional testing, such as any of the other answers listed, is not necessary or appropriate, and may reinforce the patient’s perception of a serious underlying neurologic or medical disorder.

References:


Lockhart L, Satya-Murti S. Symptom exaggeration and symptom validity testing in persons with medically unexplained neurologic presentations. Neurology Clinical Practice 2015;5(1);17-24

Question 123: Clinical Adult - Neuromuscular Disorders

Discussion:
Numerous studies and reviews have documented that using steroids early on increases the probability of recovery in patients with new-onset Bell palsy. When used early on, the addition of an antiviral agent to a steroid may modestly further increase the probability of recovery. Antivirals alone have no value in this setting. Neither physical therapy modalities nor facial nerve decompression have been shown to be of value for the treatment of acute Bell palsy. In the absence of a tick bite, erythema migrans rash, positive Lyme serology, or other clinical suggestion of Lyme disease, it is not appropriate to prescribe doxycycline for Bell palsy.

References:
Teixeira LJ, Valbuza JS, Prado GF. Physical therapy for Bell’s palsy (idiopathic facial paralysis). accessed 07/02/2016: www.cochrane.org/CD006283/NEUROMUSC
McAllister K, Walker D, Donnan PT, et al. Surgical interventions for the early management of Bell’s palsy. accessed 07/01/2016:www.cochrane.org/CD007468/NEUROMUSC

Question 128: Clinical Adult - Movement Disorders
Discussion:
The treatment of Parkinson disease is made more challenging by its association with many non-motor problems, including autonomic failure, cognitive and behavioral disturbances and pain. Hyposmia, though typically not noticed or reported by patients later diagnosed with Parkinson disease, is an early-onset problem affecting the majority of patients. Later non-motor problems include orthostatic hypotension and peripheral edema due to autonomic dysfunction and made worse frequently by use of dopaminergic medications, erectile dysfunction in male patients, as well as other urogenital complaints, and later behavioral complications such as visual hallucination.

References:

Question 131: Clinical Adult - Spinal and Root Disorders
Discussion:
This patient has subacute syndrome of pyramidal tract and dorsal column impairment due to copper deficiency. Alpha-tocopherol transfer protein gene mutation would present typically in childhood with gradual onset. A complete blood count might be abnormal, but not diagnostic. MRI of the spine might show increased T2 signal in the dorsal columns in several acquired nutritional deficiencies, but may be normal. Copper deficiency should be considered in patients with myelopathy following gastric surgery, or after zinc overdose.

References:

Question 133: Clinical Adult - Neuro-oncology
Discussion:
Cerebral abscess can be distinguished from recurrent tumor based on differences in diffusion weighted imaging and ADC maps. Both demonstrate a ring-enhancing mass with surrounding T2 hyperintensity representing brain edema. Abscesses are characterized by marked hyperintensity on DWI in the cavity and corresponding hypointensity on ADC imaging. The opposite pattern is seen with recurrent tumor.
References:


Question 139: Clinical Adult - Critical Care/Stroke

Discussion:

Hepatic encephalopathy is characterized by delirium, psychomotor slowing, asterixis, dysarthria and nystagmus. Severe cases are associated with posturing and coma. Laboratory testing can demonstrate abnormalities in liver function tests, ammonia, and clotting. CT head demonstrates generalized cerebral edema and is found in most patients with acute hepatic encephalopathy.

References:


Question 146: Clinical Adult - Epilepsy

Discussion:

Good prognostic features after temporal lobectomy include presence of mesial temporal sclerosis on MRI of the brain, a history of febrile seizures, and mostly unilateral ictal and interictal EEG abnormalities.

References:


Question 150: Clinical Adult - Movement Disorders

Discussion:

Dopa-responsive dystonia (DRD) presents in childhood as a progressive dystonia in children without a history of cerebral palsy or cognitive delay. It typically starts in a foot and progresses to become generalized. The most common inheritance pattern is autosomal dominant. The disease is unique for its robust and sustained response to low doses of levodopa. Its most notable characteristic is a diurnal variation, with symptoms usually more severe towards the end of the day and improved in the morning.

References:


Question 152: Clinical Adult - Demyelinating Disease

Discussion:

Radiologically isolated syndrome, sometimes called preclinical MS, is a recent concept used to describe MRI findings suggestive of MS in the absence of any clinical event consistent with the disease. Its significance lies in the fact that several series have shown that up to one-third of patients with this entity will develop a clinical attack consistent with MS, and the majority of patients will show radiographic progression of the disease in a short time.

References:


Question 159: Clinical Adult - Neurotoxicology
Discussion:

Isoniazid (INH), used in treatment of tuberculosis, depletes pyridoxine by increasing its excretion in urine. It can produce a predominantly sensory axonal neuropathy which is largely preventable with co-administration of vitamin B6 (pyridoxine). Cisplatin, a chemotherapeutic agent in common use for solid organ cancers, binds to and injures DNA, causing a sensory neuronopathy. There is no known preventative treatment. Stavudine is a nucleoside analog still used in some developing countries for treatment of HIV. It produces a painful neuropathy which is not preventable. Docetaxel is a powerful chemotherapeutic agent which interferes with microtubule formation and disrupts axonal transport. Multiple supplements have been tried in an effort to prevent neuropathy from this agent, thus far without success. Vincristine is another common chemotherapeutic agent used primarily in the treatment of leukemias and lymphomas. It interferes with mitosis and microtubule formation, producing a neuropathy which is not preventable.

References:


Question 165: Clinical Adult - Neuromuscular Disorders

Discussion:

Distal Acquired Demyelinating Symmetric Neuropathy is a variant of CIDP that is slower in progression and characterized by predominantly sensory symptoms that are both symmetric and distal. Cases that are associated with an IgM paraprotein tend to be resistant to immunomodulatory therapies.

References:


Question 169: Clinical Adult - Headache

Discussion:

Post lumbar puncture headache is typically a frontal or occipital headache that occurs 12 to 24 hours after lumbar puncture. The headache usually is worse in a standing position and improves when lying down. The use of higher gauge needle, orientation of the bevel parallel to the longitudinal fibers and re-inserting the stylet prior to removal of the spinal needle are all associated with reducing the risk of post-lumbar puncture headaches.

References:


Question 170: Clinical Adult - Demyelinating Disease

Discussion:

Fingolimod interacts with sphingosine-1-phosphate receptor subtypes. Potential side effects include first-dose bradycardia, macular edema, increased hepatic transaminases, blood pressure elevations and mild decrease in one second forced expiratory volume.

References:


Question 178: Clinical Adult - Dementia
Discussion:

The presence of bilateral diffuse and asymmetric corticospinal tract findings is most compatible with ischemic cerebrovascular disease and vascular dementia. The three main neuroimaging patterns in vascular dementia are large vessels stroke, small vessel disease, and microhemorrhages. Small vessel disease usually appears as either multiple bilateral lacunae or as extensive leukoaraiosis with confluent periventricular white matter hyperintensities. Cortical microbleeds are associated with amyloid angiopathy, whereas it is subcortical and periventricular microbleeds that are associated with arteriolosclerosis, subcortical ischemia and vascular dementia.

References:


Question 179: Clinical Adult - Cerebrovascular Disease

Discussion:

Cerebrovascular disease is always a concern in older individuals presenting with acute onset vertigo, especially with underlying stroke risk factors. Brain MRI may be normal within the first 48 hours in cases of posterior circulation stroke. In this setting, the presence of either skew eye deviation, normal head impulse test, or direction changing nystagmus in eccentric gaze, is highly sensitive and specific for acute brainstem or cerebellar stroke.

References:


Questions 183 - 187: Clinical Adult - Headache

Discussion:

1.During pregnancy acute migraine headache management is appropriate. Of those listed, Promethazine is the recommended first line agent during pregnancy. Although ibuprofen can be used for headache management in pregnancy, it is not advisable in the 3rd trimester. Sumatriptan is probably safe but not recommended as a first line agent. Ergotamine can trigger miscarriages and should not be used. 2.Valproic acid is associated with 5-14% known risk of major fetal malformations including neural tube defects. Topiramate has been associated with facial and oral clefts. 3. Amitriptyline can be safely used for migraine prophylaxis during pregnancy. 4.Although there is an increased risk of augmentation with levodopa compared to dopamine agonists, it is the preferred agent during pregnancy. Dopamine agonists have been reported to cause fetal malformations and intrauterine growth retardation in animals 5.Both CYP 450 enzyme inducers (eg, PHT and CBZ) and inhibitors (VPA) have been shown to impair bone health. TPM has not been shown to be deleterious to bone health.

References:

MacGregor EA. Headache in Pregnancy. Continuum Lifelong Learning Neurol 2014;20(1);1280147


Carlson C, Anderson CT. Special Issues in Epilepsy: The Elderly, the Immunocompromised, and Bone Health. Continuum Lifelong Learning Neurol 2016;22(1):246-261

Question 202: Clinical Adult - Dementia

Discussion:
The patient's presentation is strongly suggestive of Creutzfeldt-Jakob disease (CJD). The otherwise unremarkable MRI and normal routine CSF parameters are consistent with CJD. Diffusion-weighted MRI demonstrates restricted diffusion in the cortical mantle and basal ganglia, characteristic of this disorder. Of the biomarkers listed, total CSF tau is the most specific and sensitive, followed by neuron-specific enolase and 14-3-3 protein.

References:


Question 252: Clinical Adult - Spinal and Root Disorders

Discussion:

This patient has a disc space infection with an associated epidural abscess. The triad of back pain, fever, and neurologic deficit is the classic triad of this disorder. Most occur in the thoracolumbar region. Age, diabetes and the history of an invasive procedure (coronary angiogram) are the risk factors in this patient. Most disc space infections are from staphylococcus aureus.

References:


Question 262: Clinical Adult - Headache

Discussion:

This patient has a spontaneous low-pressure headache that is likely due to a dural tear that occurred during her fall. Characteristic features include improvement in the headache while lying down. The radiographic appearance of diffuse dural enhancement is typical.

References:


Question 312: Clinical Adult - Spinal and Root Disorders

Discussion:

The history is suggestive of a cervical myelopathy. A normal jaw jerk localizes a corticospinal tract lesion caudal to the brainstem. MRI findings are characteristic of spondylotic cervical myelopathy with probable spinal cord ischemia (spinal canal<13 mm, fusiform hyperintensity and pancakelike enhancement). Neither the neurologic examination nor MRI imaging are typical for ALS (no UMN signs above LMN signs, presence of MRI enhancement); metastatic cancer (no pain, no edema, temporal profile, intramedullary location, lack of ring or homogenous enhancement pattern); NMO spectrum disorder (absence of area postrema or brainstem syndrome, and of longitudinally extensive transverse myelitis); or B12 deficiency (lack of posterior column and corticospinal tract T2 hyperintensity or atrophy).

References:


Question 317: Clinical Adult - Dementia

Discussion:
Progressive supranuclear palsy is a neurodegenerative disorder that begins with falls and is characterized by vertical gaze palsy, rigidity, dysarthria, cognitive decline and parkinsonian features. Pathologic features include tau positive inclusions that are most commonly found in the basal ganglia. Idiopathic Parkinson disease typically presents with bradykinesia, rigidity and resting tremors. Lewy Body Dementia has parkinsonian features but also has prominent fluctuations in cognition and visual hallucinations. Corticobasal degeneration is a progressive asymmetric movement disorder that presents with abnormalities in one limb or on one side of the body. Multiple system atrophy has parkinsonian features as well but also has dysautonomia, pyramidal signs or cerebellar symptoms.

References:

**Question 318: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

Hypoglycemia can present with signs of focal neurologic dysfunction and mimic stroke. Serum glucose should always be checked prior to the administration of IV t-PA. MRI of the brain, echocardiogram and chest x-ray are not required prior to administration of IV t-PA.

**References:**

**Question 324: Clinical Adult - Epilepsy**

**Discussion:**

Carbamazepine is an appropriate AED for localization related epilepsy. This patient's Asian descent puts her at a 5% risk of fatal skin reactions such as Stevens-Johnson syndrome if she has the HLA-B*1502 allele. Therefore, she should be screened for the HLA-B*1502 allele prior to starting therapy. An MRI of the brain with coronal imaging, EEG, and PET studies are all appropriate for determining seizure etiology, or to help determine if the patient might be a surgical candidate, but they do not preclude starting an antiepileptic drug. Complete IgA deficiency increases the risk of anaphylactic reaction to IVIG infusions.

**References:**

**Question 327: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

This patient is at high risk of developing DVT because of age, immobility, and his prior history of thrombosis. In addition to the use of compression stockings and pneumatic compression devices, anticoagulants are useful in the prevention of DVT in ischemic stroke. Anticoagulants may be used beginning 24 hours after the administration of IV thrombolytic therapy.

**References:**
**Question 333: Clinical Adult - Demyelinating Disease**

**Discussion:**

This patient presents with an acute transverse myelitis of currently unknown etiology. Treatment with high doses of IV corticosteroids is indicated and can be initiated before a full diagnostic workup is complete so long as there is a low suspicion of infection. Plasma exchange, IVIG, and cyclophosphamide may be appropriate second line options depending on response to steroids and the underlying etiology. Beta-interferon is not appropriate for the acute management of this patient even if she were diagnosed with MS.

**References:**

Greenberg BM, Frohman EM. Immune-Mediated Myelopathies. Continuum Lifelong Learning Neurol 2015;21(1):121-131

**Question 335: Clinical Adult - Dementia**

**Discussion:**

Amnestic mild cognitive impairment (aMCI) is, neuropathologically, most often a precursor state to Alzheimer disease. As such, there is medial temporal lobe pathology demonstrated. This is reflected in MRI studies, which have shown decreased hippocampal volume as an independent predictor for the development of dementia in subjects with aMCI.

**References:**


**Question 340: Clinical Adult - Cerebrovascular Disease**

**Discussion:**

Despite limited evidence from randomized controlled trials, initial anticoagulation with heparin is the standard treatment for acute cerebral venous thrombosis. This holds even in the setting of intracerebral hemorrhage or hemorrhagic stroke.

**References:**


**Question 342: Clinical Adult - Neuro-ophthalmology/Neuro-otology**

**Discussion:**

Vestibular migraine (also sometimes referred to as migraine-related dizziness) is a syndrome defined by two or more typical episodes per year of migraine (as per IHS criteria) in concert with severe, but episodic, vestibular symptoms (vertigo, veering), with the migrainous symptoms occurring at the same time as the vestibular symptoms (i.e., temporally related). Neuro-imaging should be normal; vestibular testing, such as VNG (vestibulonystagmography) may be abnormal at times. Meniere disease is not associated with headache, and produces episodes of severe vertigo and nausea as well as aural symptoms which result eventually in hearing loss. Vestibular neuritis typically presents as an acute, debilitating bout of vertigo with severe nausea and a reluctance to move the head. Perilymphatic fistula produces bouts of vertigo with great variability, some very brief, and some lasting for days, or even chronic vertigo. There is frequently a history of trauma and symptoms can be provoked by coughing, sneezing or straining. There is no known association with headache. Basilar-type migraine is a migraine with aura, perhaps on a spectrum with vestibular migraine, but found much more often in adolescents. Additionally, per the current criteria, the aura consists of posterior circulation-type symptoms such as dysarthria, tinnitus, diplopia, visual field abnormalities and reduced level of consciousness.
References:


Question 343: Clinical Adult - Motor Neuron/Nerve

Discussion:

The history and examination conform to a diagnosis of probable ALS by revised El Escorial Criteria (both upper and lower motor neuron signs in at least 2 regions, and UMN signs above LMN signs). The pedigree suggests familial ALS (FALS) with an autosomal dominant inheritance. UBQLN2 has been linked to autosomal recessive FALS. All of the other genotypes are linked to autosomal dominant FALS with variable penetrance. Of these, the C9orf72 mutation is currently thought to be the most common genetic mutation found in FALS world-wide.

References:


Question 344: Clinical Adult - Epilepsy

Discussion:

In a patient with status epilepticus, hypoglycemia should be immediately considered as an underlying etiology, particularly in a patient with diabetes. If serum glucose is not immediately available or the value is uncertain, 50 cc of 50% dextrose should be administered intravenously concurrently with 100 mg of intravenous thiamine.

References:


Question 345: Clinical Adult - Neuro-oncology

Discussion:

Post-transplant acute limbic encephalitis (PALE) is associated with the onset of an amnestic syndrome typically within 60 days of a bone marrow transplant. The clinical picture usually progresses to an encephalitis with refractory seizures. The syndrome is usually caused by human herpes virus 6 and should be treated with anti-viral therapy.

References:


Question 351: Clinical Adult - Neuro-ophthalmology/Neuro-otology

Discussion:

Idiopathic Intracranial Hypertension presents most commonly in obese women of child-bearing age and is characterized by bilateral symmetric papilledema. In order to make a diagnosis, other causes of increased intracranial pressure should be excluded by performing an MRI brain with gadolinium to evaluate for mass lesion and MR venography to evaluate for cerebral venous thrombosis.
References:


**Question 354: Clinical Adult - Infectious Disease**

**Discussion:**

When insufficiently treated, acute Lyme disease can result in chronic neuroborreliosis. Although acute Lyme disease can be a cause of 7th nerve palsy, there are other etiologies for the same. A tick bite alone does not objectively confirm a diagnosis of Lyme neuroborreliosis. Acute brain MRI eliminates a structural CNS lesion but not Lyme neuroborreliosis. The next step is to objectively confirm whether in fact he has active Lyme disease. This is best accomplished by first checking serum Lyme ELISA with reflex Western Blot. In order to have Lyme neuroborreliosis he must have had systemic Lyme disease, which would be supported by findings of elevated serum ELISA and positive IgG Western Blot. If the abnormal presence of these antibodies is confirmed then appropriate treatment with antibiotics, using CSF/serum Lyme index as a guide, should be pursued. If these are negative, then treatment for Lyme neuroborreliosis is without foundation and another etiology for his complaints needs to be sought. SPECT brain imaging has not been shown to be either sensitive or specific for the diagnosis of Lyme neuroborreliosis.

