Brigham and Women's Hospital
Founding Member, Mass General Brigham

Board Review: Neurology

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-Clinical and research focus women's neurology

DISCLOSURES

- Consultative work for Crico
- I do consultative work for Teladocs
- I receive royalties from Oxford and up to date

Objectives

Use cases as a format:

- To review selected neurologic topics
- Discuss the differential diagnosis and neuroanatomy

Question 1:

A 47 year-old hiker has pain in the left buttock that radiates to her left lateral foot. The pain is not affected by position or movement.

Question 1 (cont'd):

She also says that her left foot tingles as do parts of her right foot and thigh, left fingers, and the left anterior part of her chest.

Question 1 (cont'd):

One month ago, a physician treated her for a right Bell palsy and arthritis.

Question 1 (cont'd):

Which of the following would be most helpful in establishing a diagnosis?

A. Electromyography and nerve conduction velocity study

B. Somatosensory evoked responses

C. Magnetic resonance imaging of the spine and spinal cord

D. Lumbar puncture

E. Lumbar spine XRay

Question 1:

The answer is D

Question 1 Answer

Which of the following would be most helpful in establishing a diagnosis?

A. Electromyography and nerve conduction velocity measurements

B. Somatosensory evoked responses

C. Magnetic resonance imaging of the spine and spinal cord

D. Lumbar puncture

E. Lumbar spine XRay

Lyme disease can present with cranial neuropathies (typically the facial nerve, may be bilateral, occurs in 5-10% of untreated patients) and inflammatory radiculopathy (may mimic a mechanical radiculopathy with pain and sensorimotor symptoms). Lumbar puncture will rule out other inflammatory, infectious or neoplastic etiologies and will be abnormal and diagnostic in Lyme disease. CSF will typically show a lymphocytic pleocytosis. EMG/NCS and MRI may be abnormal, but would not provide a definitive diagnosis. An X-ray of the lumbar spine (lumbar roetenography) would not be helpful.

Question 2:

A 78 year-old normotensive woman realizes suddenly that she cannot see to her left and that her left hand tingles.

Question 2 (cont'd):

Two years previous, she had a stroke which left her right arm and right leg weak. The family reports that during the past year she has become unreliable in daily responsibilities, and often forgets people's names.

Question 2 (cont'd):

Computed tomographic scan shows a recent, well-circumscribed homogeneous right parietal-temporal hemorrhage; an old, slit-like cavity in the left medial frontal lobe under the cortex; and moderate ventricular dilatation and cortical sulcal widening.

Question 2 (cont'd):

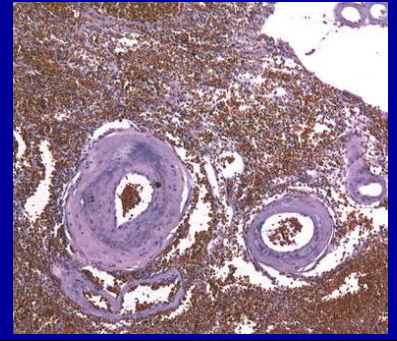
The most likely diagnosis is:

A. Hemorrhage into a brain tumor
B. Cerebral amyloid angiopathy
C. Embolization of cardiac origin with hemorrhagic infarction
D. Multiple cerebral aneurysms
E. Recurrent head trauma

Question 2:

The answer is B

Question 2 answer



The most likely diagnosis is:

A. Hemorrhage into a brain tumor

B. Cerebral amyloid angiopathy

C. Embolization of cardiac origin with hemorrhagic infarction

D. Multiple cerebral aneurysms

E. Recurrent head trauma

Her acute symptoms of a left hemifield vision loss and left hand somatosensory complaints are explained by the recent right parietal-temporal hemorrhage. Her prior stroke resulted in right sided weakness and is explained by the lesion in the left frontal lobe. A slit-like cavity is seen as a residual finding after a hemorrhage. Therefore she has had at least 2 intracerbral hemorrhages and has a possible dementia (family's report of one year of being unreliable). ICH is the most recognized complication of CAA.

Question 3:

After skiing, a 27 year-old woman developed pain in the left mastoid region that radiated to the left occiput and neck.

Question 3 (cont'd):

The next day she became dizzy, staggered, and felt pain in the left forehead and eye.

Question 3 (cont'd):

Physical examination shows normal blood pressure, but with abnormal neurologic findings including:

- decreased pin perception on the left side of the face and the right limbs and trunk;
- a left Horner syndrome;
- rotatory nystagmus;
- weakness of the left palate and pharynx;
- clumsiness and uncoordination of the left limbs.

Question 3 (cont'd):

The most likely diagnosis is:

A. Embolization to the basilar artery with pontine infarction
B. Pontine hemorrhage
C. Dissection of the left vertebral artery
D. Dissection of the left internal carotid artery
E. Artherosclerotic occlusion of the internal carotid artery

Question 3:

The answer is C

Question 3 answer

The most likely diagnosis is:

