Neurology Board Review Questions

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I have no financial disclosures
1. A 37 year old female came to your office with h/o uncontrolled seizures for past 2 years on maximal tolerated doses of CBZ and valproic acid. She has focal seizures with impairment of awareness usually manifest with a behavioral arrest and staring and last between 30 and 120 seconds. She did have prolonged febrile seizures in early childhood.

Her CNS exam is normal. MRI of brain has shown left hippocampal sclerosis with increased T2 signal within the hippocampus and atrophy of left temporal lobe. Awake EEG is normal. What is the next step in management?

A. Add Keppra  
B. D/C CBZ and add phenytoin  
C. Video EEG for possible  
Anterior temporal lobectomy  
D. Vagus nerve stimulation (VNS)  
E. Ambulatory EEG monitoring
C. Prolonged EEG for possible anterior temporal lobectomy
Epilepsy

- Drug-resistant epilepsy (DRE) (Adding 3rd drug <5% success)

- VNS -12yrs old with DRE, opposed to surgery, or not candidates, or seizures not improved by prior epilepsy surgery (30 to 40% pts >50% reduction in frequency)

- Juvenile myoclonic epilepsy provoked by alcohol or sleep deprivation and can also experience absence and myoclonic seizures that are worse in the morning

- Chronic treatment after 2 unprovoked seizures

- Single GTC seizure with abnormal EEG, MRI or neuro exam, or family history of epilepsy consider AED

- Ethosuximide for absence seizures
Epilepsy

• Broad spectrum: all seizure types
  - Lamotrigine
  - Levetiracetam (Keppra)
  - Topiramate
  - Valproate

• Narrow spectrum: focal with or without alteration in consciousness or awareness and focal evolving to bilateral convulsive seizure
  - Carbamazepine, Oxcarbazepine
  - Phenobarbital
  - Phenytoin
  - Pregabalin
  - Gabapentin
Epilepsy

- AED discontinuation, seizure free for more than two years

- Status Epilepticus: $\geq 5$ minutes of continuous seizures, or $\geq 2$ discrete seizures between which there is incomplete recovery of consciousness,

- RX lorazepam followed by phenytoin or fosphenytoin if not controlled
Epilepsy in Pregnancy

- Intrauterine hormone-releasing or depot inject of progesterone not affected AEDs

- WHO combined hormonal contraception should be avoided with CBZ, phenytoin, barbiturates, primidone, topiramate, or oxcarbazepine

- Folic acid supplementation

- Pt’s planning pregnancy should be managed on the most effective AED possibly monotherapy for their seizures (Except valproate 9%)

- Total and free plasma AED levels measured more frequently

- Vitamin K supplementation (10 to 20 mg/day) during the last month
2. A 42 year old obese female with PMH of COPD, HTN, Hyperlipidemia and migraine came to your office. She is getting migraine attack more than 1 per week which is affecting her quality of life. Her migraine was well controlled 2 years ago. Her vitals are stable. Her CNS exam is normal. She has not changed any of her medications recently. Which of the following is the most appropriate treatment?

A. Sumatriptan
B. Valproate
C. Propranolol
D. Topiramate
E. Amitriptyline
Answer

D. Topiramate
Migraine and Cluster headache

• Acute or abortive treatment: NSAIDS, Triptans, DHE, Antiemetic, Narcotics (Avoid more than 2 times a week)

• Prophylactic: Beta blockers, Tri-cyclics, Ca-blockers, topirimate, gabapentin, valproic acid, SSRI

• Cluster headache: Unilateral always, retro orbital, Predominately male, Awaken from sleep, ETOH induces, associated with Horner's syndrome, lacrimation, rhinorrhea, Last 15 to 180 minutes

• Acute Rx: Oxygen or sumatriptan SQ, If no response nasal lidocaine, DHE

• Prophylaxis: Verapamil, steroids, Lithium, Depakote
3. A 39-year-old woman is having headache for past 5 days. She describes the headache as constant, worse when she first awakens, and characterized by a feeling of increased pressure. Her only medication is a low-dose estrogen oral contraceptive. She smoke 1 PPD for past 20 years.