**References:**


**Question 358: Clinical Adult - Epilepsy**

**Discussion:**

Sudden unexplained death in epilepsy (SUDEP) remains poorly understood. Studies suggest that the most significant risk factors include uncontrolled generalized tonic-clonic convulsions, higher frequency of seizures, and the need for multiple antiepileptic drugs. Absence seizures, nonconvulsive seizures, and etiology of the epilepsy have no known bearing on the risk of SUDEP.

**References:**


**Question 359: Clinical Adult - Neuro-ophthalmology/Neuro-otology**

**Discussion:**

The history and examination is most consistent with benign positional vertigo. The other disorders listed would be associated with additional neurological deficits and/or the vertigo would be less likely to be associated with positional changes.

**References:**


**Question 360: Clinical Adult - Sleep**

**Discussion:**
This patient describes cataplexy which is a disorder characterized by the sudden loss of lower limb tone with preserved consciousness. Attacks can be triggered by laughter, anger or shock. Cataplexy is associated with excessive daytime sleepiness, sleep paralysis, and hypnagogic hallucinations. Cataplectic manifestations may be subtle and consist of mild slurring of speech, buckling of knees, jaw dropping or head nodding. Seizures, orthostatic hypotension and hypokalemia may cause transient weakness or light-headedness and are not typically provoked by laughter or other intense positive emotions.

References:

Question 362: Clinical Adult - Movement Disorders

Discussion:
This patient describes cervical dystonia. So-called "sensory tricks" (geste antagoniste), usually involving touching the chin, back of the neck or cheek, have reportedly alleviated these symptoms in up to 90% of patients with cervical dystonia.

References:

Question 366: Clinical Adult - Neuromuscular Disorders

Discussion:
The history and examination are suggestive of a rapidly progressive parainfectious polyneuropathy. The CSF is consistent with acute inflammatory demyelinating polyneuropathy (AIDP). The patient's prior history, CSF data, pain, and other features make the other diagnoses less likely than AIDP.

References:

Question 370: Clinical Adult - Neurogenetics

Discussion:
The patient described most likely has Congenital Myotonic Dystrophy Type 1, based on the clinical presentation of myotonic facies, hypotonia and swallowing problems. His mother, never previously diagnosed, has clinical myotonia, suggesting the phenomenon of anticipation wherein the mother has a much milder genetic defect -- the CTG trinucleotide expansion --and can have a severely affected child. Non-dystrophic myotonias are caused by mutations in voltage-gated ion channels; a chloride channel gene mutation causes diseases such as Myotonia Congenita and a sodium channel mutation is responsible for both paramyotonia congenita and hyperkalemic periodic paralysis. Acid maltase deficiency, in its severe infantile form, is known as Pompe disease and produces signs and symptoms consistent with glycogen deposition, such as hepatomegaly and macroglossia. X-linked mutations in the dystrophin gene are linked to Muscular Dystrophy.

References:
Question 372: Clinical Adult - Spinal and Root Disorders

Discussion:

Lumbar puncture is a safe and well tolerated procedure. Complications are rare and include post-lumbar puncture headache, cerebral herniation, infection, bleeding and radicular pain. There is no increased risk of bleeding in patients on anti-platelet therapy. In patients with bacteremia or hematologic malignancy, there is no proven risk of introducing infection or cancer into the CSF with a spinal needle. Spinal stenosis may make obtaining CSF challenging but is not a contraindication. In patients with suspected spinal epidural abscess, a lumbar puncture can introduce infection into the subarachnoid space if the needle is passed through the abscess.

References:


Question 374: Clinical Adult - Cerebrovascular Disease

Discussion:

The ECASS 3 trial demonstrated the benefit of IV tPA between 3 and 4.5 hours after symptom onset when compared to placebo. The addition of endovascular treatments, including Intra-arterial tPA and mechanical thrombectomy have not demonstrated superiority to IV tPA alone in several recently published randomized trials (SYNTHESIS, MR RESCUE and IMS III). Current evidence supports the use of IV tPA in eligible patients within 4.5 hours of symptom onset.

References:


Question 376: Clinical Adult - Critical Care/Stroke

Discussion:

All of the above parameters have been studied in depth as a predictor of poor neurologic outcome after hypoxic-ischemic brain injury (HIBI). In patients not treated with hypothermia protocols, absence of pupillary or corneal reflexes at 3 days showed a 0% false positive rate (FPR) in predicting poor neurologic outcome, while measurement of neuron-specific enolase has been shown to require much higher values than previously thought (greater than 80 micrograms/L) to achieve a 0% FPR. Additionally, the measurement of NSE may not be sufficiently standardized or readily available in some labs. The use of techniques combining DWI on MRI with other quantitative measurements is improving the prognostic value of MRI in HIBI, but these techniques are not yet accepted as having 0% FPR. In a prospective study best motor response was found to have up to a 24% FPR; finally, EEG has only Level C evidence in the absence of other prognostic factors as a predictor of poor prognosis after HIBI.

References:


Question 389: Clinical Adult - Neuromuscular Disorders

Discussion:

Both GBS and CIDP are acquired demyelinating autoimmune neuropathies. The CSF and electrodagnostic findings are quite similar, and cannot distinguish one from the other. The interval between onset of symptoms and disease plateau is the main distinguishing feature, with the vast majority of GBS patients reaching a plateau within four weeks of disease onset.
Juvenile myoclonic epilepsy is a generalized epilepsy that presents during late childhood or early adulthood. Classic presentation is often in the setting of sleep deprivation or alcohol exposure. Patients have characteristic myoclonic jerks in the early morning hours. EEG demonstrates a 6 per second spike-and-wave pattern. Treatment with valproate or lamotrigine is indicated once a diagnosis is made and is effective in most patients.

References:

West Nile Virus typically presents with fever, headaches and rash. Neurological manifestations include meningitis, encephalitis and in rare cases, an acute flaccid paralysis that involves the anterior horn cells similar to poliomyelitis.

References:

Mutations of filamin A have been linked to the presence of bilateral periventricular heterotopias, enlarged cistern magna and thinning or agenesis of the corpus callosum. In adolescents and adults, screening for an aortic aneurysm should be performed. Molar tooth sign is associated with Joubert’s syndrome; Leptomeningeal angioma is observed with Sturge Weber Syndrome. Schizencephaly is also a neuronal migration disorder with expected association of seizures and developmental delay.

References:

Myoclonic epilepsy of infancy (MEI) is a generally benign generalized myoclonic seizure disorder occurring in otherwise healthy infants, some of whom (about 25%) have a history of isolated febrile convulsions. They do not occur in clusters, and are not associated with tonic-clonic seizures.

References:

**Question 36: Clinical Pediatrics - Neuromuscular**

**Discussion:**

Only daily oral corticosteroids have been shown to prolong ambulation in Duchenne muscular dystrophy (DMD). The effect is to induce small amounts of dystrophin, not as an anti-inflammatory treatment, so other dosing strategies are not helpful. Intravenous gentamicin has been proposed for one of the rare point mutations causing DMD. Trials of oral carnitine and valproic acid are underway in SMA.

**References:**


**Question 39: Clinical Pediatrics - Neonatal**

**Discussion:**

Zellweger syndrome presents in infancy, and affected individuals have characteristic facial features with high forehead, hypoplastic supraorbital ridges, and midface hypoplasia. Severe weakness, hypotonia, and seizures are common. Diagnosis is confirmed by elevated levels of saturated and unsaturated very long-chain fatty acids in body fluids.

**References:**


**Question 45: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders**

**Discussion:**

Unilateral closed-lip schizencephaly carries nearly a 70% risk for epilepsy.

**References:**


**Question 47: Clinical Pediatrics - Vascular and Inflammatory Disorders**

**Discussion:**

Cerebrovascular disease occurs in 25% of sickle cell disease. Eighty percent of events occur at or before age 15 and most are caused by a progressive cerebral vasculopathy that can be partially arrested by chronic transfusion therapy to keep hemoglobin S below 30%. Most events are thrombotic.
Discussion:

Cerebellar mutism, also called posterior fossa syndrome, is a complication of any surgical procedure within the posterior fossa, but more likely to occur following surgeries that involve tumor resection, particularly if the vermis is involved. While various cranial nerve palsies may be seen, the syndrome is also seen without other evidence of structural damage. Typically, onset of severe dysarthria progressing to mutism is delayed hours to days after initial surgical recovery. Severe irritability is common. Recovery takes weeks to months and may never be complete, as many children with this manifestation show ongoing cognitive and behavioral problems years after successful surgery.

References:


Discussion:

Frequent seizures with a mildly slow EEG are the hallmark of catscratch encephalitis. Lyme disease is not associated with seizures; Rocky Mountain spotted fever and the arboviruses usually have fever and prominent meningeal signs. Pneumococcal meningitis would present with meningeal signs and a higher CSF WBC count and increase protein would be expected.

References:


Discussion:

Perinatal stroke occurs in about 1/4000 live births. It is frequently overlooked until children are sitting or walking. There is variable outcome, with some having primarily hand weakness, but many will have leg involvement, seizures, or learning disability. Erb and Klumpke palsies involve intrinsic hand muscle or wrist extension, not fisting. Schizencephaly is rarer; it can present with hemiparesis, but occurrences usually have more severe symptoms.

References:


The combination of dysconjugate, highly variable nystagmus, head nodding and head tilt, without ophthalmologic abnormalities, and with normal neuroimaging, is diagnostic of spasmus nutans. Latent nystagmus is a jerk nystagmus that is evoked or enhanced by covering one eye. Congenital nystagmus is usually conjugate and suppressed by convergence (in contrast to spasmus nutans, which is typically increased by convergence). Opsoclonus describes chaotic, conjugate saccades, seen classically in the paraneoplastic syndrome associated with neuroblastoma. Decompensated strabismus would not produce a paroxysmal movement disorder.

References:


Question 66: Clinical Pediatrics - Epilepsy

Discussion:

Benign idiopathic neonatal convulsions are an autosomal dominant disorder which has been strongly linked to potassium channel defects resulting from mutations of the genes KCNQ2 and KCNQ3. The normal exam makes hypoxia unlikely as most are lethargic. Most children with metabolic disorders are lethargic or even comatose. Benign rolandic epilepsy presents in adolescence.

References:


Question 70: Clinical Pediatrics - Neuromuscular

Discussion:

Spinal muscular atrophy I usually presents with hypotonia, areflexia, paradoxical respiration with narrow upper chest, tongue fasciculations, and progresses to difficulty swallowing, then respiratory distress. While infants with cerebral palsy may be initially hypotonic, they are generally hyperreflexic. Infants with infantile Gaucher disease generally present with stiffness, not initial hypotonia, and have organomeglyal at onset. Muscle stretch reflexes in children with a congenital myopathy are usually present but may be difficult to elicit due to weakness.

References:


Question 75: Clinical Pediatrics - Infectious Disease

Discussion:

Group B streptococci causes almost 50% of cases of neonatal bacterial meningitis and is the most common organism. Escherichia coli and Listeria monocytogenes are the second and third most common organisms, respectively.

References:


Question 91: Clinical Pediatrics - Movement Disorders

Discussion:

The darting random eye movements are consistent with opsoclonus, and the rapid presentation and symptom complex is most consistent with opsoclonus-myoclonus-ataxia syndrome. Typical nystagmus with rapid lateral eye movements is seen in other forms of acute ataxia such as postinfectious, Ataxia telangiectasia and neuropathy, ataxia, retinitis pigmentosa, and ptosis (NARP) do not have opsoclonus and have slower onset.

References:


Question 104: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

The typical presentation of a medulloblastoma is a midline posterior fossa mass which is hyperdense on noncontrast CT and enhances uniformly with contrast. On MRS, choline and taurine are increased. The clinical presentation is often due to the secondary hydrocephalus. Pilocytic astrocytomas also tend to occur in the posterior fossa but generally have a significant cystic component. An ependymoma would tend to be more clearly associated with the ventricle as would a choroid plexus papilloma.

References:


Question 113: Clinical Pediatrics - Epilepsy

Discussion:

The combination of tonic spasms and a burst suppression EEG is consistent with the syndromic diagnosis of early infantile epileptic encephalopathy (Ohtahara syndrome). This syndrome, which can also include fragmentary myoclonic jerks and partial seizures, frequently evolves into West syndrome at about 4 to 6 months of age. Etiological causes have included brain malformations as well as metabolic syndromes, such as glycine encephalopathy and mitochondrial disorders.

References:


Question 130: Clinical Pediatrics - Infectious Disease

Discussion:

Tuberculous meningitis typically presents in toddlers with initially nonspecific febrile illness but if not recognized progresses to hydrocephalus and infarcts in the region of basilar perforators. Unfortunately, when diagnosis is delayed, prognosis is poor.
Question 135: Clinical Pediatrics - Neuromuscular

Discussion:

Infantile botulism usually presents between 3 and 18 weeks of age. The disease is caused by the Clostridium botulinum toxin, which blocks acetylcholine release. Clinical features include constipation, hypotonia, areflexia, poor suck, impaired pupillary response to light, and ophthalmoplegia. The infants are often breast-fed. Diagnosis is made by EMG with repetitive nerve stimulation, causing an incremental response and isolation of C. botulinum toxin in the stool.

References:


Question 141: Clinical Pediatrics - Cerebral Palsy

Discussion:

Cerebral palsy is a clinical diagnosis that describes a nonprogressive delay in motor development. A diagnosis of cerebral palsy does not define its etiology. This clinical picture describes the classic tetrad of chronic bilirubin encephalopathy (kernicterus): severe athetosis, perceptive high tone deafness, supranuclear palsy of upward gaze, and enamel hypoplasia of the incisor teeth.

References:


Question 161: Clinical Pediatrics - Epilepsy

Discussion:

The nocturnal seizures associated with autosomal dominant nocturnal frontal lobe epilepsy are typically brief and consist of an aura that has been described as fear or forced thinking with associated hyperkinetic or tonic movements. Consciousness can be maintained during an event. These events do tend to cluster. The disorder has an autosomal dominant inheritance pattern and is associated with mutations in the neuronal nicotinic acetylcholine receptor.

References:


Question 176: Clinical Pediatrics - Neuromuscular
Discussion:

Hallmarks of Friedreich ataxia are a combination of weakness, ataxia, absent tendon jerks, Babinski sign, and abnormal position sense. Romberg sign is the best indication of loss of position sense, not cerebellar functioning. Hammer toes, pes cavus deformities, and scoliosis are frequent findings.

References:


Question 177: Clinical Pediatrics - Epilepsy

Discussion:

The seizure pattern described is one of benign rolandic epilepsy, for which the EEG signature is central-temporal spikes, particularly in sleep, often bilateral independent.

References:


Questions 188 - 192: Clinical Pediatrics - Headache/Other Paroxysmal Disorders

Discussion:

Night terrors typically occur in deep sleep, often 90 minutes to 3 hours after sleep onset. The child appears awake but is not. Autonomic symptoms are prominent. The child has no memory of the event.

Masturbation by infants is not often recognized immediately, causing excessive diagnostic testing. While most often seen in infant and toddler girls, boys may have similar manifestations. Usually the behavior is specific to the setting (often in highchair or car seat) but may also occur with the child lying on the floor, often with the legs extended or crossed. Facial flushing and sweating is common. Although the behavior can be interrupted, the children typically return to it as soon as they are not deterred.

Paroxysmal kinesiogenic choreoathetosis presents with unilateral choreic or dystonic movements with initiation of voluntary movement such as walking. There is no alteration of consciousness.

Cyanotic breath-holding spells occur in infants after sudden pain or fright. Brief convulsion may be precipitated by the syncope.

Hemiplegic migraine is often dominantly inherited, consists of hemiplegia followed by headache and vomiting. Some are mapped to a calcium channel gene.

References:


Question 213: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:
This clinical picture describes a typical presentation and MRI findings of Joubert syndrome; in which there is hypoplasia of the cerebellar vermis. The most common features of this syndrome include hyperpnea, hypotonia, oculomotor apraxia, ataxia, and intellectual disability. Other neurologic manifestations include seizures. The molar tooth sign, which is the result of the thickening and horizontalization of the superior cerebellar peduncle and a deep interpeduncular fossa, can also be seen in several other disorders including Dekaban-Arima syndrome, Senior-Loken syndrome, and COACH (cerebellar vermis hypoplasia, oligophrenia, ataxia, coloboma, and hepatic fibrosis).

References:


Question 215: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:

The conjunctival biopsy shows typical curvilinear bodies. This strongly suggests late-infantile neuronal ceroid lipofuscinosis (LINCL). Visual loss in LINCL often follows seizure onset by several years, and is accompanied by gradual progression of dementia. In contrast, juvenile NCL generally presents with visual loss, followed years later by seizures and dementia. Cardiomyopathy commonly occurs with several other neurodegenerative conditions including mitochondrial cytopathies and Friedreich Ataxia. Skin bronzing and adrenal insufficiency are hallmarks of adrenoleukodystrophy, while seizures are not common.

References:


Question 224: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

Dermal sinus tracts are located above the gluteal cleft and course cephalically from the skin, in contrast to coccygeal pits, which are much more common, located within the gluteal cleft, course caudally or straight down from the skin, and are not associated with intradural pathology. The tract may connect to a dermoid, either within or adjacent to the cord. Excision of the full tract, not just the skin component, is essential. As the lesion commonly connects through the dura into the spinal cord, surgery should be done by an experienced pediatric neurosurgeon, after appropriate imaging. Early excision avoids the risk of infection with subsequent significant neurologic deficits, as seen in this case.

References:


Question 257: Clinical Pediatrics - Movement Disorders

Discussion:

The MRI demonstrates a characteristic "eye of the tiger" with low T2 signal centered in a high T2 signal area. The characteristic dystonic movements in PKAN2 may follow an initial period of apparent spasticity. Oromandibular dystonia is prominent. Huntington disease presents with prominent dystonic features in children, rather than chorea, but the MRI usually shows atrophy of caudate as well as cortical atrophy. Although girls with Rett syndrome will lose purposeful movements of their hands and have progressive gait disturbance, dystonia is not a prominent feature. Patients with dopa-responsive dystonia often have early findings suggestive of spasticity, progressing to dystonia, but typically have normal MRI.