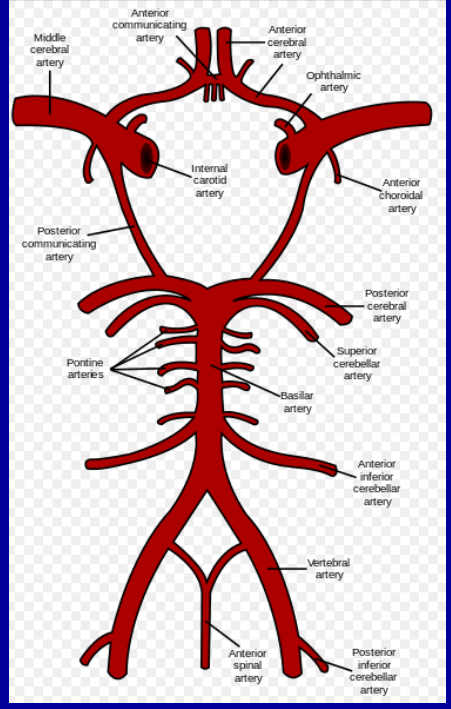
A. Embolization to the basilar artery with pontine infarction

B. Pontine hemorrhage

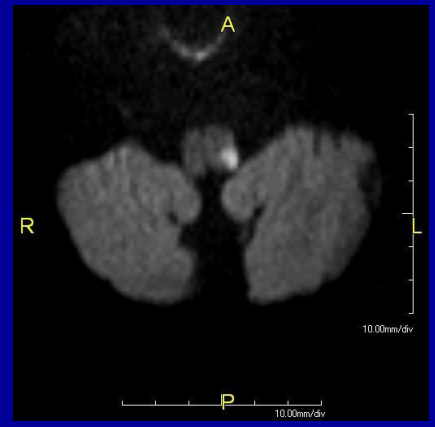
C. Dissection of the left vertebral artery

D. Dissection of the left internal carotid artery

E. Artherosclerotic occlusion of the internal carotid artery



Vertebral artery dissection can present with ipsilateral occipital or neck pain. Her examination is consistent with a left lateral medullary syndrome (Wallenburg syndrome). This area of the medulla is supplied by the PICA (posterior inferior cerebellar artery), a branch of the vertebral artery. The lateral meduallary syndrome consists of: crossed sensory findings with ipsilateral loss of pain and temperature to the face and contralateral to the body, ipsilateral horner syndrome, ipsilateral ataxia, weakness of the larynx and pharynx leading to dysphagia, dysarthria, vertigo.



Question 4:

A 64 year-old man has progressive spasticity of gait, impotence, and urinary frequency. He has occasional headaches at the vertex.

Physical examination shows increased tone in the bilateral lower extremities, 3+ patella and ankle reflexes and bilateral extensor plantar reflexes.

Question 4 (cont'd):

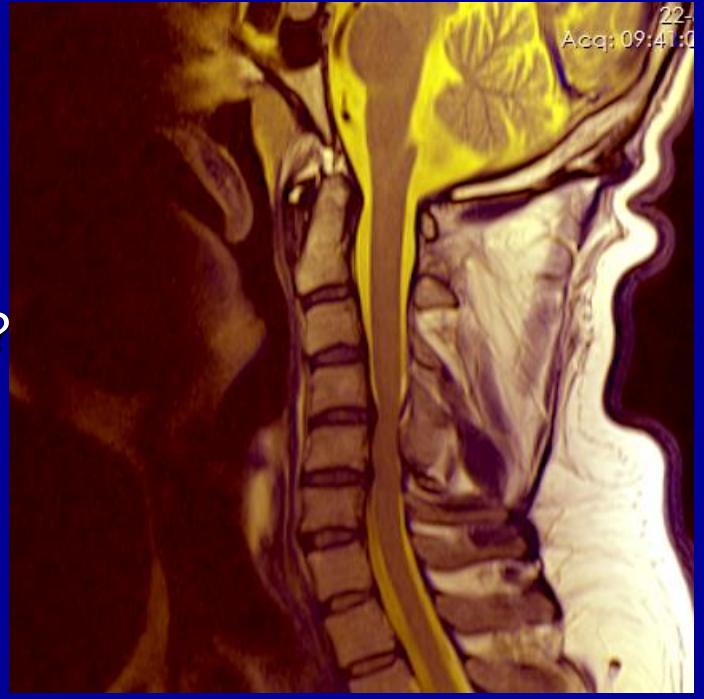
Which of the following is the next best step in his care?

A. Physical therapy
B. Magnetic resonance imaging of the cervical spine
C. EMG and NCV
D. CT of the lumbar spine
E. Cerebral angiogram

Question 4:

The answer is B

Question 4 answer:



Which of the following is the next best step in his care?

A. Physical therapy

B. MRI of the neck

C. EMG and NCV

D. CT of the lumbar spine

E. Cerebral angiogram

MRI of the neck is the test of choice to assess for cervical myelopathy. Cervical cord compression can lead to progressive spasticity in gait, sexual dysfunction, and spastic bladder (urinary frequency). The most common etiology of spinal cord dysfunction in the elderly is cervical spondylosis. Signs of cervical canal narrowing can be seen on plain films, CT, MRI, and CT myelogram. EMG/NCV are rarely helpful, but can be used when assessing for peripheral nerve pathology.

Question 5:

An 80 year-old woman with mitral valve disease and chronic atrial fibrillation becomes suddenly confused during a family dinner.

Question 5 (cont'd):

She is awake and alert, and her motor function appears intact and symmetric. She speaks in long sentences unconnected to the events of the evening or the questions asked of her.

Question 5 (cont'd):

She uses many word substitutions and nonsense words. She appears unable to understand questions put to her by family members.