BP is 122/82 mm Hg, BMI is 33. Bilateral papilledema is noted. The Valsalva maneuver increases the headache pain. Rest of the examination findings are unremarkable. Normal CBC, CMP, PT, PTT and INR. An MRI of the brain without contrast is normal. Which of the following is the most appropriate next diagnostic test?

A. Measurement of serum lupus anticoagulant level  
B. Lumbar puncture  
C. Magnetic resonance venography  
D. Cerebral angiography
Answer

C. Magnetic resonance venography
Cerebral Venous Thrombosis (CVT)

Etiology: 80-90% have provoked CVT

- Prothrombotic conditions
- OCPs
- Pregnancy and the puerperium
- Malignancy
- Infection
- Head injury and mechanical precipitants
Cerebral Venous Thrombosis (CVT)

Clinical features
- Can be acute, subacute, or chronic
- Focal syndrome (focal deficits, seizures, or both)
- Encephalopathy (multifocal signs, mental status changes, stupor, or coma)

Diagnosis
- MRI: The characteristics of the MRI signal depend on the age of the thrombus
- MR Venography
- Head CT findings are nonspecific and may be normal in 30% of pts
American Heart Association/American Stroke Association (AHA/ASA) in 2011

- Imaging of the cerebral venous system should be performed in patients with lobar intracerebral hemorrhage of otherwise unclear origin or with cerebral infarction that crosses typical arterial boundaries.

- Imaging of the cerebral venous system should be performed for pts with the c/f of idiopathic intracranial hypertension, and that imaging of the cerebral venous system is reasonable to exclude CVT in pts with headache associated with atypical features.
Management of CVT

- Warfarin with bridging for 3-12 months

- Hypercoagulable work up after finishing treatment

- Pregnant women, LMWH beginning in the third trimester and continuing up to eight weeks postpartum
4. Pt had MRV which was negative for dural venous thrombosis. LP was done while pt was lying in the lateral decubitus position with legs extended. LP opening pressure of >25 cmH2O and normal biochemical and cytological composition of CSF. Diagnosis of benign ICH (pseudotumor cerebri) was made on the basis of Modified Dandy Criteria. All of the following is TRUE except;

A. Complete Ophthalmologic evaluation with close follow up
B. Low-sodium weight reduction program
C. Acetazolamide or Topiramate with lasix
D. Long term oral corticosteroids with serial LP
E. Abducens nerve palsy can be seen
Answer

D. Long term oral corticosteroids with serial LP
**Modified Dandy Criteria**

1. Symptoms of raised intracranial pressure (headache, nausea, vomiting, transient visual obscurations, or papilledema)
2. No localizing signs with the exception of abducens (sixth) nerve palsy
3. The patient is awake and alert
4. Normal CT/MRI and MRV findings without evidence of thrombosis
5. LP opening pressure of >25 cmH2O and normal biochemical and cytological composition of CSF
6. No other explanation for the raised intracranial pressure
Management of IIH (Pseudotumor Cerebri)

• Serial examinations including visual acuity, formal visual field testing and a dilated fundus examination with optic disc photographs

• Potential agents (eg, tetracycline, sterodis, vitamin A) should be D/C

• Counseling and/or treatment for weight loss in all obese patients, Low salt

• Acetazolamide, Topiramate and other carbonic anhydrase inhibitors with lasix

• Surgical intervention with optic nerve sheath fenestration (ONSF) and/or a CSF shunting procedure
5. A 73-year-old woman is evaluated in the ED for a 6-week history of a progressively worsening and constant daily global headache. She also reports fatigue and malaise but has had no fevers, nausea, photophobia, visual changes, focal weakness or numbness, or history of headaches.

O/E Scalp tenderness in the bilateral temporal-parietal areas is noted, but no papilledema, nuchal rigidity, or focal neurologic findings are found. ESR 102 mm/h, CRP 3.0 mg/L.

Normal CBC and CMP. Pt was started on prednisone therapy for giant cell arteritis. Pt responded to prednisone treatment. She was scheduled for temporal artery biopsy.
Which of the following is FALSE regarding temporal artery biopsy?