References:
Metachromatic leukodystrophy is an autosomal recessive disorder, and in its imaging usually demonstrates involvement of the periventricular and deep white matter with relative sparing of the U-fibers. Neurocognitive symptoms can be the primary presentation in the late juvenile onset form, with relatively subtle signs on the neurological examination (long tract signs alone are common).

References:


The lesion pictured is a depigmented macule associated with tuberous sclerosis. Of the choices, only subependymal nodular lesions are associated with tuberous sclerosis. Plexiform neurofibromas, gliomas of hypothalamus and optic chiasm, and punctate "UBOs" on MRI, are associated with neurofibromatosis 1 (NF1); schwannomas on cranial nerves are associated with NF2.

References:


Chemotherapy used for leukemia, particularly l-asparaginase, may cause significant hypercoagulability, particularly secondary anti-thrombin III deficiency, which this patient had. Venous sinus thromboses, with infarctions in a venous distribution, are the result.

References:


The presence of a facial angioma in the distribution of the ophthalmic division of the trigeminal nerve, in a child who presents with a unilateral, throbbing headache and neurologic deficits, is suggestive of the diagnosis of Sturge-Weber syndrome. In this syndrome, leptomeningeal angioma may represent an embryonic remnant of the venous plexus that failed to regress. The leptomeningeal angioma is most frequently unilateral, overlying the parietal and occipital lobes, and ipsilateral to the facial angioma. The presence of leptomeningeal angioma confirms the diagnosis of Sturge-Weber syndrome, which is often complicated by headache, stroke-like episodes, hemianopsia, glaucoma, and intellectual disability.
Question 334: Clinical Pediatrics - Headache/Other Paroxysmal Disorders

Discussion:
This is a classic presentation of benign paroxysmal vertigo in a child, a syndrome that occurs in children in the first 5 years of life and is a frequent precursor of migraine. Investigations (ie, MRI, EEG) are normal. The episodes usually resolve by the end of the first decade.

References:


Question 336: Clinical Pediatrics - Epilepsy

Discussion:
Lacosamide was approved by the Food and Drug Administration in 2008 as adjunctive therapy for focal-onset seizures in patients 17 years of age and older. Patients and their families should be aware that high doses of lacosamide (300 to 800 mg) in adults have produced a mild euphoria. Although reported in less than 1% of patients enrolled in clinical trials, the risk for abuse resulted in lacosamide being approved as a schedule V controlled substance in the US.

References:

Question 341: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders

Discussion:
Rett syndrome is seen in young girls and is characterized by developmental arrest, stereotypic hand movements, deceleration of head growth and loss of communication skills. It is an X-linked disorder and 80% to 85% of classic Rett syndrome patients will have an abnormality in the MECP2 gene, while approximately 50% of atypical Rett patients will have an abnormality of this gene.

References:
Niemann-Pick disease type C is characterized by autosomal recessive inheritance with gradual and progressive neurologic deterioration. Patients have variable hepatosplenomegaly, and progressive neurodegeneration. Findings include vertical supranuclear (usually downgaze) palsy, ataxia, dystonia, seizures, gelastic cataplexy, and dementia.

References:


Question 368: Clinical Pediatrics - Neonatal

Discussion:

At birth in the healthy full term neonate, the posterior limb of the internal capsule should be myelinated. Between 2 and 4 months of age, there should be evidence on MRI of myelination of the splenium of the corpus callosum, optic radiations, and cerebellar white matter. Myelination in the frontal white matter is not evident until between 4-6 months of age.

References:


Question 383: Clinical Pediatrics - Hereditary and Metabolic Disorders

Discussion:

This clinical picture describes a typical presentation of Smith-Lemli-Opitz Syndrome. Brain abnormalities in this disorder of cholesterol synthesis (mutation of the 3-b-hydroxysterol-d-7 reductase gene) range from microencephaly, hypoplasia of the frontal lobes (holoprosencephaly), abnormal gyration, and cerebella hypoplasia.

References:


Question 386: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders

Discussion:

Meador and colleagues found that children at 3 years of age exposed to valproic acid in utero had significantly lower IQ scores than children of the same age who had in utero exposure to carbamazepine, lamotrigine, and phenytoin.

References:


Question 393: Clinical Pediatrics - Neurosurgery, Critical Care and Tumors

Discussion:

The President's Commission recommended 100% oxygen ventilation for 10 minutes, followed by passive 100% oxygen for a period long enough to achieve a PCO2 of 60. The other tests have high incidence of false-negative results.
References:

**Question 394: Clinical Pediatrics - Hereditary and Metabolic Disorders**

**Discussion:**

Menkes disease (trichopoliodystrophy) is a disorder of copper transport resulting from a mutation in a copper transporting adenosine triphosphatase. Clinical features include hypotonia, poor feeding, and medically refractory seizures. The expected laboratory finding to confirm the diagnosis is a low serum copper and ceruloplasmin level.

References:

**Question 397: Clinical Pediatrics - Develop, Learn, Lang, Behav, Psych Disorders**

**Discussion:**

Most children with Angelman syndrome have little or no spoken language. Sign language may amplify communication. The other conditions tend to be associated with milder disorders or peculiarities of language.

References:

**Contemporary Issues**

**Question 1: Contemporary Issues - Ethics/Professionalism**

**Discussion:**

In a nonemergent situation, the parents are the directors of their children’s health care. There is not a compelling reason to contradict their wishes emergently with regard to imaging in this case. In cases where their decisions would have deleterious medical consequences, ethics consultation is appropriate. Lumbar puncture should not be performed in a patient with suspected mass lesion (new seizure, hemiparesis) without imaging studies done first.

References:

**Question 23: Contemporary Issues - Ethics/Professionalism**

**Discussion:**

The patient has decision making capacity and has the autonomy to determine interventions or lack of interventions. The patient's desire to stop eating and drinking does not give the physician the right to intervene without the patient's consent to do so. The patient's spouse cannot direct care for a patient with capacity to make their own decisions, unless previously determined by advance directive.

References:
Question 33: Contemporary Issues - Practice

Discussion:

When a patient is unable to communicate his or her medical history, its value is limited. The neurologic examination also cannot be as complete if the patient is not able to understand the commands given or report on sensory testing. Cultural competency involves utilizing information about language and heritage to best treat a patient. An interpreter with training in medical information is the best interpreter for assessing and translating patient information. A relative may not be able to provide the detail needed to determine a diagnosis accurately and important details may not be understood or accurately communicated. Conducting the interview in English will provide very limited results. While old records may be useful they will most likely not give the complete story.

References:

Cultural Competency in Medicine. 7/14/09 http://www.amsa.org/programs/gpit/cultural.cfm

Question 46: Contemporary Issues - HIPAA

Discussion:

A health care provider or health plan may share relevant information with family members or friends involved in the patients’ health care or payment for health care, if you tell the provider or plan that it can do so, or if you do not object to sharing of the information. Even if family or friends accompany a patient to an appointment, this does not constitute approval to share medical information without the patient's knowledge at a later date directly to those individuals.

References:

US Department of Health and Human Services Health Information Protection. 7/24/09 http://www.hhs.gov/ocr/privacy/hipaa/understanding/consumers/familyfriends.html

Question 52: Contemporary Issues - Practice

Discussion:

Management of antiplatelets and anticoagulants perioperatively must be done weighing the risk of a thromboembolic event (TE) versus the risk of bleeding. For most dental and many dermatologic procedures, these medications can be continued. When anticoagulants are stopped, the risk of TE increases significantly after 7 days. Bridging with heparin has shown an increase in bleeding complications.

References:


Question 58: Contemporary Issues - Ethics/Professionalism

Discussion:

Websites displaying feedback from patients about their physicians are readily available. The best course of action is to seek to understand reasons behind negative comments instead of taking more escalated actions such as firing patients or bringing a lawsuit. Making a comment may be considered illegal from the standpoint of HIPAA laws.

References:


Question 68: Contemporary Issues - Evidence-based Medicine
Discussion:
The sensitivity is the number of true positives over true positives plus false negatives. In this case, that is 95/95+5 or 95%. The false positive rate is 1% (controls with an abnormal NCS) and false negative rate is 5% (normal NCS in patients with the gold standard clinical diagnosis of CTS). Specificity is the number of true negatives over true negatives plus false positives (99/99+1) or 99%.

References:


Question 103: Contemporary Issues - Driving

Discussion:
The clinical dementia rating (CDR) is a measure of severity of dementia and it has been shown to associate with decreased driving ability as judged by the ability to pass an on-road driving test. A CDR of 1.0 is more suggestive than a CDR of 0.5. The other factors listed may contribute, however, the evidence is less robust.

References:


Question 117: Contemporary Issues - Business

Discussion:
Time can be used for billing any visit, but more than 50% of the total billable time has to be spent in counseling or coordination of care. So, if 40 of the 60 minutes was spent in that fashion, it would qualify for billing by time (60 minutes). Established patients are defined as having been seen in the same department in the last 3 years. Within that timeframe, an established code should be used; outside of that timeframe a new code should be used. Although the complaint is focused, the medical decision making could be complex such that a higher level of coding could still be appropriately achieved.

References:


Question 136: Contemporary Issues - Evidence-based Medicine

Discussion:
Relative risk, odds ratios and risk differences are common measures of association in epidemiological studies. These values measure the amount of variation in the development of the disease that is explained by the presence or absence of the risk factor. Statistical significance, confidence intervals and p-values only measure the amount of random error in a study. They relate to the probability that the observed association can be explained by chance but do not inform you of the strength of the association.

References:

Question 147: Contemporary Issues - Practice

Discussion:

Informed consent must be documented in the medical record and include an explanation of the procedure with risks and benefits and alternatives to the procedure. It must be performed by an individual with knowledge of the procedure and its complications. It must be done with an adult or a delegate who has the capacity to make decisions. This applies to routine procedures (as in this case) but can be bypassed in more emergent situations.

References:


Question 158: Contemporary Issues - Business

Discussion:

Overbilling or under billing is considered Medicare fraud. HIPAA covers patient privacy and security of medical information.

References:


Question 320: Contemporary Issues - Practice

Discussion:

This is Bell’s palsy. There is Class 1 evidence supporting treatment with steroids to improve recovery. There is weak evidence for antiviral treatment. There is no benefit to combination therapy. The upper and lower facial weakness is unlikely to be a stroke.

References:

Gronseth, G and Peduga, R. Evidence-Based Guideline; steroids and antivirals for Bell's palsy. Evidence-Based Guideline; steroids and antivirals for Bell's palsy. Neurology 2012;79:15

Question 326: Contemporary Issues - HIPAA

Discussion:

As much as we sympathize with the plight of a family member, we must adhere to the patient’s wishes, especially as she is a mentally competent individual. This is the principle of confidentiality and is protected under HIPAA laws.

References:


Question 387: Contemporary Issues - Practice

Discussion:

Direct patient care in a state one does not hold a license may be considered illegal and would not be a billable service. The exchange of medical information and advice, however, directly to a licensed physician in that state is allowed assuming they then use your advice and continue to direct care themselves. Each state licenses its physicians and the risk of practicing into other
states from a legal standpoint varies by state. Interstate compacts are being evaluated which will only allow licensing to be expedited in other states.

References:
Lori A. Boyajian-O’Neill, DO; Lindsey M. Gronewold, MS; Alan G. Glaros, PhD; Amy M. Elmore, DO. Physician Licensure During Disasters: A National Survey of State Medical Boards. http://jama.jamanetwork.com/article.aspx?articleid=1149358

Neuroimaging

Question 204: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:
The MRI reveals too much white matter disease for Alzheimer disease. Metachromatic leukodystrophy typically produces more diffuse and confluent areas of demyelination by this age. Progressive multifocal leukoencephalopathy is less common, and large regions (rather than small plaques) of white matter signal change are expected. Given the diffuse white matter lesions that are characteristic of multiple sclerosis plaques, her age, and gender, multiple sclerosis is the most likely diagnosis. Vasculitis typically involves the gray/white matter junction.

References:

Question 205: Neuroimaging - Tumors/Cysts

Discussion:
The images show a cystic cerebellar hemispheric mass with an enhancing mural nodule. T2-weighted images suggest the presence of small vessels within the mural nodule as hypointense flow voids. The mass is intra-axial and infratentorial. The differential diagnosis includes ganglioglioma, medulloblastoma, cystic astrocytoma, and abscess. Demyelinating plaques may present as a mass lesion, but this is unusual and would present with a ring of enhancement, not this pattern. The finding is most consistent with a cerebellar cystic hemangioblastoma.

References:

Question 206: Neuroimaging - Spine

Discussion:
The CT scan of the thoracic spine demonstrates a hyperdense mass within the spinal canal with involvement of the adjacent posterior spinous process typical of a lytic lesion. The T2-weighted MRI similarly demonstrates infiltrating of ovoid mass extending into the posterior vertebral body within the spinal canal and within the posterior bony elements of the soft tissue. A hematoma can present as an ovoid hyperintense mass; however, one would not expect lytic destruction within the posterior spinous process or involvement of the posterior vertebral body; nor would these features be seen with an ependymoma or meningioma. Neurofibromas are slow-growing expansile lesions and result in remodeling of bone, particularly neural foramen and not a lytic mass as shown. Of the choices, a metastatic tumor is the most likely.

References:
Question 208: Neuroimaging - Critical Care/Stroke

Discussion:

The imaging of this patient demonstrate no obvious ischemic changes on the non-contrasted CT and a right M1 occlusion on the CT angiogram. The CT perfusion further confirms the vascular territory at risk of infarction strongly indicating the need for emergent intervention with TPA and/or clot retrieval techniques.

To standardize CT examination with producible grading system to assess early ischemic changes on pre thrombolytic treatment with acute ischemic stroke of the anterior circulation The Alberta ASPECTS scoring system was developed. The score divides the MCA territory into 10 different regions with 1 point is deducted from the initial score of 10 for every region demonstrating ischemic changes on a non-contrasted CT head. Patients with ASPECTS score less than 8 treated with thrombolysis do not a good clinical outcome and are at increased risk of hemorrhagic transformation. The imaging of this patient demonstrate no obvious ischemic changes on the non-contrasted CT and therefore has a 0.

References:


Question 210: Neuroimaging - Critical Care/Stroke

Discussion:

The CT angiogram shows abnormally dilated and tortuous vessels near the tentorium. These vessels are part of an arteriovenous malformation. The symmetric thalamic edema is the result of venous hypertension and impaired venous outflow.

References:


Question 211: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

The figures depict a large fluid-filled cyst within the right hemisphere with absence of much of the right hemisphere. This is not associated with a margin of gliotic changes nor is there evidence of a craniotomy defect as would be expected with a hemicraniectomy. The signal characteristics are that of CSF on all sequences, which is inconsistent with a chronic subdural hematoma. Epidermoid typically has bright signal on a diffusion scan sequence, not seen here. Of note, there is marked asymmetry to the dimensions of the right as compared to the left hemispheric calvarium indicating a long standing and possibly neonatal process. Aliasing artifact produces a wrap-around of images due to an inadequate field of view not seen on this study.

References:

The non-contrast CT demonstrates an ovoid hyperdense focus within the superior sagittal sinus typical of a thrombus and similar hyperdensity within the right transverse sinus. The appearance is that of an acute sagittal sinus thrombosis. On the CT angiographic views there is decreased contrast in the superior and right transverse sinuses due to thrombus. There is no evidence of subarachnoid hemorrhage. The hyperdensity is within the dural sinuses as compared to a subdural hematoma which would be external to the sinus. There is no evidence of cerebral edema as seen with eclampsia. An empyema would appear as a hypodense subdural collection and enhance.

References:


Question 214: Neuroimaging - Infection

Discussion:

Axial CT, T2, and post contrast T1 weighted images show an asymmetric nonenhancing lesion in the right frontal lobe with no mass effect. The lesion primarily affects the white matter. This is typical for PML.

References:


Question 216: Neuroimaging - Tumors/Cysts

Discussion:

The images depict a diffusely enhancing mass encircling the right optic nerve sheath and arising extra-axially in the ventral frontal area with a dural tail. Lymphoma, sarcoïd and tuberculosis may arise along the meninges in and have homogeneous enhancement however, a three-year history and absence of a significant response to steroids would be unlikely. Optic neuritis would result in enhancement of the nerve and not the sheath.

References:


Question 217: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

There is marked enhancement of the thecal sac and cauda equina. This is diffuse and not nodular. A 2-year history of intermittent weakness would be inconsistent with carcinomatosis or leptomeningeal seeding from a ependymoma. Cysticercosis can be associated with dural enhancement but with cyst formations would be expected, not seen on this study. Tuberculosis and sarcoïd can produce dural or leptomeningeal enhancement however a relapsing course without treatment would be inconsistent. Both acute inflammatory polyradiculitis and chronic inflammatory demyelinating polyradiculitis (CIDP) are frequently associated with leptomeningeal thickening and enhancement as seen on this study. The clinical history would be most consistent with CIDP.

References:


Question 219: Neuroimaging - Spine

Discussion:
The figures demonstrate large osteophytes with concomitant ossification with bone bridging of both the anterior and posterior longitudinal ligament. This pattern is typical of diffuse idiopathic skeletal hyperostosis. Ankylosing spondylitis forms so-called “bamboo spine” with calcification of the interspinous ligaments along with the facet joints and adjacent structures and not large osteophytes or extensive thick calcifications of the anterior and posterior longitudinal ligaments as seen in this case. Rheumatoid arthritis is associated with enlarge pannus formation, not seen on this study. Klippel-Feil syndrome is associated with congenital fusions of two or more vertebral bodies not present on this study. Spondylosis is an aging-related degenerative process of the discs with adjacent osteophyte development not present on this study.