Question 5 (cont'd):

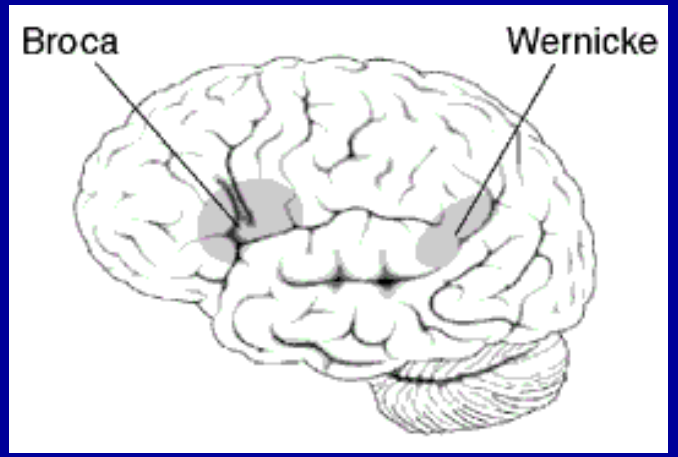
The most likely diagnosis is:

A. Complex partial seizure
B. Transient global amnesia
C. Dominant hemisphere stroke
D. Nondominant hemisphere stroke
E. Delirium

Question 5:

The answer is C

Question 5 answer



The most likely diagnosis is:

A. Complex partial seizure

B. Transient global amnesia

C. Left (dominant) hemisphere stroke

D. Right (nondominant) hemisphere stroke

E. Delirium

She has risk factors for stroke (AF) and has sudden onset of deficits. She speaks fluently but uses word substitutions (paraphasias) and nonsense words (neologisms). She has impaired comprehension. This description is classical for Wernicke's type aphasia and localizes to the left hemisphere involving the posterior temporal lobe (Wernicke's area). There are often no motor deficits because the motor pathways are spared. Sometimes there is a visual field cut on the right due to involvement of the optic radiations. A lesion involving the optic radiations in the temporal lobe will result in a superior quadrantonopsia. Over 95% of right handed people and the majority of left handed people have language in the left hemisphere. Therefore it is called the dominant hemisphere (short for dominant hemisphere for language).

Question 6:

A 48 year-old man comes to the physician complaining of right leg pain and numbness for the past 3 weeks. The pain is on the upper, lateral thigh and is burning and stinging in quality. The pain radiates to the groin and down to the knee. He has a PMH of obesity and has not seen a doctor for many years.

Question 6 (cont'd):

On examination, he is obese and otherwise well-appearing. Strength is 5/5 bilaterally. There is decreased sensation in the right lateral thigh to temperature and pinprick. There is decreased sensation to pinprick, temperature and vibration in a bilateral stocking distribution to the mid-calves bilaterally. Reflexes are 1+ at the patella and 0 at the Achilles and bilateral flexor plantar reflexes bilaterally.

Question 6 (cont'd):

Which of the following is the best diagnostic test to obtain at this time?

A. Hemoglobin A1C

B. Vitamin B12

C. EMG

D. MRI of the lumbar spine

E. CT of the pelvis

Question 6:

The answer is A

Question 6 answer:

Which of the following is the best diagnostic test to obtain at this time?

A. Hemoglobin A1C

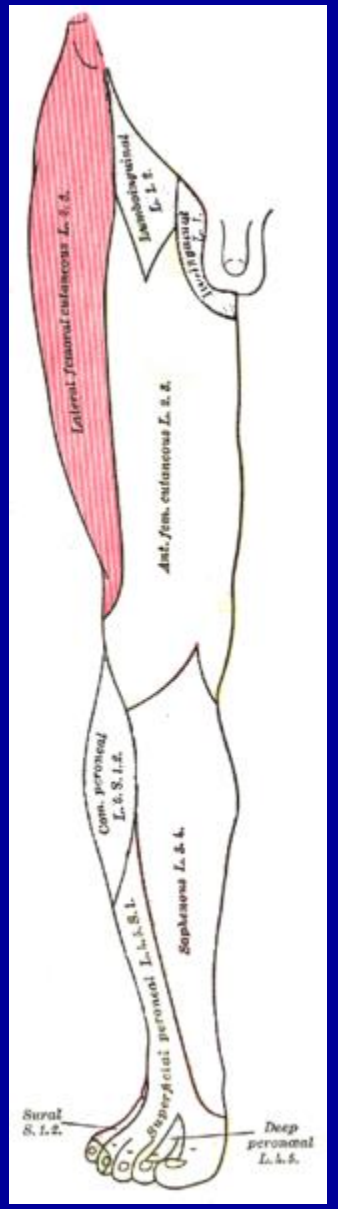
B. Vitamin B12

C. EMG

D. MRI of the lumbar spine

E. CT of the pelvis

The patient is presenting with meralgia paresthetica, a neuropathy of the lateral femoral cutaneous nerve. This nerve can be compressed by tight fitting clothing, often in association with weight gain. Weight loss can also lead to this condition, presumably by decreasing the fat padding around this nerve. The diagnosis is made clinically in the setting of characteristic sensory changes with normal motor function. It is more common in patients with diabetes mellitus. This patient has signs of a peripheral neuropathy as well that may be due to undiagnosed diabetes. A B12 deficiency causes a myelopathy as well as neuropathy and would not present in this fashion.



Question 7:

Ten days ago, a 22 year-old man had an inoculation of tetanus toxoid in his right arm after removal of a splinter.

Question 7 (cont'd):

He now has severe pain in the right shoulder and arm and parasthesias in the right hand.

Question 7 (cont'd):

Physical examination shows severe weakness of muscles around the right shoulder girdle and absence of the right biceps reflex.

Question 7 (cont'd):

Passive range of movement of the shoulder is normal. Sensory examination is normal, and other reflexes in the arms and legs are normal.