A. Temporal artery biopsy is the gold standard for the diagnosis of GCA
B. An accurate diagnosis can be made even after several weeks or months after the start of prednisone therapy with biopsy
C. For those who have undergone unilateral biopsy, a contralateral biopsy should be performed if the first biopsy is negative
D. Scheduling of temporal artery biopsy should be done before starting treatment with prednisone
E. Negative temporal artery biopsy occurs in up to 35-40 percent of patients
D. Scheduling of temporal artery biopsy should be done before starting treatment with prednisone
Giant cell (temporal) arteritis

- Age greater than or equal to 50 years at diagnosis, aging greatest risk

- Chronic vasculitis of large- and medium-sized vessels

- ESR greater than 50 mm/hour, often reaches more than 100 mm/hour

- Biopsy revealing a necrotizing arteritis with a predominance of mononuclear cells or a granulomatous process with multinucleated giant cells

- PMR occurs in about 40 to 50 percent, with decrease ROM and normal muscle strength, lack of response to steroid within 1 week question diagnosis
Diagnosis of Giant cell (temporal) arteritis

- Temporal artery biopsy should be performed in all patients suspected of GCA

- Negative temporal artery biopsy occurs in up to 40 percent of patients

- Negative biopsy may occur because of skip lesion, small sample (<2cm), or may be phenotypic variation

- Patients who manifest only symptoms of PMR do not require biopsy

- MR angiogram is required in these pts if index of suspicion is very high

- If an empiric trial of high dose of glucocorticoid fail to resolve pt’s symptoms within one week this argues strongly against the diagnosis of GCA
Polymyalgia rheumatica (PMR)

• Bilateral aching & morning stiffness for at least two weeks, and involving at least two of the following three areas: neck, shoulders, and hips

• PMR 4 times more common than GCA, PMR is the second most common autoimmune rheumatic disorder after RA

• Up to 20% of untreated PMR patients may develop full-blown GCA

• Prompt response of symptoms to low-dose glucocorticoids in PMR

• Aspirin for primary prevention if no C/I
6. A 45 year old man developed progressive difficulty walking and urinary urgency over 2 weeks. On exam cranial nerve exam is normal. He has moderate weakness in deltoid, triceps, wrist extensors, hip abductors, hamstrings, and anterior tibialis muscles. There is mild atrophy of the deltoid and biceps muscles bilaterally. Biceps tendon reflexes absent, but triceps, knee, and ankle reflexes are exaggerated with bilateral Babinski's reflexes. There is absent joint position and vibration sense in the fingers and toes. What is the most next step in diagnosis?

A. MRI of brain  
B. MRI of C-Spine  
C. Needle EMG  
D. LP to rule out GBS
Answer

B. MRI of C-Spine
Radiculopathy, Spinal stenosis

- Compressive Cervical radiculopathy: Cervical spondylosis and disc herniation
  - LMN at the level of lesion and UMN below the level of lesion
  - MRI of C-Spine, CT myelography

- Lumbar radiculopathy most commonly involves either the L5 or S1 root
  - May have normal neuro exam
  - Symptoms get worse with standing up or walking, relieve with standing or leaning forward
  - MRI of L-Spine, CT myelography
7. A 51-year-old woman is evaluated in the ED for a 2-day history of low back pain, generalized weakness, gait imbalance, and dizziness on standing. She claims that ambulation has severely affected. Three weeks ago, she had viral gastroenteritis.

Muscle strength testing reveals diffuse weakness, especially of the distal lower limbs (ankle dorsiflexor strength, 4/5). Stretch reflexes are absent. Sensory examination findings are normal. Normal CBC, CMP, UA and CXR. What of the following would be expected on further evaluation?

A. MRI brain showing T-2 enhancing subcortical lesions
B. Abnormal EEG
C. CSF showing increase protein with normal Cell count
D. Ophthalmologic exam with disc palor
E. Gadolinium enhancement on MRI of T-Spine
Answer

C. CSF showing increase protein with normal Cell count
Guillain-Barré syndrome - Acute inflammatory demyelinating polyneuropathy (AIDP)

- Ascending, symmetric, paralysis with areflexia, no sensory involvement

- Progression to nadir between few days to 4 weeks

- Albuminocytologic dissociation with <10 cells and increased protein

- Rx not recommended for ambulatory patients who are mildly affected, already recovering and present after 2-4 weeks of the presentation

- For non ambulatory pts, who are within four weeks, Rx with plasma exchange or IVIG is recommended, Steroids C/I