References:

Question 220: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders

Discussion:

Posterior Reversible Encephalopathy Syndrome (PRES) is characterized by headache, visual disturbances, altered mental. It frequently presents as patchy parietooccipital cortical/subcortical edema in patient with severe acute/subacute HTN. This the assumed result of failed autoregulation causes blood-brain barrier disruption resulting in vasogenic edema. The sagittal sinus shows flow signal and is not thrombosed on these views. The pattern of signal abnormalities is in an atypical for a cerebral infarction. Bilateral parietal occipital contusions are similarly unlikely to occur in isolation or with this appearance.

References:

Question 223: Neuroimaging - Tumors/Cysts

Discussion:

The neuroimaging studies demonstrate a heterogenously enhancing lesion in the left hemisphere with surrounding vasogenic edema and extension into the splenium of the corpus callosum. This appearance is most suggestive of a high-grade glioma. Cerebral abscesses may look similar, but typically do not have the lacy, fibrillary network of enhancement within the lesions such as the glioma in this case. Primary CNS vasculitis is best demonstrated by segmental stenosis of the intracranial arteries in a "string of beads" appearance. Neurosarcoïdosis typically shows strong enhancement of the leptomeninges and granulomas of the parenchyma. Acute intracerebral hemorrhage would be readily apparent on non-contrast CT. Arteriovenous malformation typically shows a network of flow voids that are seen on T2-weighted imaging. Tumefactive demyelination shows an open rim of enhancement, typically directed away from the ventricular surface.

References:

Question 228: Neuroimaging - Spine

Discussion:

The cerebellar tonsils fit criteria for Chiari malformation. There is a cystic cavity in the cervical spinal cord, related to a syrinx.

References:

**Question 233: Neuroimaging - Developmental/Neurogenetic Disorders**

**Discussion:**

The abnormal finding that must be recognized on these FLAIR images is the presence of tissue having signal intensity identical to that of cortical gray matter along the lateral subependymal aspect of each ventricle. This represents heterotopic gray matter. Subependymal nodules of tuberous sclerosis typically differ in signal from normal gray matter, and may have foci of heterogeneous signal related to calcifications. However, there can be some overlap in appearance in some cases.

**References:**


**Question 236: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders**

**Discussion:**

T1 hyperintensity is usually symmetric, in metabolic diseases, it could also be related to calcification or other mineralization. Bilateral T1 hyperintensity in basal ganglia, particularly globus pallidus which can be seen with underlying cirrhosis, in hyperalimentation from abnormal manganese metabolism, in patients undergoing prolonged parenteral feeding. Blood can be bright on the T1 weighted images, however, bilateral symmetric basal ganglia hemorrhage without adjacent edema or encephalomalacia would be much less likely.

**References:**


**Question 237: Neuroimaging - Spine**

**Discussion:**

There is cord contusion resulting from the combination of severe degenerative changes and trauma. There is C5-C6 fusion which serves as an anchor point for the hyper mobility with resultant cord contusion. Hyper flexion injury pattern is suggested with the vertebral body edema, and the posterior ligamentous injury. No burst fracture is present.

**References:**


**Question 239: Neuroimaging - Tumors/Cysts**

**Discussion:**

An intra-axial pontine lesion is noted that heterogeneously enhances. Moderate mass effect is present. This is not in a vascular distribution and not the ring-enhancing pattern seen with MS. The most likely diagnosis is a glioma.

**References:**


**Question 241: Neuroimaging - Dementia**
Discussion:

The patient is likely to have idiopathic calcification of the basal ganglia, also known as Fahr disease. CT scans demonstrate marked calcification of the basal ganglia and in the cerebellum. Calcification appears hypointensity on T2-weighted MRI. None of the possibilities listed can cause this degree of calcification. Leigh disease causes necrosis of the basal ganglia, particularly putamen, and results in low density of CT and hyperintensity on T2-weighted images. With amyloid angiopathy, hyperparathyroidism, and Wilson disease, there is a known disorder associated with lesser calcifications. In conditions where no such metabolic derangement can be found to explain the calcifications, the diagnosis is idiopathic calcification of the basal ganglia, or Fahr disease. Carbon dioxide toxicity would not be expected to cause progressive dementia.

References:


Question 242: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders

Discussion:

Wernicke's syndrome is associated with increased T2 and FLAIR signal and possibly diffusion signal to the mammillary bodies, medial thalamus, hypothalamus, peri aqueductal gray. Enhancement may be present in these regions and most common in the mammillary bodies. Deep venous thrombosis can produce increase signal in the thalami bilaterally general with thalamic edema not present and unlikely to produce the peri aqueductal gray signal changes seen here. Wernicke's syndrome is associate with thiamine or B1 deficiency and not vitamin B6. The artery of Perceron is an uncommon anatomic variant, in which a single dominant thalamoperforating artery supplies the bilateral medial thalami with variable contribution to the rostral midbrain. It's thrombosis is associated with bilateral paramedian thalamic infarcts and may produce midbrain infarctions. These images are atypical for such thalamic infarctions given the anterior signal extension encircling the third ventricle and pattern encircling the aqueduct. Hypoxic injury in adults has a relative predilection for the parietal occipital cortex, the medial temporal lobe, cerebellar folia, basal ganglia and less so the thalam.

References:


Question 243: Neuroimaging - Tumors/Cysts

Discussion:

Most common calcified primary brain tumor is an oligodendroglioma. Calcification occur in 70-90% of oligodendrogliomas.

References:


Question 244: Neuroimaging - Spine

Discussion:

Traversing S1 nerve roots are most likely compressed by the herniated disc. The MRI shows a focal right paracentral posterior herniation of the L5-S1 intervertebral disc. The herniated disc material causes mild narrowing of the spinal canal and severe narrowing of the lateral recess at L5-S1. The S1 nerve roots traverses at the L5-S1 level, and is therefore the most likely to be compressed by this disc herniation. The exiting nerve roots at the L5-S1 level (the L5 nerve roots) will already be lateral to the disc herniation and will not be compressed.

References:

Question 245: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

The small and medium sized vessels are ragged and demonstrate the segmented pattern of vasculitis. While vasospasm is possible, this typically affects larger vessels than those affected in this case. This appearance is known as "beads on a string".

References:


Question 246: Neuroimaging - Critical Care/Stroke

Discussion:

In global severe hypoxia, reduced diffusion in acute phase involving cerebral cortex, hippocampi, and basal ganglia, correlates with the increased metabolic demand of the grey matter. Superficial and deep water shed distribution infarcts are usually more associate with decreased blood flow rather than hypoxemia with intact or perhaps not as impaired cerebral blood perfusion.

References:


Question 253: Neuroimaging - Critical Care/Stroke

Discussion:

This may present as an acute neurological change and may necessitates immediate intervention. This patient underwent a large decompressive craniotomy then had a change in mental status with follow up CT showing marked concavity of the overlying skin flap causing mass effect and effacement of the superficial sulci.

References:


Question 255: Neuroimaging - Critical Care/Stroke

Discussion:

The middle cerebral artery and its branches are absent. The anterior cerebral artery and the posterior communicating artery and the posterior cerebral artery are present.

References:


Question 256: Neuroimaging - Critical Care/Stroke

Discussion:

The clinical history is inconsistent with the (much higher) severity of the imaged injuries. The extra-axial collections are of different signal intensities, indicating at least three different ages of subdural hematoma, strongly suggesting multiple episodes of traumatic injury. In this age group, these findings indicate nonaccidental trauma, unless several episodes of well documented accidental trauma can be substantiated (ie, multiple serious automobile accidents, which is not likely).
References:


Question 259: Neuroimaging - Critical Care/Stroke

Discussion:

The figure demonstrates a hyperdense superior sagittal sinus and lack on enhancement on the post contrast images consistent with a dural sinus thrombosis. There is right parietal hemorrhage demonstrated. Intracranial hemorrhages are frequently a sequela of these thrombotic events.

References:


Question 261: Neuroimaging - Infection

Discussion:

The scan demonstrates findings highly suggestive of acute or subacute herpes simplex virus (HSV) encephalitis involving the temporal lobe. The lesion crosses the vascular boundaries of the middle and posterior cerebral arteries, making an ischemic insult unlikely. The contrast-enhancing pattern would be unusual for glioblastoma or lymphoma. A contusion could occur in this location but would contain evidence of hemorrhage and MR evidence of parenchymal blood degradation products, which is not seen in this case. HSV encephalitis primarily involves limbic structures. MR typically shows temporal and inferior frontal lobe swelling, with low signal on T1-weighted images and high signal on T2-weighted images. Postcontrast studies in the acute and subacute setting often show cortical and leptomeningeal enhancement. The lesion may not be hemorrhagic in the early stages.

References:


Question 264: Neuroimaging - Spine

Discussion:

There is an enhancing nodule within the thecal sac. Most likely considerations would be nerve sheath tumor (schwannoma or neurofibroma) or a drop metastasis. Epidural abscess, chordomas and disc fragments are not in the intradural space. A lipoma would have a much higher signal on unenhanced T1.

References:


Question 265: Neuroimaging - Tumors/Cysts

Discussion:

Chordomas arise from remnants of the embryonic notochord which is a mesodermal derivative. Cranial chordomas are most common in the third and fourth decades of life and males are affected more often than females. Cartilaginous tumors occur between 20 and 60 years of age. They are extradural and over half arise in or adjacent to the body of the sphenoid bone. Radiologically the normal, high signal marrow cavity of the clivus is replaced by lower signal intensity tumor. Large areas of calcification may be seen as void-phenomenon. Meningiomas are isointense to the brain before contrast. This is an extra pontine lesion; the brainstem is not involved. The pituitary gland is normal. Parapharyngeal abscesses are usually smoothly contoured.
References:

Question 266: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:
Leptomeningeal enhancement, parenchymal enhancement, and, especially, pituitary stalk/hypothalamic involvement in a patient with cranial nerve symptoms is characteristic of neurosarcoidosis. About 5% to 10% of sarcoidosis patients develop CNS involvement. Lack of hemiatrophy and prominent (asymmetrical) choroid plexus enhancement make the diagnosis of Sturge-Weber syndrome less likely. No gyral enhancement is present. Low CSF pressure would not be limited to one hemisphere and would not have masslike and leptomeningeal enhancement. The enhancing lesion is intra-axial and therefore is not a meningioma.

References:

Question 267: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:
Arachnoid cysts are congenital space occupying lesions that may result in distortion and displacement of brain tissue. These also may result in deformity of the overlying skull with alteration in contour, and thinning of the calvarium, as in this case. This lesion is extra-axial, excluding the tumor diagnosis. Cortex is preserved, excluding the diagnosis of MCA infarction. The calvarium is not eroded. Arachnoid cysts (correct diagnosis) do not have a high likelihood of causing seizure, and typically are clinically silent, neurologically.

References:

Question 270: Neuroimaging - Infection

Discussion:
There are multiple cystic lesions containing a high intensity/density dot that corresponds to the scolex of Taenia solium. The patient has cysticercosis. As often happens in this disorder, intraventricular cysts block the cerebrospinal fluid pathways and hydrocephalus develops. Note that the cyst in the anterior third ventricle also contains a scolex. Hydrocephalus ex vacuo refers to enlargement of cerebrospinal fluid spaces due to atrophy, which is not present in this case. Colloid cysts usually do not contain a central area of focal enhancement. Schistosomiasis usually presents with multiple intraparenchymal edematous lesions with hemorrhage.

References:

Question 271: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders

Discussion:
Focal infarctions associated with MELAS was proven the correct diagnosis in this child who had lactic acidosis and multiple stroke-like episodes with localized brain tissue loss resulting in regions of gliosis and focal infarctions and localized compensatory ventricular enlargement. Leukodystrophies demonstrate diffuse lack of myelin formation, not seen here. Periventricular leukomalacia produces a characteristic pattern of gliosis aligning the lateral ventricles not present here. Moyamoya disease is very rare in childhood and not per se related associated with elevated lactate levels. Perinatal hypoxia is most frequently associated with parietal occipital encephalomalacia and would not be similarly associated with elevated lactate.

References:


Question 272: Neuroimaging - Multiple Sclerosis/Autoimmune Disorders (Non-MS)

Discussion:

On biopsy, this woman had a demyelinating disorder of the type known as tumefactive sclerosis, Marburg disease or Schilder disease. The lesions are large, with a cystic appearance on MRI and often have a ring of enhancement that is open: the “open-ring sign.” The ring of enhancement is open where the lesion touches the gray matter. Lesions circumscribed to the white matter may have a closed ring, as present in the lower left panel. Lesions caused by the other diagnoses listed tend to have more homogenous or closed-ring enhancement.

References:


Question 273: Neuroimaging - Critical Care/Stroke

Discussion:

The images demonstrate subarachnoid hemorrhage. This case is most likely due to Call-Fleming syndrome which is a reversible segmental vasoconstriction of cerebral arteries manifested by a “thunderclap” headache and which can be associated with subarachnoid hemorrhage. It is associated with antidepressant agents such as SSRIs, vasoactive sympathomimetic drugs, and antimigraine agents. Intracranial hypotension shows increase FLAIR signal along the pachymeninges not subarachnoid. The sagittal sinus is clearly patent. There is no history or imaging findings to suggest trauma. Posterior reversible encephalopathy syndrome results in cortical and juxtacortical white matter edema not present on this study.

References:


Question 275: Neuroimaging - Spine

Discussion:

The rounded lesion that enhances with gadolinium is located in the center of the conus medullaris, which is a location preferred by ependymomas, particularly the myxopapillary variant. Metastasis here is less frequent, as is the inflammatory lesion of schistosomiasis. Herniated discs and neurofibromata are extramedullary lesions.

References:
Question 276: Neuroimaging - Tumors/Cysts

Discussion:

The figures demonstrated a large homogeneously enhancing mass encompassing the left nasal passage, ethmoid sinuses, and extending through the cribriform plate into the ventral frontal region. An olfactory groove meningioma can have a similar intracranial appearance with extension into the upper nasopharynx; it would be unlikely to have this degree of paranasal sinus involvement disproportionate to its intracranial extension. A hemangiopericytoma is a soft tissue sarcoma that originates in the pericytes in the walls of capillaries. When inside the nervous system, although not strictly a meningioma tumor, it is a meningeal tumor with aggressive behavior but it is unlikely to present with this pattern which is most typical of an esthesioblastoma, which is a tumor arising from neural crest cells in the superior nasal cavity and can be associated with extension intracranially through the cribriform plate. A mucocele typically has little or no enhancement. A craniopharyngioma is most often a suprasellar cystic mass. Metastatic disease can be present within the nasal passage with intracranial extension although it would be unlikely to have this pattern of homogenous enhancement and absence of vasogenic edema. Mucocele, and extending through the cribriform plate into the ventral frontal region. This is homogeneously enhancing. While an olfactory groove meningioma can take this course, it would be unlikely to have this degree of paranasal sinus involvement disproportionate to intracranial extension. This pattern is most typical of an esthesioblastoma, which is a tumor arising from neural crest cells in the superior nasal cavity and can be associated with extension intracranially via the cribriform plate. A mucocele typically has little or no enhancement.

References:


Question 279: Neuroimaging - Critical Care/Stroke

Discussion:

The acute subdural hematoma is crescentic and extends past the coronal and lambdoid sutures, assuming the typical configuration of a collection expanding the subdural space. It is hyperdense relative to brain, consistent with acute subdural hematoma.

References:


Question 290: Neuroimaging - Infection

Discussion:

Treatment of HIV patients with combination antiretroviral therapy with transient paradoxical worsening of infectious processes is recognized as immune reconstitution inflammatory syndrome (IRIS). Immune reconstitution inflammatory syndrome is due to rapid, dysregulated restoration of the immune system in HIV patients with resultant worsening of active or subclinical infectious processes. This is often seen in the setting of infection by intracellular pathogens such as mycobacteria (tuberculous and non-tuberculous), cryptococcus, CMV, HSV, VZV and JCV. The inflammatory reaction is usually self-limited but occasionally may be severe enough to result in long-term sequelae and fatal outcomes. As pointed out in the question, the patient has PML, since his CSF was positive for JC virus, the causative agent of PML on polymerase chain reaction testing.

References:


Question 292: Neuroimaging - Metabolic, Movement, CSF Circulatory Disorders
Discussion:

The white arrows point to a band of decreased signal anterior to the cervical spine. This is a typical saturation pulse placed by the MRI technicians with the goal of reducing artifact from movements such as blood flow and respirations. Aliasing/wrap-around is artifact due to a smaller field of view than the average band image such that parts of the image are placed on the opposite side as compared to their actual position. Magnetic susceptibility artifact results in segments of decreased signal. There tends to be a margin of bright signal associated with such artifact, and this cylindrical pattern as seen with a saturation pulse would be unexpected. Truncation artifact is a series of lines parallel to segments with large changes of signal intensity such as adjacent to CSF and fat. Chemical shift artifact is due to the difference between resonance frequencies for fat and water and the frequency encoding direction, resulting in a misshapen or boundary pattern of artifact.

References:


Question 294: Neuroimaging - Spine

Discussion:

The CT demonstrates the pattern seen with a so-called “bamboo spine,” which is formed by diffuse syndesmophytic ankylosis as a result of interspinous ligament calcifications, diffuse ossification of the spinal ligaments, joints and enthesophyte formation. While the study does show osteopenia which can be seen with osteoporosis, this would not be the best answer. Rheumatoid arthritis is associated with enlarged pannus formation not seen on this study. Diffuse idiopathic skeletal hyperostosis is associated with bone proliferation along tendinous and ligamentous insertions of the spine resulting in calcification of the anterior and posterior longitudinal ligament and large anterior and posterior osteophytes not seen on this study. Spondylosis is a term for typical degenerative changes of aging of both the spinal discs and associated formation of adjacent osteophytes not seen on this study.

References:


Question 296: Neuroimaging - Dementia

Discussion:

The patient has cerebral amyloid angiopathy, most commonly seen in patients with Alzheimer disease. The other choices have no correlation with intraparenchymal microhemorrhages.