Question 7 (cont'd):

The most likely diagnosis is:

A. Brachial neuritis
B. Herniated C-7 disc
C. Epidural cervical spinal abscess
D. Cervical spinal cord tumor
E. Rotator cuff injury

Question 7:

The answer is A

Question 7 answer:

The most likely diagnosis is:

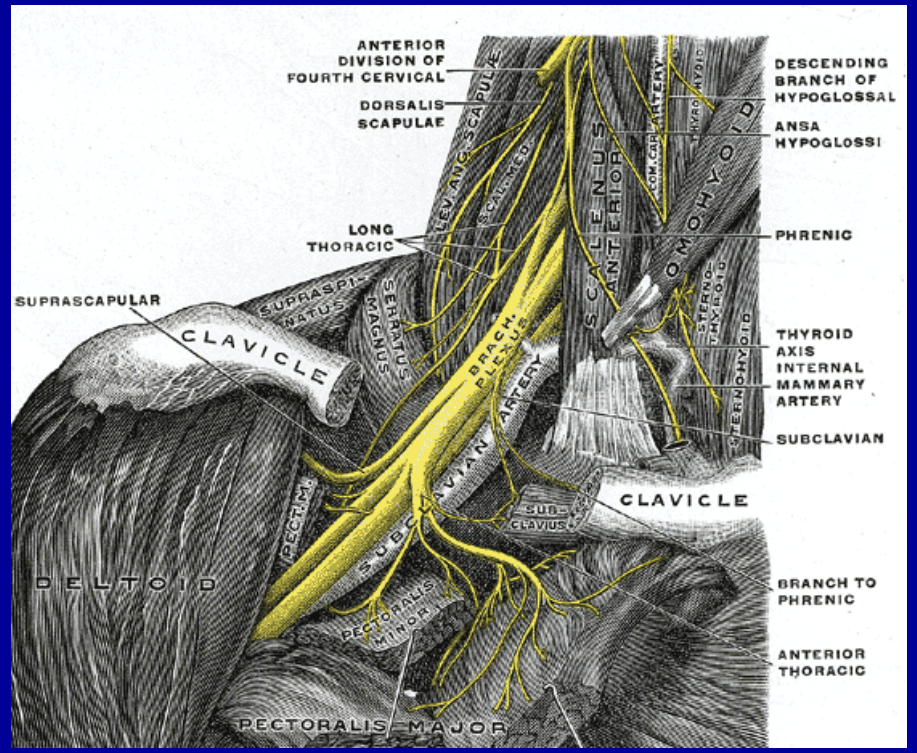
A. Brachial neuritis

B. Herniated C-7 disk

C. Epidural cervical spinal abscess

D. Cervical spinal cord tumor

E. Rotator cuff injury



"Parsonage-Turner syndrome" is characterized by inflammation of the brachial plexus. It typically presents with severe shoulder pain and then as the pain is improving, there is weakness in the deltoid and periscapular muscles. It is often idiopathic, but may be autoimmune and triggered by immunization. There is typically good recovery and treatment is supportive with physical therapy and pain control. There is no role for immunosuppression. A C7 disc herniation would lead to weakness in the triceps and wrist extensors and the biceps reflex would not be affected.

Question 8:

A 60 year-old man with a long history of back pain recently began feeling weakness and tingling in his legs when he walks more than a half a block. The symptoms disappear when he sits.

Question 8 (cont'd):

He has no symptoms when doing bicycling-like exercises supine on his bed even after 30 minutes.

Question 8 (cont'd):

Except for an absent left ankle reflex, neurologic examination is normal. Foot and femoral pulses are normal.

Question 8 (cont'd):

The most likely diagnosis is:

A. Aortic atherosclerosis with claudication
B. Polyneuropathy
C. Herniated lumbar L-5 disk
D. Lumbar spinal stenosis
E. Cervical spondylitic myelopathy

Question 8:

The answer is D

Question 8 answer:

The most likely diagnosis is:

A. Aortic atherosclerosis with claudication
B. Polyneuropathy
C. Herniated lumbar L-5 disk
D. Lumbar spinal stenosis
E. Cervical spondylitic myelopathy

"Neurogenic claudication"
Lumbar stenosis can be asymptomatic, associated with low back pain, cause symptoms and signs of focal nerve root compression, or give rise to neurogenic claudication Neurogenic claudication refers to pain and discomfort in the low back, buttocks, and legs that occurs after walking and is relieved by sitting. Relief of symptoms with flexion of the spine explains why it is often easier to walk up an incline than on a level surface, and forms the basis of the bicycle test. A patient with neurogenic claudication will be able to cycle (spine flexed), but will not be able to walk erect (spine extended) for an equivalent time. A patient with vascular claudication is expected to have the same tolerance for both activities.

Question 9:

A 25 year-old previously healthy man is found unconscious in his apartment. There is no evidence of trauma.

Question 9 (cont'd):

On examination, he is responsive to voice and painful stimulation. There is no evidence of meningeal irritation. The pupils are 3 mm and unreactive.

Question 9 (cont'd):

There is no inducible eye movements by the doll's eye maneuver or irrigation of a tympanic membrane with ice water.

Question 9 (cont'd):

The blood pressure is 90/70 mm Hg, and the pulse rate is 54/min. Respiratory function is depressed.