- No vaccination within 1 year, after that individualized
Indications for intubation-GBS

Consider intubation for 20, 30, 40 rule:

- Forced vital capacity <20 mL/kg
- Maximum inspiratory pressure <30 cmH2O
- Maximum expiratory pressure <40 cmH2O
GBS diagnosis doubtful

- Sensory level (decrement or loss of sensation below a spinal cord root level as determined by neurologic examination)
- Marked, persistent asymmetry of weakness
- Severe and persistent bowel and bladder dysfunction
- More than 50 white cells in the CSF
Chronic inflammatory demyelinating polyneuropathy (CIDP)

- The classic form is fairly symmetric and motor involvement is greater, hallmark pattern is presence of weakness in both proximal and distal muscles (>8 weeks)

- Sensory involvement distal to proximal, affect large myelinated fibers

- Most pts also globally diminished or absent reflexes

- Cranial nerve and bulbar involvement occur in 10 to 20 percent of patients
Chronic inflammatory demyelinating polyneuropathy (CIDP)

- The classic albuminocytologic dissociation in CSF, MRI enlarged nerves
- NCV study might show reduced Conduction velocity, prolonged distal motor latencies, conduction block
- Nerve biopsy evidence of segmental demyelination with or without inflammation
- Intravenous immune globulin (IVIG), glucocorticoids, or plasma exchange
A 21-year-old woman presented because of a 1-week history of bilateral leg weakness and numbness accompanied by urinary incontinence that began after a viral gastrointestinal illness. Bilateral leg weakness, loss of sensation below the umbilicus, and hyperreflexia in the lower extremities are noted.

A T2-weighted MRI of the T-spine reveals hyperintensity at the T9 level, which enhances with administration of gadolinium. CSF showed glucose 57, protein 45, WBC 50, Cultures for CSF bacteria are negative & PCR for herpes negative. RPR, ANA, HIV antibodies & Lyme titers are negative. What would you do next?

A. Glatiramer acetate
B. IV Methyprednisolone for 3-5 days
C. Methotrexate
D. Plasmapheresis
E. Cyclophosphamide
Answer

B. IV Methyprednisolone for 3-5 days
Transverse Myelitis

- Sensory, motor or autonomic dysfunction attributable to the spinal cord, Bilateral signs and/or symptoms, Clearly defined sensory level

- Post infectious (40-60%), CNS autoimmune disorders (MS, NMO, ADEM), Neurosarcoidosis, Paraneoplastic syndromes, CNS infections, autoimmune

- Serum NMO-IgG antibodies, B12, TSH, methylmalonic acid, HIV antibodies, RPR, ANA, Ro/SSA, and La/SSB antibodies

- Suspected acute Myelopathy → MRI of the entire spine → LP to evaluate for (IgG index) cord inflammation → Define extent and site of inflammation

- Progression to nadir between four hours and 21 days, monophasic, symptoms beyond 30 days may be MS
9. A 34-year-old woman is evaluated for a 3-day history of partial vision loss in the left eye and pain on movement of that eye. She has a history of migraine. She denies any other neurological symptom. Visual acuity is 20/200 in the left eye and 20/20 in the right eye. A left afferent pupillary defect is noted. Funduscopic examination shows no papilledema. Normal CNS exam otherwise.

An MRI of the left orbit shows contrast enhancement of the left optic nerve. An MRI of the brain shows three white matter lesions, each measuring approximately 3 mm; two of these lesions are periventricular. What would you tell her?

A. She does NOT need IV steroid for acute Rx
B. She meets full Macdonald’s criteria for MS
C. She has Relapsing-remitting MS (RRMS)
D. She has Secondary progressive MS (SPMS)
E. Beta IFNs, glatiramer acetate, & IV immune globulin can delay conversion to clinically definite MS
E. Beta IFNs, glatiramer acetate, & IV immune globulin can delay conversion to clinically definite MS in patient with Clinically isolated syndromes (CIS)
<table>
<thead>
<tr>
<th>Mc Donald criteria for MS</th>
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<tr>
<td><strong>Dissemination in space</strong></td>
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<tr>
<td>1 T2 lesion or more in at least two MS typical CNS regions:</td>
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<tr>
<td>• Periventricular</td>
</tr>
<tr>
<td>• Juxta cortical</td>
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<tr>
<td>• Infratentorial</td>
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<tr>
<td>• Spinal cord</td>
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Multiple Sclerosis (MS)

- Clinically isolated syndromes (CIS), Radiologically isolated syndrome (RIS), Relapsing-remitting MS, Secondary progressive MS, Primary Progressive MS

- Early disease-modifying Rx in pts with CIS, with clinically silent lesions on MRI reduces the risk of MS development, but do not prevents long-term disability??