References:


Question 298: Neuroimaging - Epilepsy

Discussion:

Multiple intraparenchymal cystic lesions are seen, found to be metastatic cystic adenocarcinoma from a lung primary. Gliomas would be more infiltrative; meningiomas and arachnoid cysts would be extraaxial. Tuberous sclerosis does not lead to cystic rim-enhancing lesions.

References:

**Question 300: Neuroimaging - Spine**

**Discussion:**

The spinal lesion is multisegmental, elongated, and is in the lower cervical and thoracic levels. The pattern and extent of this lesion is atypical for multiple sclerosis in its size and extent and most characteristic of a form of transverse myelitis. The presence of anti-aquaporin antibodies (NMO antibodies) is a diagnostic marker of neuromyelitis optica (also known as Devic disease) which is a distinct form of demyelinating disease. The other choices would be highly unlikely to have these auto-antibodies.

**References:**


**Question 301: Neuroimaging - Tumors/Cysts**

**Discussion:**

The pituitary lesion shown on the left side of the gland demonstrates classic features of a pituitary microadenoma on the noncontrast MRI, with focal hypointensity less than one centimeter in size. On the image shown, there is less enhancement of the microadenoma as compared to normal pituitary tissue. An epidermoid is a possibility but much less likely. The sella does not appear empty in the images. Rathke cleft cyst would have less signal intensity on T1 and no enhancement. Craniopharyngiomas are suprasellar, whereas this one is within the sella.

**References:**


**Question 303: Neuroimaging - Tumors/Cysts**

**Discussion:**

Sagittal T1WI MR shows a well-circumscribed high signal lesion in the pineal region. The axial T2 image was fat saturated, with complete nulling of the signal from the "fatty" mass. This should not be confused with a flow void. Chemical shift artifact in frequency-encoding direction is extremely helpful in confirming the high fat content in this lesion.

**References:**


**Question 304: Neuroimaging - Developmental/Neurogenetic Disorders**

**Discussion:**
This MRI demonstrates a classic choroid fissure cyst, which has similar signal characteristics as CSF. The choroid fissure is a CSF space between the fimbria of the hippocampus and the diencephalon and is a common site for other neuroepithelial cysts and arachnoid cysts. Colloid cysts are usually seen in the anterior 3rd ventricle / Foramen of Monro region, and are most commonly hyperintense on T1WI and hypointense on T2WI (although this can be variable). Intracerebral hemorrhage is usually associated with surrounding vasogenic edema on T2WI and hypointense signal on T2-star Gradient Echo views. Mesial temporal sclerosis should demonstrate gliosis (hyperintensity on T2 FLAIR) and atrophy of the hippocampus and mesial temporal lobe. Cavum septum pellucidum is a developmental abnormality that results from the persistence of a normal fetal cavity and is seen as a CSF filled space contained between the two septal leaves.

References:

Question 305: Neuroimaging - Spine

Discussion:

There is symmetric abnormal T2 hyperintensity in the dorsal columns of the cervical spinal cord that can be seen associated with both B12 and copper deficiencies. The clinical correlate will be pseudoathetosis due to loss of proprioceptive sensation. This pattern of pathology spares the anterior motor columns, spinothalamic tracts, and autonomic nervous system and would not directly affect pain and temperature sensation, motor strength, or bladder or bowel continence or be associated with a Horner's syndrome.

References:

Question 306: Neuroimaging - Infection

Discussion:

The images demonstrate a smooth ring-enhancing mass within the left temporal lobe with markedly bright signal on the diffusion scan sequence. The diffusion findings are consistent with hypercellularity as seen with pus. Other massed lesions such as lymphoma or a PNET can have increased diffusion signal although it would be unlikely to be this bright. There is marked vasogenic edema more typical of an abscess as compared to a neoplasm. The figure also depicts enhancement within the left mastoid air cells, consistent with an acute mastoiditis, the most likely etiology of the adjacent abscess. IV drug use can result in septic emboli resulting in abscesses, although the figure clearly demonstrates mastoiditis and would be the best answer. A metastasis is unlikely to have this pattern of uniformly smooth ring enhancement and the degree of increased diffusion signal within the central aspect of the mass.

References:

Question 307: Neuroimaging - Tumors/Cysts

Discussion:

There is a calcified soft tissue mass in the pineal region, most consistent with a pineoblastoma. Mass effect on the aqueduct results in hydrocephalus. Because of the high incidence of subarachnoid seeding in these tumors, contrast-enhanced imaging of the entire spine is indicated to assess for drop metastases. Chordoma usually arises from clivus, which is intact. No pituitary calcification is present to suggest craniopharyngioma.

References:
Question 308: Neuroimaging - Epilepsy

Discussion:

Both the location of the lesion and the presence of precocious puberty favor a hamartoma in the tuber cinereum. In the sagittal image, the intact pituitary gland can be seen in the sella, above the suprasellar cistern.

References:


Question 309: Neuroimaging - Dementia

Discussion:

There is severe bilateral atrophy in the tip of the temporal lobes, most consistent with a neurodegenerative process such as frontotemporal dementia. Temporal gyri are atrophied, but the gyral crowns are preserved: in trauma they tend to be affected. The symptoms could be related to a prion disorder, but this is not given as an alternative. There is no hydrocephalus. The atrophy is atypical for mesial temporal sclerosis, in that it is very marked and does not predominantly affect the medial temporal region.

References:


Question 310: Neuroimaging - Developmental/Neurogenetic Disorders

Discussion:

This sagittal view of the brain demonstrates marked cerebellar atrophy that can be seen in spinocerebellar ataxias. No other malformations of the cerebellum are seen. The corpus callosum has normal appearance. This view of the brain also includes coverage of the upper cervical spine; it is important to review the appearance of these structures even when suspecting a lesion in the brain. In this case, no abnormalities of the cervical spine are seen.

References:


Pathology

Question 24: Pathology - Infectious Disease

Discussion:

Naegleria fowleri causes acute hemorrhagic meningoencephalitis, whereas entamoeba histolytica typically causes amoebic abscess. Toxoplasma likewise causes necrotizing abscesses within the brain. Plasmodium falciparum causes cerebral malaria with numerous petechial hemorrhages within the brain. Echinococcus produces hydatid cysts.

References:

Question 72: Pathology - Basic Reactions

Discussion:

Microglia are the resident macrophages of the brain. They share antigens with circulating monocytes, and both cell lines are probably derived largely from the same bone marrow precursors. The microglia are activated by injury to become large scavengers identical to macrophages. When activated, they may undergo mitosis. These cells play no role in maintaining the blood-brain barrier but are conspicuous at the circumventricular organs where the blood-brain barrier is absent.

References:


Question 96: Pathology - Demyelinating Disease

Discussion:

Megalencephaly is seen in a wide variety of conditions including some other inborn errors of metabolism, but among the choices the only correct answer would be Canavan disease.

References:


Question 125: Pathology - Demyelinating Disease

Discussion:

Progressive multifocal leukoencephalopathy (PML) is an opportunistic viral infection with tropism for oligodendrocytes. As of May 6, 2010, 49 confirmed cases of PML have been documented among patients receiving natalizumab worldwide. In addition to general characteristics of a demyelinating disease shared with active multiple sclerosis (such as abundant foamy macrophages, astrocytic atypia, and relative sparing of axons) intranuclear oligodendrocytic viral inclusions are only seen in PML. Perivascular inflammatory infiltrates are not typically a feature of PML.

References:


Question 143: Pathology - Critical Care/Stroke

Discussion:

Cerebral contusions are often associated with overlying subarachnoid hemorrhage; cerebral contusions and lacerations cannot by themselves produce epidural hemorrhage. Tearing of dural veins produces subdural hemorrhages, while middle meningeal artery lacerations are associations with epidural hemorrhages. Vertebral artery lacerations may produce large subarachnoid hemorrhages overlying the base of the brain.

References:


Question 153: Pathology - Neuromuscular Disease
Discussion:

In males suspected of having Duchenne's or Becker's muscular dystrophy, dystrophin analysis may reveal the presence of a mutation in affected individuals. Carrier analysis can also be performed. The precise location and size of the dystrophin mutation determines whether the more severe Duchenne's phenotype or the milder Becker's phenotype will be seen.

References:


**Question 155: Pathology - Toxic/Metabolic Disease**

Discussion:

Methanol is converted by the liver to formic acid and formaldehyde. These toxic compounds may result in bilateral necrosis of the lateral putamen and claustrum. This contrasts with the bilateral necrosis of the globus pallidus produced by carbon monoxide intoxication.

References:


**Question 166: Pathology - Cerebrovascular Disease**

Discussion:

The most common lesion attributed to sickle cell anemia is cerebral infarction. This is often due to occlusion of large arteries. The second most common complication is intracerebral hemorrhage. Subarachnoid hemorrhage is less common and attributed to rupture of aneurysms. Sagittal sinus thrombosis is rare.

References:


**Question 203: Pathology - Neuromuscular Disease**

Discussion:

Mitochondrial crystalloid inclusions may be seen in mitochondrial myopathies, or found incidentally, but with a history of other common features of mitochondrial diseases, the most likely other finding among the choices would be ragged red fibers.

References:


**Question 207: Pathology - Tumors**

Discussion:

The photo shows a poorly circumscribed variegated mass with foci of necrosis and hemorrhage most consistent with glioblastoma multiforme. The lack of central purulent material is against an abscess. The lesion is not confined within a single vascular territory, nor is it hemorrhagic, arguing against embolic infarct. The intra-axial location argues against most meningiomas. The lesion is a mass, ruling out Huntington disease.

References:
Question 218: Pathology - Tumors

Discussion:

The photograph shows an extra-axial mass anterior and lateral to the brainstem consistent with a schwannoma of the eighth cranial nerve. Schwannomas at this location, although arising from the vestibular division of the nerve and more correctly termed vestibular schwannomas, have been historically called "acoustic neuromas" or "acoustic neurinomas," and produce hearing impairment and tinnitus early in the clinical course. As they enlarge, adjacent cranial nerves may be encroached upon, producing facial sensory loss, facial weakness and dysphagia. Sporadic schwannomas of the 8th nerve typically present in adults between 4th and 6th decades. Bilateral lesions are associated with neurofibromatosis, type II, not type I.

References:


Question 221: Pathology - Tumors

Discussion:

Chordoid meningioma is a rare variant which resembles chordoma with cords of eosinophilic cells in a mucoid matrix. There may be vacuolated cells also. Usually more typical areas of meningioma are also present. This variant has a high rate of recurrence when it is the predominant pattern within the tumor and is a WHO grade II tumor. Chondrosarcomas and chordomas are S100 positive. Carcinomas are usually keratin positive. Pituitary adenomas may be keratin positive depending on the type of keratin and type of tumor, but are generally not EMA positive.

References:


Question 226: Pathology - Epilepsy

Discussion:

Patients with colloid cysts tend to present with headaches form obstructive hydrocephalus, but may have seizures and sudden death. The cysts are bright on precontrast T1 and do not enhance with contrast. The cyst contents are proteinaceous and do not suppress on T2 FLAIR. The epithelium is pseudostratified, ciliated and has goblet cells, but may be flattened and unrecognizable. The lining seen here is not consistent with the epithelium in a papilloma or craniopharyngioma. The rathke cleft cyst may have a similar lining but would be in the pituitary, not the third ventricle.

References:


Question 227: Pathology - Neuromuscular Disease

Discussion:

Tomaculous neuropathy (also known as pressure sensitive neuropathy) is characterized by focal hypermyelination (tomacula) with redundant myelin folds best seen on teased-fiber nerve preparations. Afflicted individuals have susceptibility to pressure palsies following relatively trivial compression.
References:


Question 230: Pathology - Developmental

Discussion:

The images showed numerous thin-walled cysts of varying sizes in the brain, characteristic of severe multicystic encephalomalacia due to intrauterine hypoxia/ischemia. The formation of cysts reflects cavitation which is the end stage of maturation of infarcts. The infarcts must be weeks old, and therefore, reflect intrauterine events. The threadlike strands within the cysts are gliovascular remnants and thus signify an event that occurred after astrocytes were capable of participating in the reaction, roughly after 27 weeks gestational age.

References:


Question 231: Pathology - Infectious Disease

Discussion:

Bacterial meningitis in adults is often characterized by leptomeningeal opacity caused by accumulations of bacteria and acute inflammatory cells, typically concentrated over the convexities of the brain. The causes vary with age and immunocompetence (or lack thereof). Common etiologies for immunocompetent adults include S. pneumoniae, N meningitidis, and less likely Staphylococcus.

References:


Question 232: Pathology - Neurodegenerative Disease

Discussion:

The images show disproportionate atrophy of the frontal and temporal lobes, a finding which would support the diagnosis of frontotemporal dementia.

References:


Question 234: Pathology - Toxic/Metabolic Disease

Discussion:

The biopsy shows a severe vacuolar myopathy due to extensive glycogen storage. These findings are consistent with Pompe disease (glycogen storage disease type II), which is caused by a genetic deficiency of acid-alpha-glucosidase (acid maltase). In its most severe form, the disease presents in infancy with cardiomegaly, hepatomegaly, progressive muscle weakness, macroGLOSSIA, and hypotonia (“floppy baby”). Of the choices provided, Pompe disease is the only disorder that presents findings of vacuolar myopathy on a muscle biopsy. Pathologic findings of Haltia-Santavuori disease, (infantile neuronal ceroid lipofuscinosis), and
Alexander disease primarily affect the brain. Krabbe disease is a leukodystrophy affecting CNS white matter. Kufs disease is an adult form of neuronal ceroid lipofuscinosis.

References:


Question 238: Pathology - Critical Care/Stroke

Discussion:

The hematoxylin and eosin stained sections demonstrate numerous enlarged, round axonal swellings which stain positively with the silver stain. This is the histologic picture of axonal spheroids, the finding in diffuse axonal injury. Also known as "shear injury," patients with this condition are rendered immediately unconscious with trauma, and subsequently have a course of chronic "closed head injury." Axonal spheroids may be seen at the edge of a cerebral infarction but not in this clinical context and diffuse pattern. Multiple sclerosis may also eventually result in axonal injury with spheroids but with more extensive demyelination than is evident in the figure showing intact myelin.

References:


Question 240: Pathology - Demyelinating Disease

Discussion:

This is an example of acute disseminated encephalomyelitis (also known as perivenous encephalomyelitis), a disorder that is most common in first and second decades and one from which the usual outcome is complete clinical recovery. The photograph illustrates the classic perivenous sleeves of demyelination and mononuclear cells and is illustrated on a myelin stain (Luxol fast blue-periodic acid Schiff for myelin). Death is fortunately uncommon. The most common antecedent illness today is a nonspecific respiratory infection, although formerly many fatal cases followed measles or smallpox vaccination. The severe variant of this disease is acute hemorrhagic leukoencephalitis (Hurst disease).

References:


Question 247: Pathology - Neurodegenerative Disease

Discussion:

The histologic hallmarks of Alzheimer disease are neuritic plaques, neurofibrillary tangles, Hirano bodies, neuronal granulovacuolar degeneration, and the deposition of amyloid in the walls of blood vessels. The pyramidal neuron illustrated showed granulovacuolar degeneration, a neurofibrillary tangle, and a Hirano body.

References:


Question 249: Pathology - Demyelinating Disease

Discussion:

The brain shows the characteristic features of a central, triangular lesion of central pontine myelinolysis, due to electrolyte disturbances in most well documented cases. Rapid and sustained elevations in serum sodium will experimentally produce similar lesions in rats and dogs.
References:


Question 254: Pathology - Developmental

Discussion:

Open neural tube defects (NTDs) including anencephaly are associated with elevated alpha fetoprotein (AFP) in maternal serum because the central nervous system makes this protein and the lack of closure of the neural tube allows the protein into the amniotic fluid. It subsequently enters the maternal circulation. NTDs are associated with trisomy 18 or 13. Anencephaly may be associated with spinal NTDs of various sizes.

References:


Question 258: Pathology - Hypothalamus/Pituitary

Discussion:

Tumors centered in the sella and expanding into the suprasellar space frequently also invade down into the sinuses. "Nasal biopsies" may be misdiagnosed if the pathologist isn't aware of the actual center (origin) of the lesion. The MRI and prolactin level make an adenoma more probable. Both olfactory neuroblastomas and pituitary adenomas are neuroendocrine (synaptophysin and chromogranin positive). CAM5.2 is more likely to be positive of all the possible keratins in adenomas. Rarely, though, even neuroblastomas may be keratin positive.

References:


Question 280: Pathology - Neurodegenerative Disease

Discussion:

In the figure, myelin-stained cross sections of the spinal cord show pallor and Wallerian degeneration of the corticospinal tracts with sparing of the posterior columns and spinocerebellar tracts. These findings are most compatible with amyotrophic lateral sclerosis.

References:


Question 281: Pathology - Demyelinating Disease

Discussion:

Multiple periventricular areas of demyelination are characteristically seen in multiple sclerosis. These plaques have a brownish gray hue because of the loss of myelin that normally confers a glistening white appearance to the white matter.

References:
Question 285: Pathology - Tumors

Discussion:

The photomicrographs show classic perivascular pseudorosettes characteristic of ependymoma.

References:


Question 286: Pathology - Neuromuscular Disease

Discussion:

Formation of onion bulbs results from repeated episodes of demyelination and remyelination. Onion bulbs are especially conspicuous in hypertrophic Charcot-Marie-Tooth disease, Dejerine-Sottas disease and Refsum disease. In addition, approximately one half of the cases of chronic inflammatory demyelinating polyradiculoneuropathy show variable numbers of onion bulbs.