Question 9 (cont'd):

The most likely diagnosis is:

A. Sedative drug overdose
B. Subarachnoid hemorrhage
C. Intracranial mass
D. Brain stem stroke
E. Narcotic overdose

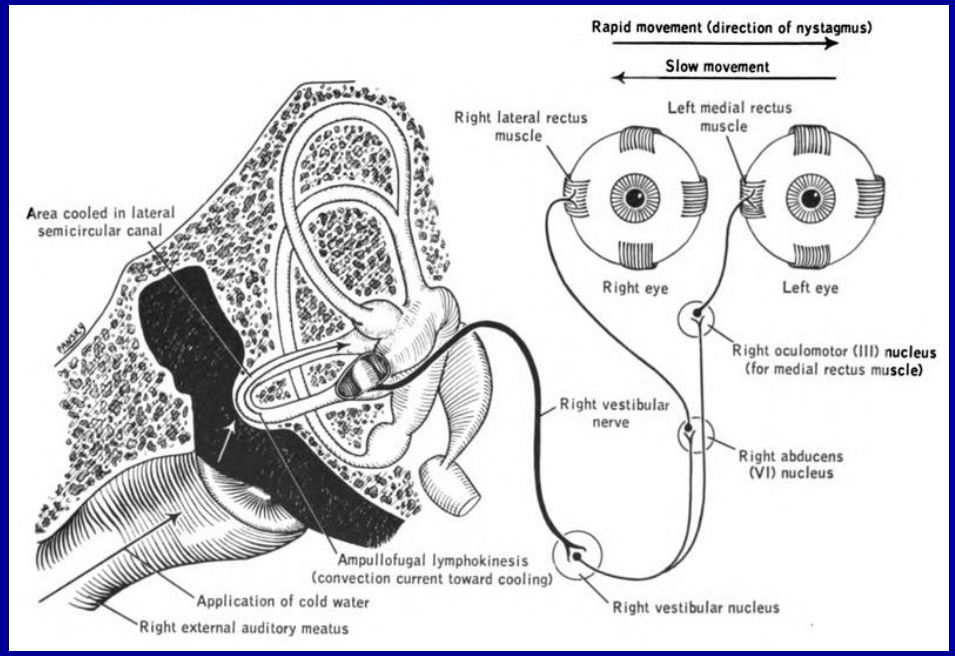
Question 9:

The answer is E

Question 9 answer

The most likely diagnosis is:

A. Sedative drug overdose
B. Subarachnoid hemorrhage
C. Intracranial mass
D. Brain stem stroke
E. Narcotic overdose



The clues to this diagnosis are that he has small pupils and has depressed respiratory rate. This patient has impaired brain stem function, which can occur temporarily with drug overdose. Both sedative and narcotic drug overdose can lead to absent oculovestibular reflexes, but pupils are not small with a sedative drug overdose.

Question 10:

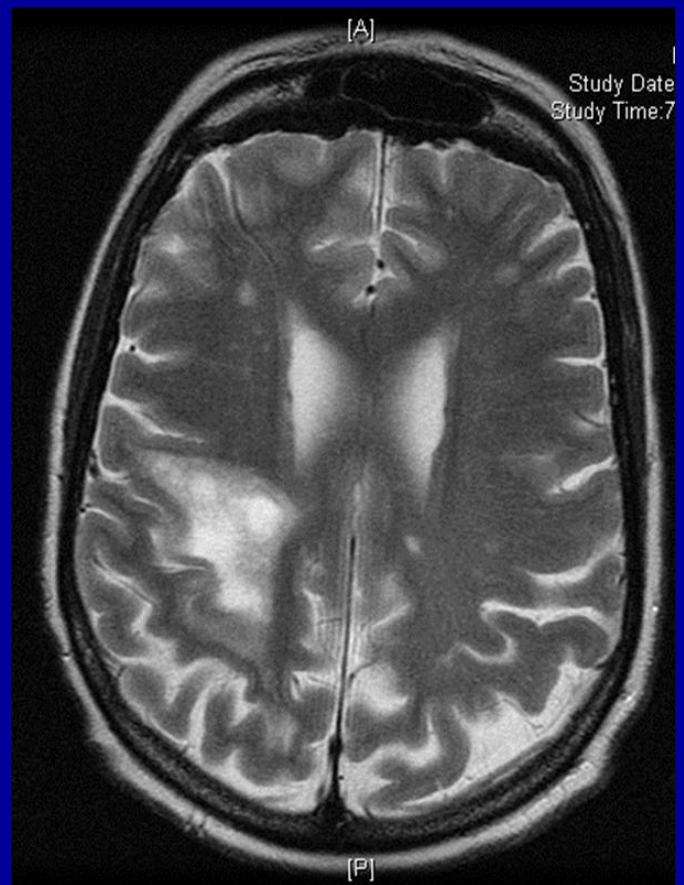
A 35 year-old woman, who had a renal transplant 4 years ago for renal failure due to membranous glomerulonephritis, is hospitalized because of progressive left sided weakness and dysarthria.

Question 10 (cont'd):

She has been treated with prednisone and cyclosporine. A test for the human immunodeficiency virus is negative.

Question 10 (cont'd):

MRI of the brain shows, in the right frontal lobe, a focal area of abnormal signal, that spared the cortical gray matter.



Question 10 (cont'd):

There is minimal mass effect and no contrast enhancement.
There are also similar smaller lesions throughout the white matter.