- No indication for the use of disease-modifying treatment for patients with RIS

- Rarely cause seizure, pain, aphasia (gray matter involvement?)
CLASSIFICATION OF MULTIPLE SCLEROSIS

**PRMS** Progressive Relapsing MS
Steady decline since onset with super-imposed attacks.

**SPMS** Secondary Progressive MS
Initial RRMS that suddenly begins to decline without periods of remission and relapses.

**PPMS** Primary Progressive MS
Gradual progression of the disease from its onset with no relapses or remissions in 50% of the cases. More common in older patients above 40.

**RRMS** Relapsing/Remitting MS
Unpredictable attacks which may or may not leave permanent deficits followed by periods of remission accounts for 85% of total cases.
Multiple Sclerosis (MS)

- The Optic Neuritis Treatment Trial: Combination drugs speed recovery, White matter lesions predict MS, Vision recovers regardless of the Rx

- Immunomodulatory agents, including Interferon beta, glatiramer acetate, Dimethyl fumarate, Natalizumab, in pts with RRMS:
  - A decreased relapse rate
  - A slower accumulation of brain lesions on MRI

- SPMS and PPMS usually required IV glucocorticoid monthly pulses OR Methotrexate, oral or SQ, 7.5 to 20 mg per week, with or without monthly glucocorticoid pulses

- UMN spasticity and hyperreflexia- Tizanidine, baclofen and cyclobenzaprine & Amantadine and modafinil for fatigue
10. A 52-year-old man is evaluated for a 3-day history of episodic dizziness for 35-40 seconds with nausea. He noted the onset abruptly and compares the feeling to “being on a roller coaster.” His most severe episodes occurred while arising from bed and when parallel parking his car. He has no recent fever, headache, tinnitus, hearing loss, double vision, dysarthria, weakness, or difficulty walking. He had a similar episode 4 years ago.

O/E, vital signs are normal. Results of cardiac and neurologic examinations are normal. The Dix-Hallpike maneuver precipitates severe horizontal nystagmus after about 20 seconds. With repeated maneuvers, the nystagmus is less severe

A. Benign paroxysmal positional vertigo  
B. Vestibular neuronitis  
C. Meniere disease  
D. Cerebellar infarction
Answer

A. Benign paroxysmal positional vertigo
Benign Paroxysmal Positional Vertigo (BPPV)

- BPPV is most commonly attributed to calcium debris within the posterior semicircular canal

- Nystagmus 30-45 seconds latency, fatigable, last for less than 1 minute

- Unidirectional, fast component toward the normal ear; never reverses direction, Horizontal with a torsional component, never purely torsional or vertical

- Vestibular rehabilitation exercises of Epley’s maneuverer fails

- Medications like Meclizine usually have no role
### Clinical features of central versus peripheral vertigo

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<tr>
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<th>Peripheral</th>
<th>Central</th>
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<tr>
<td><strong>Nystagmus</strong></td>
<td></td>
<td></td>
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<tr>
<td>Direction</td>
<td>Unidirectional, fast component toward the normal ear; never reverses direction</td>
<td>Sometimes reverses direction when patient looks in the direction of slow component</td>
</tr>
<tr>
<td>Type</td>
<td>Horizontal with a torsional component, never purely torsional or vertical</td>
<td>Can be any direction</td>
</tr>
<tr>
<td>Effect of visual fixation</td>
<td>Suppressed</td>
<td>Not suppressed</td>
</tr>
<tr>
<td><strong>Other neurologic signs</strong></td>
<td>Absent</td>
<td>Often present</td>
</tr>
<tr>
<td><strong>Postural instability</strong></td>
<td>Unidirectional instability, walking preserved</td>
<td>Severe instability, patient often falls when walking</td>
</tr>
<tr>
<td><strong>Deafness or tinnitus</strong></td>
<td>May be present</td>
<td>Absent</td>
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11. A 58-year-old man is complaining of imbalance when he walks. He also has an intermittent tremor of the right hand, when is not engaged in purposeful activities. His tremor gets worse when he perform mental calculations or voluntary repetitive movements of the left arm. He has slowed down a lot according to his wife. She complain that pt is having difficulty performing simple tasks, such as buttoning clothes, tying shoelaces, typing, or lifting coins from a pocket or purse. Pt is also having symptoms of mood changes, difficulty with sleep and fatigue. As per his wife his memory is intact.