References:


Question 287: Pathology - Cerebrovascular Disease

Discussion:

The photograph shows a massive basal ganglionic hemorrhage with rupture into the ventricular system. The location of this hematoma is common for hypertensive intracerebral hemorrhage. Hypertension is a major risk factor for this type of hemorrhage, and hypertension is associated with pathologic and electrocardiographic evidence of left ventricular hypertrophy. Prostate cancer only rarely metastasizes to brain parenchyma and is usually not hemorrhagic. Visceral cysts are not relevant to intracerebral hemorrhage, but are frequently seen in von Hippel-Lindau disease with associated CNS hemangioblastoma.

References:


Question 291: Pathology - Neurodegenerative Disease

Discussion:

The illustration shows a brain with severe atrophy of the pons. This is grossly consistent with olivopontocerebellar atrophy which is a type of multiple system atrophy.

References:

Question 295: Pathology - Infectious Disease

Discussion:

Neurosarcoid is a diagnosis of exclusion. In the orbit, it most often affects the lacrimal gland, but may cause optic neuritis. The most common cranial nerve palsy involves facial nerve. The granulomas of sarcoid tend to have less well-developed rims of lymphocytes than do those of infections.

References:


Question 297: Pathology - Toxic/Metabolic Disease

Discussion:

A coronal section of the brain at autopsy shows mammillary bodies that are congested and discolored, which is a characteristic finding in thiamine deficiency manifesting as Wernicke encephalopathy. Few other disorders affect the mammillary bodies. Vitamin B12 deficiency can cause subacute combined degeneration, a spinal cord lesion. Methanol poisoning classically causes hemorrhagic putaminal lesions.

References:


Question 299: Pathology - Tumors

Discussion:

The biopsy of a right parietal lobe mass showed a tumor, composed of cells with monotonous round nuclei, surrounded by prominent perinuclear halos (“fried egg” appearance). These features are characteristic of oligodendroglioma. Of the other choices listed, ependymomas characteristically show perivascular pseudorosettes. The clear cell variant may mimic oligodendroglioma. Fibrillary astrocytomas and gemistocytic astrocytomas show eosinophilic cytoplasm rather than the clear perinuclear halos of oligodendrogliomas, with multiple elongated cytoplasmic processes in fibrillary astrocytomas and large rounded globular cytoplasm in gemistocytic astrocytomas. Neurocytomas may resemble oligodendrogliomas microscopically but are usually intraventricular.

References:


Question 302: Pathology - Epilepsy

Discussion:

The dysembryoplastic neuroepithelial tumor (DNT) is composed of neurons and oligodendrogial-like cells clustered within intracortical nodules. A prominent myxoid stroma is typically present within the tumor nodules. This indolent tumor is frequently associated with cortical migration abnormalities. Surgery is curative.

References:

Question 316: Pathology - Neuromuscular Disease

Discussion:

GNE myopathy, also known as hereditary inclusion body myopathy or distal myopathy with rimmed vacuoles, is one of several neuromuscular disorders in which rimmed vacuoles may be identified on muscle biopsy. Inclusion body myositis, which is the most common, typically presents in much older individuals. Oculopharyngeal muscular dystrophy may also have rimmed vacuoles but has a different pattern of muscle weakness.

References:


Question 328: Pathology - Epilepsy

Discussion:

Balloon cells are seen in focal cortical dysplasia type IIB (Palmini, et al system). Hippocampal sclerosis is seen in medial temporal lobe. Hirano bodies and granulovacuolar degeneration are found in Alzheimer disease and aging.

References:


Question 332: Pathology - Neurodegenerative Disease

Discussion:

Tau protein deposition is characteristic of chronic traumatic encephalopathy. Senile plaques, amyloid angiopathy, granulovacuolar degeneration, and Hirano bodies are typical of Alzheimer disease. Spongiform degeneration is characteristic of Creutzfeldt-Jakob disease.

References:


Question 365: Pathology - Cerebrovascular Disease

Discussion:

Kawasaki's disease and polyarteritis nodosa (PAN) both have necrotizing vasculitis as their sole pathologic vascular lesions, while Takayasu's arteritis and temporal arteritis are both forms of granulomatous arteritis. Primary angiitis of the CNS may present as both granulomatous and non-granulomatous vasculitis, the latter taking the form of a PAN-like necrotizing vasculitis.

References:


Question 382: Pathology - Cerebrovascular Disease

Discussion:

Familial forms of cavernous malformations follow an autosomal dominant pattern of inheritance. The proportion of familial cases is approximately 50% among patients of Mexican descent and seems to be less in other populations.
References:


Question 400: Pathology - Cerebrovascular Disease

Discussion:

Saccular ("berry") aneurysms of the circle of Willis are present in 10-30% of patients with adult polycystic kidney disease, in whom they are responsible for significant morbidity and an important cause of death. Saccular aneurysms are also associated with fibromuscular dysplasia, coarctation of the aorta and moyamoya disease.

References:


Pharmacology/Chemistry

Question 4: Pharmacology/Chemistry - Epilepsy

Discussion:

This patient fulfills the clinical criteria for propofol infusion syndrome. The clinical features of propofol infusion syndrome are acute refractory bradycardia leading to asystole, in the presence of one or more of the following: metabolic acidosis (base deficit > 10 mmol/l, rhabdomyolysis, hyperlipidemia, and enlarged or fatty liver. There is an association between propofol infusion syndrome and propofol infusions at doses higher than 4 m/kg/hour for greater than 48 hours. An early sign of cardiac instability associated with propofol infusion syndrome is the development of right bundle branch block with convex-curved ("coved type") ST elevation in the right praecordial leads (V1 to V3) of the electrocardiogram.

References:


Question 11: Pharmacology/Chemistry - Movement Disorders

Discussion:

This patient has clinical manifestations of parkinsonism with bradykinesia, postural instability, and rigidity. However, this patient also has additional manifestations included impaired voluntary eye movements, with an intact oculocephalic reflex. This presentation is most consistent with progressive supranuclear palsy, which is pathologically characterized by tau inclusions in the substantia nigra. Alpha-synuclein inclusions are seen in the substantia nigra in Parkinson disease and in cortical neurons in Dementia with Lewy Bodies.

References:


Question 17: Pharmacology/Chemistry - Neurogenetics

Discussion:

Ataxia telangiectasia (AT) is characterized by progressive cerebellar ataxia, oculocutaneous telangiectasia, progressive cerebellar dysfunction, and recurrent sinopulmonary infections secondary to progressive immunological and neurologic dysfunction. AT patients are significantly predisposed to cancer, particularly lymphomas and leukemia. Other manifestations of the disease include sensitivity to ionizing radiation, premature aging, and hypogonadism. AT has been a major interest of scientists since the
1960's because it may yield an insight into numerous other major health problems, such as cancer, neurologic disease, immunodeficiency, and aging.

AT is characterized by: early-onset progressive cerebellar ataxia, oculocutaneous telangiectasia (dilated blood vessels in the eyes and skin), immunodeficiency mostly thorough lowering of IgA, IgG and IgE levels, chromosomal instability, hypersensitivity to ionizing radiation, increased incidence of malignancies primarily lymphoid, and raised alpha-fetoprotein levels.

References:


Question 19: Pharmacology/Chemistry - Epilepsy

Discussion:

Grapefruit juice leads to inhibition of CYP3A4, predisposing to increased plasma levels and toxicity of carbamazepine. There is no interaction between dietary proteins or carbohydrates nor is there an effect of carbamazepine on zinc metabolism. Carbamazepine does not increase the risk of nephrolithiasis.

References:


Question 28: Pharmacology/Chemistry - Movement Disorders

Discussion:

Amiodarone has been associated with drug-induced postural tremor. Beta-blockers such as propanolol and sotalol may improve postural tremor. Diltiazem and digoxin have not been associated with tremor.

References:


Question 30: Pharmacology/Chemistry - Epilepsy

Discussion:

Lamotrigine is metabolized in the liver predominantly by glucuronic acid conjugation; the major metabolite is an inactive 2-N-glucuronide conjugate. Carbamazepine, phenytoin, and phenobarbital induce glucuronic acid conjugation of lamotrigine resulting in a 40% reduction of the lamotrigine level. Valproic acid inhibits glucuronic acid conjugation of lamotrigine doubling the lamotrigine level. Coadministration of lamotrigine with eslicarbazepine acetate has been shown to modestly decrease the plasma concentrations of lamotrigine by an average of 15%. The mechanism of interaction has not been established. This change is not considered clinically significant, and no dosage adjustments are required.

References:

**Question 37: Pharmacology/Chemistry - Headache**

**Discussion:**

Medication overuse headache (MOH) is a secondary form of chronic daily headache, in which excessive use of acute medications (e.g., butalbital, opioids, NSAIDs, triptans) causes transformation to headache occurring 15 or more days per month for 4 or more hours per day if left untreated. The risk of developing medication overuse headache is highest with butalbital (5 days of use per month), followed by opioids (8 days of use per month), triptans (10 days of use per month), and NSAIDs (10-15 days per month).

**References:**


**Question 54: Pharmacology/Chemistry - Neuromuscular Disorders**

**Discussion:**

This patient has the classic presentation of acute polyneuropathy following oxaliplatin treatment. Patients with acute oxalplatin polyneuropathy have increased sensitivity to touching cold items and swallowing cold items (71%), throat discomfort (63%), and muscle cramps (42%). These symptoms peak 3 days after treatment. Electromyography in patients with acute oxaliplatin neuropathy demonstrates spontaneous high frequency bursts of muscle unit action potentials and repetitive compound muscle action potentials. Paclitaxol causes a chronic large and small-fiber sensory predominant polyneuropathy. Thalidomide causes a distal sensory predominant polyneuropathy with tingling when tapping the fingertips and autonomic neuropathy manifesting as constipation in more than 80% of patients. Vincristine causes a distal sensory motor axonal polyneuropathy with more than a third of patients having autonomic neuropathy, manifesting as orthostatic hypotension, constipation, bladder dysfunction or impotence. Polynynepathy. Following treatment with brentuximab vedotin, 50% of patients treated with brentuximab had a sensory predominant polyneuropathy and 8% had a motor predominant polyneuropathy.

**References:**

Pachman DR, Qin R, Seisler DK, et al.. Clinical Course of Oxaliplatin-Induced Neuropathy: Results From the Randomized Phase III Trial N08CB (Alliance). J Clin Oncol. 2015;33(30):3416-3422


**Question 57: Pharmacology/Chemistry - Movement Disorders**

**Discussion:**

Paroxysmal kinesigenic dyskinesia may be sporadic or inherited in an autosomal dominant fashion. It responds well to low-dose anticonvulsants, such as carbamazepine.

**References:**


**Question 65: Pharmacology/Chemistry - Neuromuscular Disorders**

**Discussion:**

...
Sporadic inclusion body myositis (IBM) is the most common acquired myopathy in patients over 50 years of age. IBM classically presents with asymmetric finger flexor and knee extensor weakness. IBM is resistant to immunosuppressive therapy. In 2011, Salajegheh et al. reported an autoantibody against a 43-kDa muscle antigen highly specific for IBM. The antigen was later identified as cytoplasmic 5'-nucleotidase 1A (cN1A; NT5C1A). Two independent case control studies found 60-70% sensitivity and 91-92% specificity for this antibody in IBM.

References:


**Question 76: Pharmacology/Chemistry - Neurogenetics**

**Discussion:**

This patient has Fabry's disease, an X linked recessively inherited disorder associated with deficiency the enzyme alpha galactosidase A (ceramide trihexosidase). The enzyme deficiency results in accumulation of trihexoside and other glycosphingolipids in various tissues including skin, nervous system, vascular endothelium, kidney, and cardiovascular system. Deposition in autonomic and dorsal root ganglia, unmyelinated and myelinated axons are associated with dysautonomia and painful distal neuropathy. Diagnosis is by assaying the enzyme in leukocytes or skin fibroblasts. Enzyme replacement therapy leads to a modest improvement in clinical manifestations of neuropathy.

References:


**Question 79: Pharmacology/Chemistry - Epilepsy**

**Discussion:**

In 1965, diazepam was first used to treat status epilepticus in humans. Clonazepam was introduced in the 1970’s primarily as an anti-epileptic drug and clobazam, a 1,5 benzodiazepine, was later developed as an anti-epileptic drug with less sedative effect. Induction of tolerance limits the use of benzodiazepines as anti-epileptic drugs. The 1,5 benzodiazepines like clobazam produce less tolerance than the 1,4 benzodiazepines like diazepam and lorazepam.

References:


**Question 80: Pharmacology/Chemistry - Toxic/Metabolic Disease**

**Discussion:**

Methanol intoxication results in severe metabolic acidosis due to oxidation of methanol to formic acid. Dilated, unreactive pupils and reduced vision are typical, due to destruction of retinal ganglion cells. Bicarbonate is the keystone of treatment of methanol poisoning. Concomitant administration of Fomepizole, a competitive substrate of alcohol dehydrogenase, may have some benefit. A loading dose (15 mg/kg) followed by four maintenance dose (10 mg/kg) every 12 hours should be administered as soon as the diagnosis of methanol poisoning is made.

References:


Question 84: Pharmacology/Chemistry - Epilepsy

Discussion:

Valproic acid is associated with idiosyncratic hyperammonemia. Valproate-associated hyperammonemic encephalopathy (VHE) is characterized by a decreasing level of consciousness, focal neurologic deficits, cognitive slowing, asterixis, drowsiness, and lethargy. The pathophysiology of VHE is poorly understood but a disturbance of urea cycle metabolism has been postulated; this is independent of hepatic function.

References:


Question 92: Pharmacology/Chemistry - Toxic/Metabolic Disease

Discussion:

Organophosphorous pesticides phosphorylate the hydroxyl group of the serine residue of acetylcholinesterase, which irreversibly inhibits the function of this enzyme. As a result, there is an accumulation of acetylcholine in the synaptic cleft, which continuously binds to the acetylcholine receptor on the muscle cell membrane raising the resting muscle cell membrane potential. Voltage gated sodium channels on the muscle remain in an inactive state at this raised resting muscle cell membrane potential, preventing muscle cell depolarization and contraction.

References:


Question 93: Pharmacology/Chemistry - Movement Disorders

Discussion:

Livedo reticularis is a skin condition associated with rheumatologic and hematologic conditions including: Systemic lupus erythematosus, antiphospholipid syndrome and rheumatoid arthritis. Amantadine is also known to cause this potentially reversible dermatologic manifestation. Treatment is considered unnecessary unless symptomatic. However, discontinuation of or decrease in Amantadine may be attempted.

References:


Question 98: Pharmacology/Chemistry - Other Pain Syndromes

Discussion:

This woman has right Horner syndrome, likely due to right internal carotid artery (ICA) dissection as a result of chiropractic maneuvers. ICA dissection causes of oculosympathtic fibers that travel along the extracranial ICA. With Horner syndrome of any cause (central, pre-ganglionic, post-ganglionic), there is denervation hypersensitivity at post-ganglionic sympathetic receptors of the pupillary dilator muscle. Apraclonidine is a weak alpha adrenergic agonist that binds to post-ganglionic receptors to cause pupillary dilatation in all types of Horner syndrome. Hydroxyamphetamine will cause pupillary dilatation in Horner syndrome of central (e.g. brainstem) and pre-ganglionic origin (e.g. Pancoast tumor), when the third-order post-ganglionic neuron is intact (and can thus release norepinephrine in response to hydroxyamphetamine). In carotid dissection, the post-ganglionic neuron is impaired and there is no response to topical amphetamine. Topical cocaine produces no pupillary response in Horner syndrome,
as there is a synaptic deficit of norepinephrine. Pilocarpine in a cholinergic agent, producing pupillary constriction and timolol is a beta-blocker, preventing pupillary dilatation.

References:


Question 100: Pharmacology/Chemistry - Headache

Discussion:

Botulinum toxin injected subcutaneously into the bilateral forehead, temples, occiput, neck, and shoulders was approved by the Food and Drug Administration for the prophylaxis of chronic migraine in October of 2010 based on a demonstrated reduction in headache-free days by about one additional day when compared to placebo (-11.7 versus -10.8, respectively).

References:

Aurora SK. Winner P. Freeman MC. Spierings EL. Heiring JO. DeGryse RE. VanDenburgh AM. Nolan ME. Turkel CC.. Onabotulinumtoxin A for treatment of chronic migraine: pooled analyses of the 56-week PREEMPT clinical program.. Headache 2011; 51(9):1358-73


Question 107: Pharmacology/Chemistry - Cerebrovascular Disease

Discussion:

For patients on anticoagulant therapy undergoing a surgical intervention, the risk of bleeding from anticoagulant therapy should be weighed against the risk of thromboembolism. If INR is not supratherapeutic, the risk of bleeding from simple dental extraction is considered low. It is recommended that anticoagulant therapy not be interrupted. Local measures such as pressure and/or local hemostatic agent should be used to control bleeding.

References:


Question 118: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Daclizumab is a humanized antibody that binds to CD25, the alpha subunit of the high-affinity interleukin-2 (IL2) receptor expressed on T-lymphocytes. Through this interaction, daclizumab reduces IL-2 signaling, which is thought to be important in the pathogenesis of MS and other autoimmune disorders. In a randomized, double-blind study, daclizumab demonstrated significant reduction in relapse rate (45%) and MRI activity when compared with intramuscular interferon beta-1a. Patients treated with daclizumab had higher incidence of infection, cutaneous disorders, and transaminitis. Autoimmune dermatitis and hepatitis have also been associated with daclizumab, and monthly monitoring of liver enzymes has been mandated by the FDA for patients treated with daclizumab. ITP, Goodpasture disease, and autoimmune thyroid disease have been associated with alemtuzumab, a CD-52 monoclonal antibody, in patients with RRMS.