Question 10 (cont'd):

The most likely diagnosis is:

A. Multiple sclerosis
B. Glioma
C. Embolic stroke
D. Progressive multifocal leukoencephalopathy
E. Primary central nervous system lymphoma

Question 10:

The answer is D

Question 10 answer

The most likely diagnosis is:

A. Multiple sclerosis

B. Glioma

C. Embolic stroke

D. Progressive multifocal leukoencephalopathy

E. Primary central nervous system lymphoma

The patient is immunosuppressed leaving her vulnerable to reactivation of the JC virus. This disease is usually associated with AIDS, but is also seen patients immunocompromised due to other reasons. This virus causes destruction of the CNS white matter by infecting the oligodendrocytes. With acute lesions of multiple sclerosis, glioma and with lymphoma there is often enhancement due to break down of the blood brain barrier. Additionally, MS requires 2 separate episodes of demyelination in time and space. A stroke affects the gray matter as well as the white matter.

Question 11:

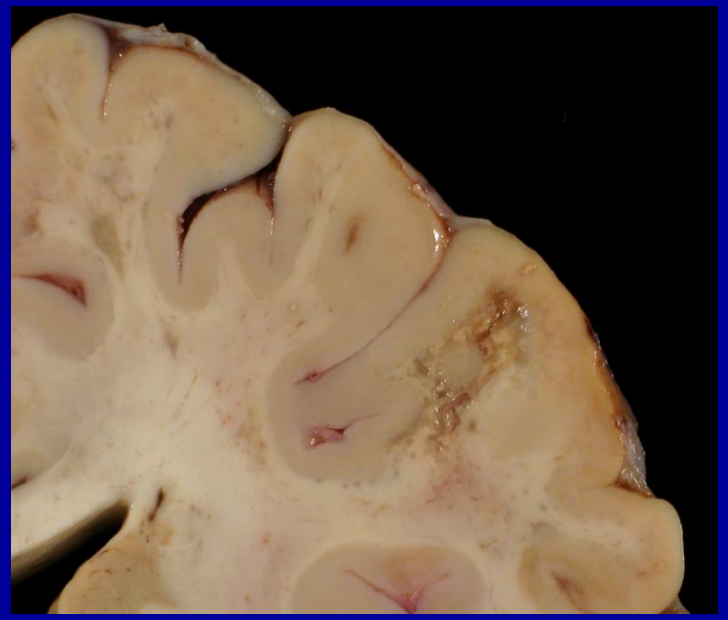
The diagnosis can be established by:

A. Measuring beta$_2$-microglobulin in cerebrospinal fluid
B. Electroencephalography
C. Arteriography
D. Magnetic resonance imaging
E. Measuring CSF polymerase chain reaction for JC virus

Question 11:

The answer is E

Question 11 answer:

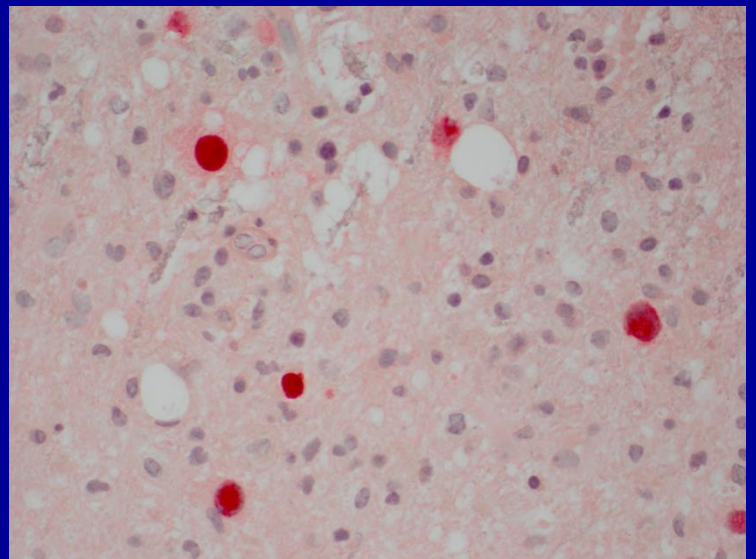


The diagnosis can be established by:

A. Measuring beta$_2$-microglobulin in cerebrospinal fluid
B. Electroencephalography
C. Arteriography
D. Magnetic resonance imaging

E. Measuring CSF polymerase chain reaction for JC virus

A diagnosis of PML can be made via CSF with polymerase chain reaction (PCR) for the JC virus or by brain biopsy. PCR of the CSF has been shown to be highly specific (92-99%) and sensitive (74-93%) for the detection of JC virus in patients with PML. Brain biopsy has a sensitivity of 74-92% and a specificity of 92-100%.



Question 12:

A 60 year-old businessman suddenly becomes confused at a meeting. He has no perceivable motor impairment and recognizes his colleagues.

Question 12 (cont'd):

He continually asks the same questions about the subject matter under consideration at the meeting.

Question 12 (cont'd):

One week later, the patient is normal, but unable to remember the events of the meeting.