O/E, vital signs are normal. AAO X 3, MMSE 26. He is bradykinetic with a right hand rest tremor. Tone is increased in the right arm and right leg. His walking is slow and shuffled and he falls backward when pulled from a neutral position.

Normal CBC, CMP, TSH, Vit b12, A1C and lipid profile. Normal CXR and EKG.
What is the next step in management?

A. Perform autonomic reflex testing
B. A treatment trial with cholinesterase inhibitors to ameliorate cognitive and behavioral symptoms as pt is having Dementia with Lewy bodies (DLB)
C. Perform MRI of brain to rule out Binswanger disease
D. Avoid carbidopa-levodopa because of concerns of toxicity in early Parkinson disease
E. Prescribe a gradually increasing dose of carbidopa-levodopa
Answer

E. Prescribe a gradually increasing dose of carbidopa-levodopa
**Parkinson Disease**

- The cardinal features of PD are tremor, bradykinesia, rigidity and gait disturbance

- Bradykinesia, plus one of the other two cardinal manifestations (tremor, rigidity) must be present in order to make the diagnosis of idiopathic PD

- C/F that are supportive of the diagnosis are unilateral onset, presence of a rest tremor, and a persistent asymmetry throughout the course of the disease

- Acute dopaminergic challenge test VS long-term trial of dopaminergic or levodopa therapy

- Significant therapeutic response to moderate doses of levodopa (400 to 600 mg daily)

- Absence of response to a dose of 1000 to 1500 mg/day for at least two months strongly suggests that the original diagnosis of PD was incorrect

- The true "gold standard" for diagnosis is neuropathologic examination
Parkinson Disease

• Dopamine agonist (DAs) in younger patients (age <65 years), and with levodopa in older patients (age >65 years)

• DAs as adjunctive treatment for advanced PD complicated by reduced levodopa response, motor fluctuations, dyskinesia, and other adverse effects of levodopa

• Selegiline, a selective MAO) type B inhibitor, is modestly effective as symptomatic treatment in early PD and may have neuroprotective properties

• Anticholinergic for younger patients in whom tremor is the predominant problem

• COMT inhibitors as levodopa extenders, reduces the peripheral (entacapone) and central (tolcapone) methylation of levodopa and dopamine
12. A 74-year-old man is evaluated for a 3-month history of hallucinations. He describes frequently seeing small imaginary animals resembling badgers in the house, a situation he finds annoying rather than threatening. Additionally, the patient often has disturbed sleep, but he has no recollection of nightmares the next morning. Wife often describe episodes in which pt appear to "blank out" or lose consciousness, become confused or behave in a bizarre manner, have speech or motor arrest, or become excessively somnolent. These episodes can last seconds to days, and they can be interspersed with periods of near-normal function.

He has a history of parkinsonism characterized by slowness, a stooped posture, and a tendency to shuffle when he is tired. Not on any medications. Results of physical examination are unremarkable. MMSE showed impaired figure copying (overlapping pentagons), clock drawing, and serial sevens (or spelling WORLD backward). His MMSE score is 20 with preserved short memory.
What is the most likely diagnosis?