References:


**Question 122: Pharmacology/Chemistry - Other Pain Syndromes**

**Discussion:**

Arsenic is a heavy metal with a name derived from the Greek word "arsenikon," meaning potent. Once arsenic is in the body, it binds to hemoglobin, plasma proteins, and leukocytes and is redistributed to the liver, kidney, lung, spleen, and intestines. Over a period of weeks, deposits may be found in skin, hair, nails (Mee's lines), bone, muscle, and even nervous tissue. Arsenic produces cellular damage through a variety of mechanisms. In some forms, arsenic is caustic, exerting a direct toxic effect on blood vessels and large organs. Long-term exposure results in nerve damage and may lead to lung, skin, or liver cancer. Once inhaled, arsine gas combines with hemoglobin in red blood cells, causing severe hemolysis and anemia. Patients develop hemoglobinuria and hematuria within several hours of exposure. Patients with acute exposure present with gastrointestinal distress characterized by nausea, vomiting, abdominal pain, and profuse watery or bloody diarrhea. Patients often are hypotensive and tachycardic and may complain of a metallic taste in their mouth and have a garlic odor on their breath. Patients frequently exhibit signs of delirium upon examination. Patients with chronic arsenic exposure often present with painful paresthesias. Neuropathy results in diminished sensitivity to pinprick, light touch, temperature, and vibration and in motor deficits in a stocking-glove distribution. Muscle wasting and foot drop sometimes are noted. Other examination findings include cyanosis of distal extremities, pallor from anemia, hyperpigmentation of skin, and Mees lines. Patients may develop cardiovascular effects, diabetes mellitus, or cancer, as well. Current approved treatment lies in the administration of chelating agents that form an insoluble complex with the metal and remove it. They have been used clinically as antidotes for treating acute and chronic poisoning. The most widely used chelating agents are calcium disodium ethylenediamine tetra acetic acid (CaNa2EDTA), D-penicillamine, and British anti-lewisite (BAL).

Lead has direct effects on porphyrin metabolism, by inhibiting gamma-aminolevulinic acid dehydrase. Lead intoxication produces a motor neuropathy that affects predominantly, but not exclusively, the radial nerve. Associated features include abdominal pain, bluish discoloration of the gums just below the teeth, microcytic hypochromic anemia with basophilic stippling of the red cells, and increased coproporphyrin levels. Arsenic reacts with sulfhydryl groups of proteins and interferes with several steps of oxidative metabolism in the neuron, producing dying back type axonal degeneration, particularly in myelinated fibers. Thallium ions act interchangeably with potassium in respect to their transport by the Na/K ATPase system. Alopecia, and cranial nerve and autonomic involvement help to distinguish thallium from arsenic intoxication. Organophosphates (OP) inhibit acetylcholinesterase and OP intoxication produces an early syndrome (type I) of excessive muscarinic activation, followed by an intermediate syndrome (type II) of excessive nicotinic activation and muscle weakness. In addition, OP induces a delayed axonal, predominantly motor neuropathy associated with manifestations of CNS involvement (ataxia, spasticity); this is attributed to phosphorylation and inhibition of a neuropathy target esterase (NTE) and impairment of axonal transport. Acrylamide impairs axonal transport causing accumulation of neurofilaments and paranoidal swelling mostly in large myelinated axons. This produces a dying-back axonopathy, affecting both the peripheral nerves and the central tracts. (eg, gracile tract and dorsal spinocerebellar)

**References:**


**Question 129: Pharmacology/Chemistry - Neuromuscular Disorders**

**Discussion:**

Lead has direct effects on porphyrin metabolism by inhibiting gamma-aminolevulinic acid dehydrase. Lead intoxication produces a motor neuropathy that affects predominantly, but not exclusively, the radial nerve. Associated features include abdominal pain, bluish decoloration of the gums just below the teeth, microcytic hypochromic anemia with basophilic stippling of the red cells, and increase coproporphyrin levels.

**References:**

**Question 132: Pharmacology/Chemistry - Cerebrovascular Disease**

**Discussion:**

Idarucizumab has been shown to reverse the anticoagulant effect of Dabigatran. Andexanet is a potential drug developed to neutralize the anticoagulant effect of Apixaban, Rivaroxaban and Edoxaban but is pending FDA approval.

**References:**


**Question 140: Pharmacology/Chemistry - Neuromuscular Disorders**

**Discussion:**

This patient's clinical presentation of fluctuating extra-ocular, bulbar, and proximal muscle weakness that worsens as the day progresses is most consistent with generalized myasthenia gravis. Approximately 80% of all generalized myasthenia gravis patients have autoantibodies against the nicotinic acetylcholine receptor in their serum. Autoantibodies against the tyrosine kinase muscle-specific kinase (MuSK) are responsible for 5-10% of all generalized myasthenia gravis cases. Low-density lipoprotein receptor-related protein 4 (LRP4) has been identified as the agrin receptor. LRP4 interacts with agrin, and the binding of agrin activates MuSK, which leads to the formation of most if not all postsynaptic specializations, including aggregates containing acetylcholine receptors (AChRs) in the junctional plasma membrane. Antibodies to LRP4 have been reported to occur in up to 50% of patients with myasthenia who were negative for acetylcholine receptor and tyrosine kinase muscle-specific kinase antibodies. Antibodies to aquaporin-4 are associated with neuromyelitis optica. Antibodies to 3-hydroxy-3-methylglutaryl-coenzyme A reductase (HMG CO-a reductase) are associated with statin-induced necrotizing myopathies. Antibodies to leucine-rich, glioma inactivated 1 protein (LGI-1) and contactin-associated protein-like 2 (Caspr2) are autoantigens of encephalitis and peripheral nerve hyperexcitability (PNH) previously attributed to voltage-gated potassium channels antibodies.

**References:**


**Question 145: Pharmacology/Chemistry - Demyelinating Disorders**

**Discussion:**

Depression, suicidal ideation, and new or worsening other psychiatric disorders have been observed to be increased in patients using interferon compounds. Adverse reactions most commonly reported in patients are flu-like and other symptoms occurring within hours to days following an injection. Symptoms can include myalgia, fever, fatigue, headaches, chills, nausea, and vomiting. The most frequently reported adverse reactions resulting in discontinuation or the need for concomitant medication to treat an adverse reaction symptom) were flu-like symptoms and depression.

**References:**


**Question 154: Pharmacology/Chemistry - Dementia**

**Discussion:**
Tau protein is a heat-resistant phospho-protein that promotes microtubule polymerization and stabilization. Tau accumulates in neurons and in glia in a wide range of neurodegenerative disorders and in the aging brain. Disorders in which tau pathology is considered the major contributing factor to neurodegeneration are referred to as “primary tauopathies.” Tau protein in the brain is heterogeneous due to alternative splice forms, as well as post-translational modifications, including phosphorylation. Exon 10 of the MAPT gene is alternatively spliced to generate tau species with either three or four conserved ~32 amino acid repeats in the microtubule binding domain of tau protein referred to as 3R and 4R tau. In Alzheimer disease, neurofibrillary pathology is composed of an equimolar ratio of 3R and 4R tau. In Pick disease, Pick bodies are primarily composed of 3R tau. In progressive supranuclear palsy and corticobasal degeneration, the cytoplasmic inclusions are primarily composed of 4R tau.

References:


Discussion:

Tardive dyskinesia (TD) primarily involves the tongue, lips, and jaw. A combination of tongue twisting and protrusion, lip smacking and puckering, and chewing movements in a repetitive and stereotypic fashion is often observed. TD results from chronic exposure to dopamine receptor blocking agents - drugs primarily used to treat psychosis. TD has not been reported with agents that deplete dopamine (such as reserpine) and is only rarely reported with "atypical" antipsychotic drugs (such as clozapine). Some drugs for nausea (such as metoclopramide or prochlorperazine) and depression (such as amoxapine) are actually dopamine receptor blocking agents and therefore can cause TD.

The weight of evidence favors the notion that chronic blockade of dopamine receptors leads to increased receptor sensitivity. After 2 weeks of therapy with dopamine receptor blocking agents, the affinity and number of dopamine D2 receptors increased in rodent models of TD. Yet, it remains unclear how this leads to the development of TD, or why only a minority of patients with similar drug exposure develop TD.

References:


Question 164: Pharmacology/Chemistry - Aging, Degenerative Diseases

Discussion:

Some patients with autonomic failure exhibit a degree of hypoproliferative anemia. Administration of recombinant human erythropoietin-alpha, 25-75 U/kg subcutaneously two to three times a week for 6 weeks, rapidly corrects the anemia and improves orthostatic tolerance.

References:


Question 180: Pharmacology/Chemistry - Epilepsy

Discussion:

Valproic acid binds to plasma proteins, displacing phenytoin from protein binding sites and thus causing higher free phenytoin levels. The other agents do not interact significantly with phenytoin at all.

References:

French J.. Treatment with antiepileptic drugs, new and old.. Continuum, volume 13, 2007, 71-90
**Question 311: Pharmacology/Chemistry - Neuromuscular Disorders**

**Discussion:**

Dystrophin is a membrane-bound protein distributed along the intracellular surface of the sarcolema and is a member of the superfamily of cytoskeletal proteins; dystrophin interacts with actin and may contribute to structural and functional stability of the plasma membrane.

**References:**


**Question 315: Pharmacology/Chemistry - Dementia**

**Discussion:**

Memantine (Namenda) is a moderate-affinity, non-competitive, voltage-dependent, N-methyl-D-aspartate (NMDA) -receptor antagonist with fast on/off kinetics that inhibits excessive calcium influx induced by chronic overstimulation of the NMDA receptor.

**References:**


**Question 319: Pharmacology/Chemistry - Cerebrovascular Disease**

**Discussion:**

Although infrequent, orolingual angioedema has been more recognized due to increased use of IV thrombolysis. Emergent intubation may be needed. IV tPA increases plasma plasmin activity as well as bradykinin which potentiates vascular permeability and vasodilation. Angiotensin converting enzyme (ACE) normally inactivates bradykinin and ACE inhibitor will therefore increase the risk of angioedema among patients receiving IV tPA. Interesting findings are the asymmetry of angioedema, preferentially for the hemiparetic side. Patients with middle cerebral artery infarction are more likely to have this tPA-related angioedema than those with brainstem infarction. It is believed that damage of the insular cortex leads to autonomic dysfunction and vasomotor changes in the contralateral side. This tPA-related condition may be treated with intravenous ranitidine, diphenhydramine, and methylprednisolone.

**References:**


**Question 323: Pharmacology/Chemistry - Sleep Disorders**

**Discussion:**

Suvorexant is an orexin receptor antagonist, which inhibits the wakefulness-promoting orexin neurons of the arousal system. Suvorexant is the first drug in this class for the treatment of insomnia. (Ambien, Sanofi), eszopiclone (Lunesta, Sunovion),...
Zolpidem and zaleplon both bind to the GABA-BZ receptor at a distinct location from either benzodiazepines or barbiturates. The mechanism of action of eszopiclone is unknown but is believed to bind to the GABA receptor. Temazepam also binds to the GABA receptor.

References:


Question 325: Pharmacology/Chemistry - Headache

Discussion:

This patient has episodic cluster headache and is in a cluster cycle. A brief course of corticosteroids can achieve rapid, short-term headache remission while titrating prophylactic treatment. Sumatriptan (parenteral or intranasal) and oxygen inhalation are effective for abortive therapy for acute cluster headache. Verapamil and lithium carbonate are used for long-term prophylaxis for patients whose cluster headache cycles last more than 2 weeks. Since their onset of benefit is delayed, they should be initiated concomitantly with corticosteroids, to ensure effective headache prophylaxis after completion of steroid therapy.

References:


Question 329: Pharmacology/Chemistry - Epilepsy

Discussion:

Hepatic enzyme-inducing AEDs (carbamazepine, phenytoin, phenobarbital, primidone, and oxcarbazepine) cause elevated production of sex hormone-binding globulin. This results in increased clearance of oral contraceptives, thereby reducing birth control efficacy. Lamotrigine, and topiramate are weak-inducers of the production of sex hormone-binding globulin at higher doses. Non-enzyme inducing AEDs, such as zonisamide, levetiracetam, gabapentin, pregabalin, and lacosamide, do not appreciably alter metabolism of oral contraceptives and are thus recommended first-line agents for women with epilepsy who are on an oral contraceptive. While valproic acid does not affect oral contraceptive levels, it is highly teratogenic and thus is typically avoided in young women.

References:


Question 330: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

Fingolimod is one of the robust disease-modifying drugs for multiple sclerosis. Because of its potentially fatal cardiac complications, careful cardiac monitoring, in addition to other standard blood testing, is deemed necessary. Similar first-dose cardiac monitoring is also indicated if fingolimod therapy is interrupted for more than 2 weeks.

References:


Novartis Pharmaceuticals Corporation. Medication Guide: Gilenya (Fingolimod). East Hanover, New Jersey 07936 (Revised February 2016)

Question 331: Pharmacology/Chemistry - Epilepsy

Discussion:
Perampanel is the first-in-class anticonvulsant that inhibits the post-synaptic AMPA receptor. The mechanism of action that carbamazepine, lamotrigine, phenytoin, topiramate, valproic acid, and zonisamide have in common is slow recovery of voltage-gated sodium channels. Phenobarbital and primodone increase the duration of GABA channel opening. Clobezam increases the frequency of GABA channel opening.

References:


Question 369: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

This patient has area postrema syndrome, one of the core features of NMO spectrum disorder (NMOSD), which is associated with serum autoantibody against aquaporin-4. Other core features of NMOSD include optic neuritis and longitudinally extensive transverse myelitis. Aquaporin-4 (AQP4) water channels are located on CNS oligodendrocyte foot processes. Antibody binding to AQP4 leads to complement activation, resulting in inflammatory demyelination.

References:


Question 378: Pharmacology/Chemistry - Demyelinating Disorders

Discussion:

This patient has clinically isolated right optic neuritis, with a normal brain MRI. Based upon the optic neuritis treatment trial, a short course of intravenous steroids followed by an oral steroid taper is appropriate to hasten clinical recovery; oral prednisone increased the risk of developing optic neuritis in the fellow eye. Patients who present with a clinically isolated demyelinating syndrome and a normal brain MRI are at relatively low risk to develop clinically definite multiple sclerosis and generally should not be offered MS disease-modifying therapy initially.

References:


Question 381: Pharmacology/Chemistry - Aging, Degenerative Diseases

Discussion:

ALS is classified into sporadic ALS and familial ALS. A meta-analysis using prospective population registries suggested that about 5% of ALS is familial. Clinically, sporadic ALS and familial ALS are indistinguishable. Screening for mutations in the genes currently linked to ALS identifies known genetic variants in approximately 60% of all patients with familial ALS, with chromosome 9 open reading frame 72 (C9ORF72) representing 40%, Cu/Zn-superoxide dismutase (SOD1) 20%, fused in sarcoma (FUS) 5%, and TAR DNA-binding protein 43 (TARDBP) 3%. Mutations in C9ORF72, fused in sarcoma (FUS) 5%, and TAR DNA-binding protein 43 all affect RNA processing. Expansion of GGGGCC hexanucleotide repeats within the C9ORF72 gene produces toxic RNA that sequesters RNA binding proteins disrupting normal regulation of RNA metabolism.

References:

**Physiology**

**Question 9: Physiology - EMG**

**Discussion:**

The H reflex is a true reflex with a sensory afferent, a synapse, and a motor efferent segment. It is typically elicited in the leg by stimulating the tibial nerve and recording over the soleus muscle. The response is the electrical correlate of the S1 tendon ankle reflex. If the ankle reflex is present clinically, the H reflex should always be present. Any lesion that might decrease the ankle reflex will likely result in an abnormal H reflex (eg, polyneuropathy, proximal tibial neuropathy, sciatic neuropathy, L-S plexopathy, S1 radiculopathy).

**References:**


**Question 13: Physiology - EEG**

**Discussion:**

Favorable prognostic factors on EEG are variability, reactivity to external stimuli, varying sleep patterns, and increase in background frequencies. Poor prognostic factors are invariant pattern, no reactivity, monorhythmic pattern, burst suppression, generalized periodic discharges, very low voltage tracing, and generalized suppression. Over 96% of the patients with poor prognostic findings on the EEG following a cardiac arrest either die within a few days after the cardiopulmonary arrest, or if they survive, do so in a persistent vegetative state.

**References:**


**Question 14: Physiology - EEG**

**Discussion:**

Patients with Lennox-Gastaut have multiple seizure types, refractory to AEDs. Interictal EEG in these patients: slow spike and wave discharges.

**References:**


**Question 16: Physiology - EMG**

**Discussion:**

Jerking the arms overhead with a sudden deceleration caused a stretch injury to the long thoracic nerve in the patient. The long thoracic nerve supplies the serratus anterior muscle and a lesion of this nerve causes winging of the medial border of the scapula, medial displacement of the scapula with attempted shoulder movements, and internal rotation of the inferior angle of the scapula. The nerve has no sensory component. The mechanism of injury and clinical manifestations support a lesion in the long thoracic nerve. The axillary nerve supplies the deltoid and the suprascapular nerve supplies the supraspinatus; a lesion of either would produce shoulder abduction weakness, but not scapular winging. The dorsal scapular nerve supplies the rhomboids; a lesion of
this nerve would produce winging of the medial border of the scapula, although would produce lateral displacement of the scapula with use of the shoulder, rather than medial displacement. The thoracodorsal nerve supplies the latissimus dorsi, which abducts the humerus.

References:


Question 18: Physiology - EMG

Discussion:

Acid maltase deficiency in its most severe form, Pompe disease, presents as a hypotonic infant with macroglossia and hepatomegaly. Distinctive needle EMG findings include myotonic discharges without clinical myotonia, diffuse fibrillation potentials, and complex repetitive discharges.

References:


Question 26: Physiology - Basic Physiology

Discussion:

Fusimotor fibers transmit impulses from gamma motorneurons in the ventral horn of the spinal cord to muscle spindles. This system allows muscle spindles to remain sensitive over a broad range of muscle size by shortening the spindle as the muscle shortens.

References:


Question 41: Physiology - EMG

Discussion:

The extensor indicis proprius (EIP) is innervated by the posterior interosseous nerve. This is derived from the posterior cord, lower trunk of the brachial plexus. The ulnar and median innervated muscles of the hand are supplied by the lower trunk, medial cord of the brachial plexus. Thus, if the EIP is involved, then the medial cord could not be the primary site of the lesion.