Question 12 (cont'd):

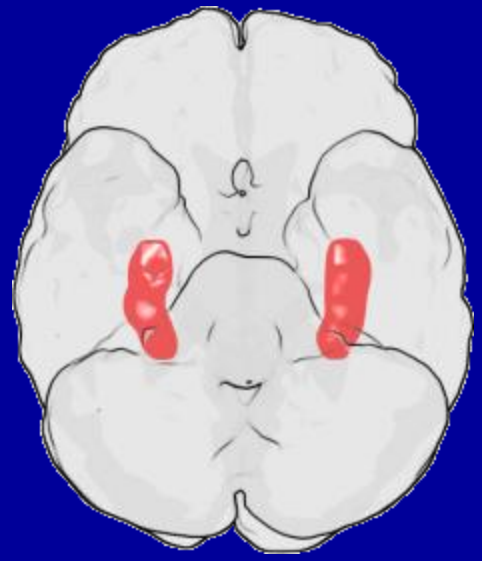
The most likely diagnosis is:

A. Hysterical fugue state
B. Pulmonary embolism
C. Stroke syndrome
D. Transient global amnesia
E. Complex partial seizure

Question 12:

The answer is D

Question 12 answer



The most likely diagnosis is:

A. Hysterical fugue state

B. Pulmonary embolism

C. Stroke syndrome

D. Transient global amnesia

E. Complex partial seizure

TGA is characterized by <24 hours of inability to form new memories (anterograde amnesia), which cannot be attributed to other causes such as stroke or seizure, with normal level of consciousness and otherwise normal cognitive functioning and neurologic examination. It most commonly affects the middle-aged or elderly. The underlying cause of TGA is unclear, but there appears to be reversible dysfunction of the hippocampus possibly due to ischemia or excitotoxicity.

Question 13:

Four years after having lumpectomy and radiation treatment for breast carcinoma, a 45 year-old woman develops pain and weakness of the left leg that spreads over a period of 1 week to involve the right leg.

Question 13 (cont'd):

She also has local back pain in the midthorax and a circumferential band-like sensation.

Question 13 (cont'd):

In the past day, she has become incontinent of urine after brief urgency, and her genitalia are numb.

Question 13 (cont'd):

The patient weighs 160 kg (352 lbs). Reflexes are 3+ with unsustained ankle and knee clonus; toes are extensor and the legs occasionally jerk into a flexed posture.

Question 13 (cont'd):

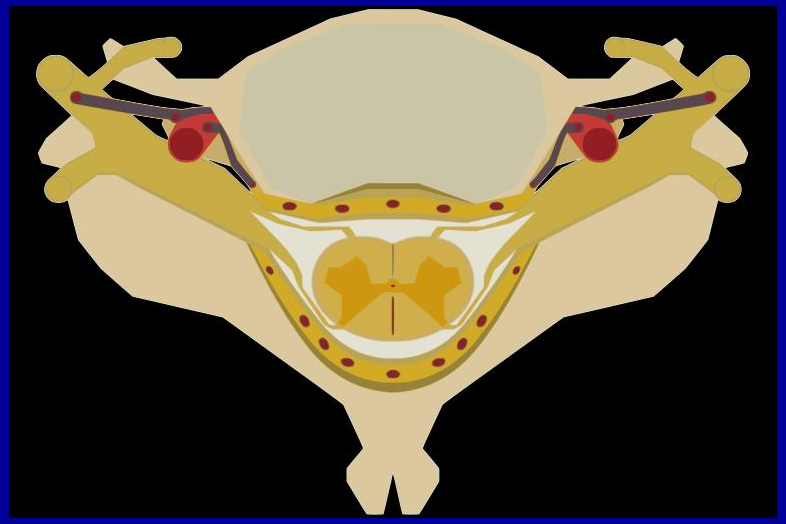
The most likely diagnosis is:

A. Intramedullary metastasis
B. Epidural metastasis
C. Carcinomatous meningitis
D. Metastasis to the sagittal sinus
E. Radiation necrosis of the spinal cord

Question 13:

The answer is B

Question 13 answer



The most likely diagnosis is:

A. Intramedullary metastasis

B. Epidural metastasis

C. Carcinomatous meningitis

D. Metastasis to the sagittal sinus

E. Radiation necrosis of the spinal cord

Cord compression at the thoracic level
In a patient with bilateral leg weakness, it is crucial to check for a sensory level, bowel and bladder dysfunction, and saddle anesthesia. This presentation is a neurologic emergency. The most common metastases leading to cord compression are prostate, breast and lung cancer. Metastasis usually arises in the posterior aspect of vertebral body with later invasion of epidural space. The most common complaint is pain, and two thirds of patients will have motor signs at time of diagnosis of spinal epidural metastasis.

Question 14:

A 25 year-old woman had lost the vision in the right eye for 6 weeks at age 18. The loss was attributed to “nerves” when her father died.

Question 14 (cont'd):

Her visual acuity has been as bad as 20/400, but is now 20/20 with 20/15 acuity in the left eye.

Question 14 (cont'd):

She has been having recurrent 20- to 30-minute episodes of dimming of vision related to exercise and hot showers. She has no other neurologic symptoms.

Question 14 (cont'd):

Which of the following statements about this patient is (are) true?

A. She has a larger pupil on the right
B. She probably has a normal visually evoked potential
C. She has a right afferent pupillary defect
D. She has definite multiple sclerosis
E. She has an optic nerve tumor

Question 14:

The answer is C

Question 14 answer

Which of the following statements about this patient is (are) true?

A. She has a larger pupil on the right

B. She probably has a normal visually evoked potential

C. She has a right afferent pupillary defect

D. She has definite multiple sclerosis

E. She has an optic nerve tumor

She has a right afferent pupillary defect. She had an initial presentation consistent with R optic neuritis, which is associated with ipsilateral afferent pupillary defect. 20-30% of the time optic neuritis is the presenting sign of multiple sclerosis (MS). Up to 50% of patients with MS will develop an episode of optic neuritis.

Uhthoff's phenomenon, episodic transient worsening of symptoms with increased body temperature (due to exertion, hot weather, hot showers), occurs in isolated optic neuritis and in MS.

Question 14 (cont'd):

Six months later she returns after having an episode of right leg weakness and numbness that resolved. MRI of the brain at that time showed an enhancing lesion of the left periventricular white matter. She begins treatment for MS, but develops fever, chills, malaise, muscle aches and fatigue.

Question 14 (cont'd):

Which of the following is the most likely treatment that was initiated?

A. Intravenous methylprednisolone
B. Rituximab
C. Glatiramer
D. Interferon beta
E. Natalizumab

Question 14:

The answer is D

Question 14 answer

Which of the following is the most likely treatment that was initiated?

A. Intravenous methylprednisolone

B. Rituximab – nausea, vomiting, dizziness, h/a, pruritus, asthenia, shivering

C. Glatiramer – post-injection reactions (systemic & local)

D. Interferon beta

E. Natalizumab – depression, nausea/GI, **PML**

Interferon beta frequently causes flu-like side effects and this is the most common side effect requiring intervention. These side effects usually appear early in therapy, last about 1 day after injection and may subside over time. NSAIDs and acethaminophen are used to manage symptoms.

Question 15:

A 60 year-old mechanic fractured his left tibia. Two weeks later he develops a severe, constant ache in his leg.

Question 15 (cont'd):

The leg becomes pale with some cyanotic mottling, feels cold and sweaty, and movement is limited. The toenails are brittle and short.

Question 15 (cont'd):

The most likely diagnosis is:

A. Tarsal tunnel syndrome
B. Occlusion of the tibial artery
C. Psychophysiologic disorder
D. Complex regional pain syndrome
E. Dermatophyte infection

Question 15:

The answer is D

Question 15 answer

The most likely diagnosis is:

A. Tarsal tunnel syndrome

B. Occlusion of the tibial artery

C. Psychophysiologic disorder

D. Complex regional pain syndrome

E. Dermatophyte infection

Reflex symptathetic dystrophy (RSD)/ Complex regional pain syndrome (CRPS). CRPS is a chronic progressive disease characterized by severe pain, swelling and changes in the skin. The cause of this syndrome is currently unknown. Precipitating factors include injury and surgery, although there are documented cases that have no demonstrable injury to the original site. The syndrome is diagnosed by: the presence of an initiating noxious event or a cause of immobilization (type 1) or after nerve injury (type 2), continuing pain, allodynia (perception of pain from a nonpainful stimulus), or hyperalgesia disproportionate to the inciting event, evidence at some time of edema, changes in skin blood flow, or abnormal sudomotor activity in the area of pain.

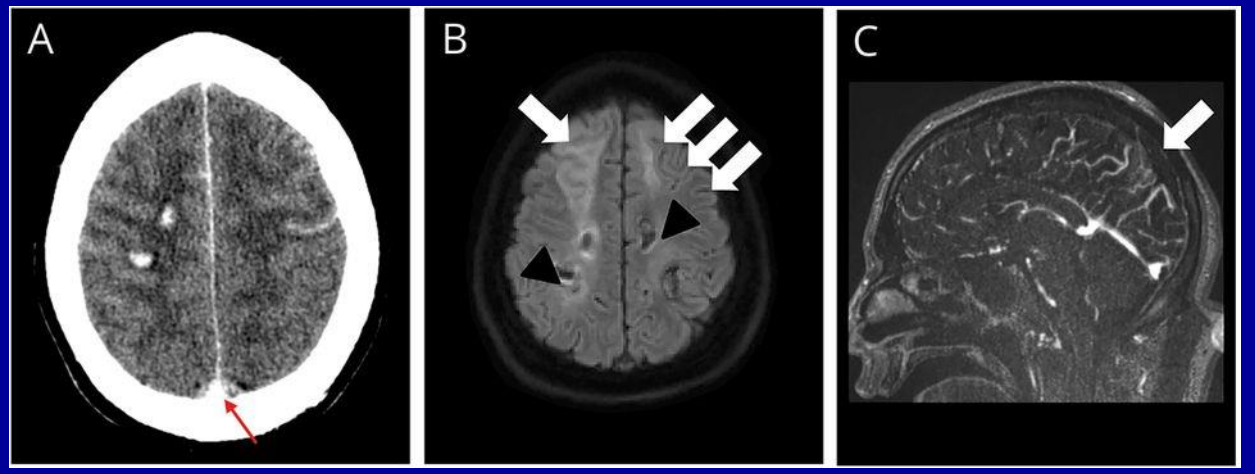
Question 16

A 26-year-old previously healthy woman on CHCs presented 2 weeks after receiving a vaccine with several days of headache and a new left sided weakness

Question 16 continued

Routine blood work demonstrated platelet count 38 × 10^9/L, fibrin D-dimer >20,000 µg/L, international normalized ratio 1.3, and fibrinogen 80 mg/d

Imaging



Ginette Moores et al. Neurol Clin Pract 2021;11:e929-e931

The most likely diagnosis is CVT due to

A. Antiphospholipid antibody syndrome
B. Heparin induced thrombocytopenia
C. Vaccine induced immune thrombotic thrombocytopenia
D. A hypercoagulable state due to CHC

Question 16

The answer is C

Question 16 Answer

HIT is a transient prothrombotic disorder that is triggered by exposure to heparin. “Vaccine induced immune thrombotic thrombocytopenia syndrome” is when a clinical picture identical to HIT (thrombocytopenia, thrombosis, and detectable HIT antibodies) is triggered by a vaccine and is mediated by platelet- activating antibodies to platelet factor 4

Good Luck!