A. Alzheimer disease  
B. Dementia with Lewy bodies  
C. Frontotemporal dementia  
D. Neurosyphilis
Answer

B. Dementia with Lewy bodies
Dementia with Lewy bodies (DLB)

- Progressive dementia (100%), deficits in attention and visuospatial function; fluctuating cognition; recurrent visual hallucinations; and spontaneous motor features of parkinsonism

- Repeated falls, syncope, autonomic dysfunction, neuroleptic sensitivity, delusions, hallucinations in other modalities, sleep disorders, and depression

- Visual Hallucinations DLB vs AD, Parkinsonism features DLB vs PD, REM Sleep disorders DLB vs PD

- SPECT and PET scans show generalized decreased perfusion and metabolism, respectively, which is most noticeable in the occipital areas

- Cholinesterase inhibitors may represent a first line pharmacologic Rx, Atypical neuroleptic agent for disabling psychotic symptoms
13. Pt was diagnosed with DLB. Non pharmacologic therapies and behavior intervention showed minimal benefit for his neuropsychiatric symptoms. Wife has requested pharmacologic therapy. Which of the following is FALSE regarding pharmacologic therapies in pts with DLB?

A. Atypical neuroleptics are the agents of choice for treating hallucinations
B. Cholinesterase inhibitors for mild to moderate dementia
C. REM Sleep disorders respond to melatonin or clonazepam
D. IV Haloperidol is FDA approved to control agitation
D. IV Haloperidol is FDA approved to control agitation
Front temporal dementia

• Patients present with progressive change in behavior and personality within 1-2 yrs

• Pts seem unaware or incompletely aware of their deficits at presentation

• Loss of social awareness, voiding urine or feces in inappropriate places without apparent concern, may commit antisocial activities, diminished libido

• Stereotyped or ritual behaviors, hoarding, Change in eating patterns, Emotional blunting and loss of empathy

• Progressive aphasia includes word-finding difficulty and impaired object naming, dysfluent speech, speech errors

• SSRI, SARI for neurobehavioral symptoms, atypical antipsychotic agents last resort
14. A 39 year old male with PMH of HTN came to ER with right sided weakness. His EKG showed NSR and 48 hrs telemetry showed no abnormal rhythm. After having carotid doppler, MRA, Echo and basic blood work up, etiology of stroke is not clear. He was taking aspirin and Lisinopril. What is next best step in management of this patient?

A. Add plavix or Aggrenox to his current regimen  
B. His CHADS2 is 3 now start him on OAC  
C. D/C lisinopril and start him metoprolol  
D. Prolonged continuous cardiac monitoring  
E. Do hypercoauguble work up  
F. Start him on Low intensity statin
Answer

E. Do hypercoagulable work up
Stroke

• Tissue based definition of TIA/Stroke

• Prolonged Telemetry Cryptogenic stroke (30%) in older patients, paroxysmal PAF

• Aspirin, clopidogrel, and aspirin-extended-release dipyridamole (Aggrenox)

• TEE- ascending aortic atheromatous disease, PFO, emboli 3 mm or less

• SBP>220 mmHg or DBP >120 mmHg, (ACS, CHF, dissection, hypertensive encephalopathy, ARF, or pre-eclampsia/eclampsia)

• Persistent disabling neurologic deficit, symptoms <4.5 hours, Age ≥18 ys, consider IV alteplase (persistent SBP ≥185 or DBP≥110 mmHg C/I)
15. A 58-year-old woman is evaluated in the ER for sudden onset of a severe generalized headache that began 36 hours ago and has not responded to over-the-counter medications. The patient has a history of hypertension and 30 pack year of smoking. Do not take any medications.

Normal Vital. Nuchal rigidity is noted. Normal CNS exam. Normal CBC, CMP, and INR. CT scan of the head without contrast is normal. Which of the following is the most appropriate next diagnostic test?

A. CT of the head with contrast  
B. Lumbar puncture  
C. MRA of the head and neck  
D. MRI of the brain without contrast  
E. Carotid Doppler
Answer

B. Lumbar puncture
SAH

• Sensitivity of CT for detecting SAH is highest in the first 6 to 12 hours

• Elevated opening pressure and an elevated RBC that does not diminish tube one to tube four, Xanthochromia

• Digital subtraction angiography, CT and MR angiography to find out the etiology

• Repeat the angiogram in 4 to 14 days if the initial angiogram is negative

• First 48 hours after hemorrhage, aneurysm rerupture and hydrocephalus

• Starting at day 5, the risk of cerebral arterial vasospasm emerges

• PO Nimodipine prevents vasospasm
16. A 65-year-old female is evaluated for an episode of left-handed weakness involving all five digits that occurred 9 months ago and gradually subsided over 3 hours. She started taking aspirin after this episode. On exam, bp is 156/78 mm Hg and HR is 76/min and regular. Cardiac examination reveals a right carotid bruit. Normal exam. LDL 156 mg/dL. She did not have insurance 9 months ago and just got Medicare.

EKG - NSR. Carotid duplex ultrasound: 50% to 69% stenosis of Rt, ICA, which is confirmed by CT angiography. MRI of the brain: 5-mm old infarct in the right MCA distribution. What is the next step in management?

A. Right Carotid Artery Stenting
B. Right Carotid endarterectomy (CEA)
C. Stop aspirin and start plavix or Aggrenox
D. Lipitor 80mg and ACE inhibitor with No CEA
Answer

D. Lipitor 80mg and ACE inhibitor with No CEA
Carotid Artery Stenosis-Asymptomatic/Symptomatic

- Symptomatic (within 6 months) transient or permanent focal neurologic symptoms related to the affected artery (eg, ipsilateral amaurosis fugax, contralateral weakness or numbness of an extremity or the face, dysarthria or aphasia)

- CEA rather than medical Rx alone for stable men with asymptomatic stenosis of 70 to 99 percent who have a life expectancy of at least 5 years

- For men with recently symptomatic carotid stenosis of 50 to 69 percent who have a life expectancy of at least 5 ys, CEA rather than medical management (2A)
Carotid Artery Stenosis-Asymptomatic/Symptomatic

- For women with recently symptomatic carotid stenosis of 50 to 69 percent, medical management rather than CEA (2B)

- No benefit if Near occlusion (100%) of the symptomatic ipsilateral carotid artery & Coexisting severe contralateral stenosis increase perioperative risk but does not cancel out benefit from CEA

- No benefit if Near occlusion (100%) of the symptomatic ipsilateral carotid artery & Coexisting severe contralateral stenosis increase perioperative risk but does not cancel out benefit from CEA
17. A 56-year-old man with 6-month history of generalized fatigue, slowly progressive weakness in the legs, difficulty climbing stairs and raising his arms above his head, and a 2-month history of dry mouth and impotence. He reports that on two occasions in the past month he almost lost consciousness when rising quickly from a sitting position. 38-pack-year smoking history.

Muscle strength testing reveals moderate weakness in the arms and proximal legs, especially when the patient rises from a low chair. Repeated testing slightly improves strength. Stretch reflexes are diffusely reduced or absent but increase to normal after brief exercise. Orthostatic vitals positive. Chest X ray showed lymphadenopathy. What would you do next?

A. Acetylcholine receptor antibody assay
B. Lumbar puncture
C. Muscle biopsy
D. Nerve conduction studies
Answer

D. Nerve conduction studies
# Neuromuscular Disorders

<table>
<thead>
<tr>
<th>Myasthenia Gravis</th>
<th>Eaton-Lambert Syndrome</th>
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<tr>
<td>Antibodies to post-synaptic acetylcholine receptors</td>
<td>Pathophysiology</td>
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<td>Ptosis, Diplopia</td>
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<td>Association</td>
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<td>Normal</td>
<td>Reflexes</td>
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<tr>
<td>Symptoms worsen</td>
<td>Repetitive movements</td>
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</table>

*Lambert enjoys EATING OATs. EATING will make one stronger with repetition*
Management

- The aggressive search for and treatment of a primary underlying malignancy

- Guanidine, Aminopyridines, Acetylcholinesterase inhibitors

- IVIG, Immunosuppressive agents, Glucocorticoid tapering, Plasma exchange

- MG-Acetylcholine receptor antibodies, MuSK antibodies (in AChR Ab negative patients), Repetitive nerve stimulation, Single fiber electromyography

- MG-anticholinesterase agents, chronic immunotherapies (glucocorticoids and immunosuppressive), immunotherapies (plasma exchange and IVIG)

- Thymectomy is indicated in all patients with thymoma, regardless of the status of MG (eg, generalized, bulbar, ocular)
THANKS

ANY QUESTIONS
References


References


References


12. Unpublished data, International study on cerebral vein and dural sinus thrombosis (ISCVT) and Cerebral venous thrombosis Portuguese collaborative study group (VENOPORT) registries.


References


References