References:


Question 53: Physiology - EMG

Discussion:

The needle EMG shows evidence of axonal injury to the right femoral, obturator nerve and the proximal branch innervating iliopsoas muscle with absent sensory response in the saphenous nerve. The findings localize the lesion to the lumbar plexus.

References:
Question 69: Physiology - EMG

Discussion:

The patient has severe facial weakness secondary to severe facial neuropathy. Based on symptoms, the lesion is intracranial, between the branches of the facial nerve to the stapedius and to the tongue. The nerve conduction studies, with stimulation distal to the lesion, are performed two weeks from onset, beyond the time when Wallerian degeneration should have occurred in any transected axon, and the CMAP amplitudes are the same bilaterally, arguing against axonal loss and thereby supporting profound demyelination as the underlying pathophysiology for her severe facial weakness. In this regard, the absence of abnormal spontaneous or insertional activity is supportive as well, arguing against axonal loss, but is still early (prior to 3 weeks) for full development of denervation changes. While axonal lesions tend to have a poor prognosis, demyelinating lesions tend to do well. In sum, the patient has an intracranial facial neuropathy with severe demyelination as the causative pathophysiology, which generally carries a good prognosis for recovery of facial function.

References:


Question 85: Physiology - EEG

Discussion:

Small sharp spikes, also known as benign epileptiform transients of sleep, are benign sporadic sleep spikes occurring mainly in adults during drowsiness and light sleep. They are usually low-voltage, short duration, diphasic spikes with steep descending limb. They do not distort the background and are not associated with rhythmic slow wave activity. They have no significance for the diagnosis of epileptic seizures.

References:


Question 87: Physiology - EMG

Discussion:

Early signs of Guillain-Barré Syndrome include diminished or lost muscle stretch reflexes. Electrophysiological studies reveal slowing of peripheral conduction velocity as well as increased central conduction time, including prolonged F waves and distal motor latencies. Interestingly, the disease tends to spare the sural nerve sensory action potential, often regarded as one of the first affected in other neuropathies. Reduction of amplitude of compound muscle action potentials with distal stimulation implies a poor prognosis.

References:


Question 89: Physiology - EMG

Discussion:
Homozygous mutation in the SMN gene results in spinal muscular atrophy. These infants will show normal sensory conduction studies with reduced motor CMAP amplitude and normal motor nerve conduction velocity for age.

References:

Question 101: Physiology - EEG

Discussion:
Multiple technical criteria are used for recording EEG data in suspected cases of brain death, criteria essentially designed to ensure technically adequate quality recordings and to maximize the detection of even low-amplitude cerebral activity. These criteria include the use of at least 8 scalp electrodes, 10 cm interelectrode distances, and recording sensitivities of at least 2 microvolts per millimeter; other criteria exist as well, the reader is referred to the reference below for a full listing.

References:

Question 109: Physiology - EMG

Discussion:
The posterior interosseous nerve, a distal branch of the radial nerve, supplies the extensor muscles in the forearm, with the exception of the ECRL. The radial nerve supplies the triceps and extensor carpi radialis longus muscles proximal to the branchpoint of the posterior interosseous nerve (PIN). As such the EMG findings are limited to the distribution of the PIN, sparing more proximal radial-innervated muscles.

References:

Question 115: Physiology - Basic Physiology

Discussion:
Patients with demyelinating neuropathies develop paralysis primarily because of conduction block, rather than slow conduction velocity. Conduction block causes failure of nerve transmission, leading to inability of the nerve to cause muscle contraction and, thereby, loss of power.

References:

Question 124: Physiology - Basic Physiology

Discussion:
Depolarization of action potentials causes an initial negative departure from baseline due to rapid opening of voltage-sensitive sodium channels. Closing of these channels due to an intrinsic inactivation time constant as well as slow opening of voltage-sensitive potassium channels results in repolarization of the neuron.

References:
Question 126: Physiology - EMG

Discussion:

The short head of the biceps femoris is innervated by the common peroneal nerve. This muscle would be spared in a lesion of the peroneal nerve at the fibular head.

References:


Question 134: Physiology - EEG

Discussion:

The entity of benign seizures of childhood (Sylvian or Rolandic) is characterized by focal tingling or jerking of the side of the face or hand, salivation, inability to speak and progression to a generalized tonic-clonic seizure. The EEG shows spike discharges over the central midtemporal region which are characteristic of this entity.

References:


Question 138: Physiology - Autonomic Studies

Discussion:

Deep breathing with monitoring of heart rate and blood pressure can be used in evaluating both the afferent and the efferent portions of the vagal system. With inspiration heart rate rises and blood pressure falls while the opposite occurs during exhalation. Vagal dysfunction can produce a loss of these changes and particularly of heart rate variability with respiration.

References:

Iodice V and Sandroni P. Autonomic Neuropathies. Continuum (Minneap Minn); 2014; 20(5): 1373-1397

Question 149: Physiology - EMG

Discussion:

The tibialis posterior muscle is derived from the L5 and S1 myotomes, but it is supplied by the tibial nerve. An abnormality in this muscle would rule out a peroneal mononeuropathy at the fibular head. The peroneus longus and extensor hallicus longus could both be involved in an L5 radiculopathy or peroneal mononeuropathy and would therefore not distinguish the two. The gastrocnemius is an S1 muscle supplied by the tibial nerve, and the vastus lateralis is an L3 and L4 muscle supplied by the femoral nerve.

References:


Question 157: Physiology - EMG

Discussion:
This patient most likely has Guillain-Barre' syndrome. While the earliest abnormality in GBS on NCS is prolonged or absent F-waves, the earliest finding on needle EMG is reduced recruitment. It is too early in the course to see active denervation, which usually does not appear for 1-2 weeks. Enlarged motor units indicate a chronic denervating/reinnervating condition and small motor units indicate a muscle disorder.

References:


Question 162: Physiology - EEG

Discussion:

Alzheimer disease is associated with a decrease or loss of alpha and beta activity at an earlier stage than other disorders associated with dementia. Generalized periodic sharp waves and invariant alpha pattern are poor prognostic indicators on EEG and are usually seen after cardiopulmonary arrest. Triphasic waves are seen in 50% of patients with hepatic coma. Excessive beta activity may be seen due to medications like benzodiazepines or barbiturates.

References:


Question 172: Physiology - EMG

Discussion:

The patient's examination localizes to the right L5 root. Both the gluteus medius and peroneus longus muscles receive their major innervation from the L5 nerve root.

References:


Question 174: Physiology - EMG

Discussion:

Fasciculation potentials result from spontaneous discharges of a whole or possibly part of a motor unit. The generator source of nearly all fasciculations has a motor axonal origin. Fasciculation potentials, although typically associated with diseases of anterior horn cells, are also seen in radiculopathy, entrapment neuropathy, and the muscular pain-fasciculation syndrome.

References:


Question 175: Physiology - EEG

Discussion:

Rare frontal sharp waves can be seen on EEG in a normal full-term baby and does not indicate underlying abnormality or epileptiform activity.
References:


Question 182: Physiology - EMG

Discussion:

Sensory studies aid in the localization of predorsal or postdorsal root ganglion regions. Lesions proximal to dorsal root ganglia, like radiculopathy, have normal sensory nerve action potentials. Sensory nerve action potentials are abnormal in lesions distal to dorsal root ganglia. This patient has C8-T1 radiculopathy with likely abnormalities in first dorsal interosseus and abductor pollicis brevis muscles.

References:


Question 209: Physiology - EEG

Discussion:

Panayiotopoulos syndrome is associated with unilateral occipital spikes. Panayiotopoulos syndrome is a benign (non-lesional), age-related focal epilepsy of childhood. It differs from benign rolandic epilepsy primarily in the length of episodes, the frequent occurrence of vomiting, and the lack of typical central-temporal spikes on EEG. While acute, confusional migraine may include vomiting, episodes do not typically begin during sleep. Juvenile absence epilepsy (JAE) has 3-5 Hz generalized spike and polyspike and slow wave bursts. Benign neonatal convulsions may be associated with a range of paroxysmal abnormalities superimposed upon a normal background. Rasmussen encephalitis usually has unilateral slowing and spikes.

References:


Question 229: Physiology - EEG

Discussion:

An EEG showing triphasic waves is consistent with a metabolic encephalopathy, most commonly hepatic encephalopathy though it is certainly not entirely specific.

References:


Question 235: Physiology - EMG

Discussion:

In patients with Lambert-Eaton myasthenic syndrome, a characteristic feature of motor nerve conduction studies is reduced compound muscle action potential (CMAP) amplitude. The diagnostic finding in LEMS is a marked increase in CMAP amplitude (increment) following brief exercise or rapid repetitive stimulation (more than 5 hz). This phenomenon occurs because exercise or rapid repetitive stimulation causes calcium ingress into the nerve terminal to exceed egress, resulting in a brief rise in
intracellular calcium in the nerve terminal. Calcium is needed for fusion of acetylcholine quanta with the pre-synaptic membrane. Thus, brief exercise or rapid stimulation facilitate release of Ach from the nerve terminal. This returns CMAP amplitude to normal briefly.

References:


**Question 250: Physiology - EEG**

**Discussion:**

The 14 and 6 phantom spikes occur bilaterally synchronous and diffuse fashion. The bursts are brief and last 1 to 2 seconds. At times the 14 Hz or, more commonly, the 6 Hz pattern may be seen in isolation. The pattern is considered a normal variant but can easily be confused with epileptiform discharges.

References:


**Question 251: Physiology - EEG**

**Discussion:**

Hypsarrhythmia is a characteristic pattern seen in a majority of infants with infantile spasms. It is characterized by high-voltage and irregular slow waves with multiple sharp and spike discharges.

References:


**Question 268: Physiology - EEG**

**Discussion:**

Periodic lateralizing epileptiform discharges (PLEDS) indicate unilateral hemispheric lesion and are the most common finding in herpes encephalitis. 3 Hz spike/wave discharge is an diffuse epileptic discharge composed of a spike with after-coming slow wave at 3/sec. FIRDA is a slow bifrontal discharge without a sharp component. Triphasic waves are seen in metabolic encephalopathy and are bilateral and frontally predominant. Breach rhythm is seen overlying a skull defect in which normal discharges are seen at higher than normal amplitudes.

References:


**Question 269: Physiology - EEG**

**Discussion:**

The background activity is posteriorly dominant alpha rythm at 9 to 9.5 Hz. This is normal waking occipital activity. The normal occipital background rhythm disappears on eye opening which is shown in the EEG tracing.

References:

**Question 274: Physiology - Evoked Potentials**

**Discussion:**

As Wave I is generated by the VIIIth nerve, it is normal for it to be absent on the side opposite the stimulated ear. After this point in the ascending auditory pathway the signals are traveling both ipsilaterally and contralaterally.

**References:**


**Question 277: Physiology - EEG**

**Discussion:**

Tonic seizures are manifested on EEG by a burst of generalized fast activities, often preceded by a generalized spike and wave. These patterns often have a feel of generalized suppression due to low voltages. Focal sensory or motor seizures would be expected to demonstrate focal evolving rhythmic electroencephalographic seizure patterns. Likewise, complex partial seizures (e.g., producing lip-smacking) should demonstrate a focal temporal lobe electroencephalographic seizure pattern. Generalized clonic seizures should have a similar, although generalized, evolving electroencephalographic seizure pattern.

**References:**


**Question 282: Physiology - EMG**

**Discussion:**

The tracing shows a markedly prolonged distal latency with a small, dispersed motor response and decreased velocity. The presence of temporal dispersion and/or conduction block favors an acquired demyelinating polyneuropathy such as chronic inflammatory demyelinating polyneuropathy (CIDP).

**References:**


**Question 288: Physiology - EEG**

**Discussion:**

The EEG pattern shown in the question is that of electrocerebral inactivity. ECI is the pattern observed in brain death evaluations. The characteristic EEG pattern of West Syndrome is hypsarrhythmia. Following MCA infarction, focal abnormalities might be expected. ECI is not the typical pattern associated with status epilepticus. Multiple patterns may be seen in encephalopathy with the classic pattern that of diffuse slowing with decreased reactivity.

**References:**
Discussion:

The EEG finding in the figure is increased amplitude and beta frequencies in the left central region, consistent with a breach rhythm. Breach rhythm is often seen following craniotomy and involves increased voltages and increased amounts of faster (typically beta) frequencies recorded from the region of the skull defect. This excess beta activity often creates a sharply-contoured appearance to the underlying rhythms or may appear to displace a normal physiologic rhythm (for example, the alpha rhythm may appear to be maximal more anteriorly than typically seen, if there is a more anterior skull defect). The EEG depicted otherwise demonstrates normal waking background characteristics except for increased voltages and faster frequency elements in the left central lead. Breach rhythm is seen following procedures that violate the integrity of the skull, such as ventriculoperitoneal shunting. No change in the EEG background is expected with pontine infarction. Subarachnoid hemorrhage might have a normal EEG pattern or result in findings of depressed levels of consciousness. Hydrocephalus is not associated with focal EEG findings. Subdural hematoma would result in focal decrease in recorded voltages, often with suppression of faster frequency elements, which is not seen in this case.

References:


Question 293: Physiology - EEG

Discussion:

Juvenile myoclonic epilepsy is characterized by generalized polyspike wave discharges. These findings are often brought out by photic stimulation. Studies demonstrate that "photoparoxysmal discharges" - polyspike wave discharges evoked by photic stimulations - that outlast stimuli are often associated with epilepsy, whereas those that do not outlast stimuli can be seen in asymptomatic individuals.

References:


Question 313: Physiology - EEG

Discussion:

Nonconvulsive status epilepticus can be prolonged and is associated with fairly abrupt deterioration in mental function and a paroxysmal EEG. These features differentiate nonconvulsive status from the dementias.

References:


Question 321: Physiology - Sleep

Discussion:

Patients with excessive daytime sleepiness must be evaluated for sleep apnea, narcolepsy, idiopathic hypersomnia, restless legs syndrome, or parasomnias. The absence of any abnormality on the overnight polysomnography and multiple sleep latency test other than less-than-5-minute sleep latency is consistent with idiopathic hypersomnia. A normal PSG the night prior to the MSLT is important to ensure that sleepiness is not due to undiagnosed OSA, for example.

References:


Question 348: Physiology - EMG

Discussion:

Myotonic dystrophy is a multisystem disease that can cause cardiac conduction defects, early cataracts, frontal balding, ptosis, facial weakness, hip girdle and finger flexor weakness as well as hypogammaglobulinemia, endocrine and CNS abnormalities. Electrophysiologic studies demonstrate myotonic discharges, even when there is minimal to no clinical myotonia elicitable.

References:


Question 350: Physiology - Evoked Potentials

Discussion:

In upper extremity somatosensory evoked potential (SEP), after stimulation of the median or ulnar nerve in the wrist, activity can be recorded at the elbow, Erb’s point cervical spine, and scalp. The N5 potential is recorded in the median or ulnar nerve. The N9 potential is recorded at the Erb’s point. The N11 potential is recorded at the dorsal root entry zone. The N13 potential is created by dorsal column of the cervical cord and N14 potential represents activity in the nucleus cuneatus or medial lemniscus at the cervicomedullary junction. The activity recorded over the frontal lobe is reflected in the N30 peak.

References:


Question 353: Physiology - Sleep

Discussion:

The goal of the multiple sleep latency test is to quantitate sleepiness during waking hours and to determine the occurrence of REM sleep near sleep onset. The goal of polysomnography is to quantitate the amount of time spent in various stages of sleep during the night and to document clinically relevant events such as cardiopulmonary abnormalities or sleep-related abnormal motor activity. Electrooculogram, surface electromyography of all four limbs, and EEG recording are all used while performing polysomnography and multiple sleep latency test. It is important to perform a PSG the night prior to performing an MSLT in order to assess the quality of sleep the night prior. This is particularly true when assessing for narcolepsy as sleepiness due to poor quality fragmented sleep could result in an MSLT which appears to indicate narcolepsy.

References:


Question 371: Physiology - EEG
Discussion:

The location of the ictal focus is the most important factor in determining seizure onset during chronic intracranial EEG monitoring and this is best represented by ictal onset. The subsequent expression of the ictal discharge, while important, is less useful in localizing the epileptogenic zone.

References:


Question 373: Physiology - Sleep

Discussion:

REM sleep behavior disorder is seen in older patients and is characterized by REM sleep without muscle atonia and various abnormal motor activities during sleep. Benign sleep myoclonus would be characterized by occasional very brief shock-like jerks and wouldn't be characterized as violent leg movements. This patient had no evidence given of a sleep associated breathing disorder like obstructive sleep apnea - particularly, no oxygen desaturations were noted. Restless legs syndrome is a disorder of wakefulness while periodic limb movements of sleep may be seen during sleep - either as a disorder or as an occurrence without impact on overall sleep quality (PLMD vs PLMS).

References:


Question 379: Physiology - Basic Physiology

Discussion:

Pacinian corpuscles are responsible for perception of vibration. Free nerve endings are associated with pain perception. Ruffini endings are also associated with vibration. Meissner corpuscles and Merkel receptors are associated with superficial touch.

References:


Question 385: Physiology - EMG

Discussion:

Lesions of the upper trunk of the brachial plexus will cause weakness in proximal upper extremity with abnormal SNAPs on NCS in the radial and median nerves. The deltoid muscle is innervated by the axillary nerve which arises from the upper trunk/posterior cord. The biceps muscle is innervated by the musculocutaneous nerve which arises from the upper trunk/lateral cord.

References:


Question 388: Physiology - EEG

Discussion:
Absence seizures can be misdiagnosed as attention deficit disorder. Observance at school may be helpful in gaining an appropriate history. Family history of seizures is also helpful. EEG shows 3 Hz spike-and-wave.

References:


Question 391: Physiology - EMG

Discussion:

In compression neuropathies, sensory fibers are usually affected first with conduction changes. The median palmar sensory latency prolongation is the earliest change seen on nerve conduction studies in patients with mild carpal tunnel syndrome.

References: