

## Movement Disorders: Review Questions

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### QUESTIONS

Choose the single best answer for each question.

Questions 1 to 3 refer to the following case.

A 56-year-old man with a history of hypertension presents to the clinic with complaints of progressive hand tremor. The patient is left-handed. He first noticed the tremor 9 months ago, mostly when he is at rest. On examination, the patient has resting tremor with rigidity that is worse on the left side and decreased facial expressions. Rapid alternating movements are slow on the left side. Sporadic Parkinson's disease is suspected.

- All of the following may help to identify this patient's diagnosis EXCEPT**
  - HIV history
  - Laboratory tests, including complete blood count
  - Manganese exposure
  - Medication history
  - Trauma history
- All of the following are cardinal features of Parkinson's disease EXCEPT**
  - Bradykinesia
  - Dementia
  - Postural instability
  - Rigidity
  - Tremor
- All of the following are appropriate treatment options for this patient EXCEPT**
  - Amantadine
  - Carbidopa-levodopa
  - Duloxetine
  - Pramipexole
  - Selegiline
- A 72-year-old man presents to the emergency department after a fall. He states that he has fallen frequently over the past 8 months. On examination, he has no tremor, but he has generalized rigidity (mostly axial), bradykinesia, increased gag reflex, and difficulty with vertical gaze. What is this patient's most likely diagnosis?**
  - Corticobasal degeneration
  - Multiple system atrophy
  - Parkinson's disease
  - Parkinsonism-dementia-amyotrophic lateral sclerosis
  - Progressive supranuclear palsy
- Alien limb phenomenon is typically seen in which of the following disorders?**
  - Corticobasal degeneration
  - Essential tremor
  - Huntington's disease
  - Multiple system atrophy
  - Parkinson's disease
- All of the following statements regarding essential tremor are correct EXCEPT**
  - Essential tremor is the most common adult-onset movement disorder
  - Essential tremor is secondary to decreased dopaminergic neurotransmission in the basal ganglia
  - Symptoms may improve with alcohol intake
  - Symptoms respond to propranolol and primidone
  - Tremor is typically postural and kinetic
- Which of the following movement disorders persists during sleep?**
  - Dementia with Lewy bodies
  - Essential tremor
  - Huntington's disease
  - Palatal tremor
  - Parkinson's disease

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8. A 37-year-old man presents to the clinic with clumsiness and infrequent jerking of all limbs. He has a recent history of admission to the psychiatry unit for paranoia. Family history is significant for dementia at young age and suicide. Magnetic resonance imaging (MRI) reveals atrophy of the caudate nucleus. What is this patient's most likely diagnosis?
- (A) Brief psychotic episode
  - (B) Essential tremor
  - (C) Huntington's disease
  - (D) Paranoid schizophrenia
  - (E) Tardive dyskinesia

Questions 9 and 10 refer to the following case.

A 72-year-old woman is admitted to the psychiatry unit with agitation and visual hallucinations. The patient's symptoms started 1 year ago, and 6 months later, the patient started to have fluctuating cognitive impairments. Physical examination reveals tremor and rigidity.

9. Which of the following is this patient's most likely diagnosis?
- (A) Alzheimer's disease
  - (B) Corticobasal degeneration
  - (C) Dementia with Lewy bodies
  - (D) Multi-infarct dementia
  - (E) Progressive supranuclear palsy
10. Which of the following medications is the most appropriate for the long-term management of the patient's visual hallucinations and agitation?
- (A) Amitriptyline
  - (B) Chlorpromazine
  - (C) Haloperidol
  - (D) Quetiapine
  - (E) Thioridazine
11. Wilson's disease is most accurately diagnosed with which of the following?
- (A) Biopsy of the liver
  - (B) Computed tomography
  - (C) MRI
  - (D) Serum ceruloplasmin
  - (E) Urinary copper excretion

ANSWERS AND EXPLANATIONS

1. (B) **Laboratory tests, including complete blood count.** There are currently no blood or laboratory tests that have been proven to help in diagnosing sporadic Parkinson's disease. The diagnosis is

based on a thorough medical history and neurologic examination. Patients exposed to toxins, such as manganese, MPTP (1-methyl-4-phenyl-1,2,3,6-tetrahydropyridine), or carbon monoxide; patients with a history of repetitive head trauma or infections, such as HIV or Creutzfeldt-Jakob disease; or patients taking neuroleptic or antiemetic medications may display signs and symptoms of Parkinson's disease.

2. (B) **Dementia.** Parkinson's disease is an idiopathic, slowly progressive, degenerative central nervous system disorder that usually starts on one side of the body and later affects the contralateral side. The 4 primary symptoms of Parkinson's disease are tremor at rest at a frequency of 4 to 5 Hz, rigidity, bradykinesia, and postural instability.
3. (C) **Duloxetine.** Carbidopa-levodopa, pramipexole, ropinirole, amantadine, selegiline, and trihexyphenidyl are used to treat symptoms of Parkinson's disease. Duloxetine is used for the treatment of depression and neuropathic pain. Although depression may be associated with Parkinson's disease, it is not necessary in this patient, as there is no mention of depression in the history.
4. (E) **Progressive supranuclear palsy.** Progressive supranuclear palsy is a neurodegenerative disease that presents with primarily vertical gaze dysfunction accompanied by extrapyramidal symptoms and cognitive dysfunction. The disease usually develops after the fourth decade of life, and the diagnosis is purely clinical.
5. (A) **Corticobasal degeneration.** Alien limb phenomenon is seen in corticobasal degeneration, a Parkinson plus syndrome. It presents with rigidity-dystonia affecting the involved arm that is typically unilateral along with cortical signs of apraxia, alien limb phenomenon, cortical sensory loss, and cortical reflex myoclonus.
6. (B) **Essential tremor is secondary to decreased dopaminergic neurotransmission in the basal ganglia.** Essential tremor is the most common adult-onset movement disorder and is characterized by an 8 to 12 Hz postural and kinetic tremor of the arms that may involve the head, voice, and legs. The pathophysiology of essential tremor is unknown. No pathologic findings are known to be consistently associated with essential tremor. Treatment of

essential tremor includes primidone and  $\beta$ -blockers, such as propranolol. Parkinson's disease is caused by decreased dopaminergic neurotransmission in the basal ganglia.

7. **(D) Palatal tremor.** Palatal tremor is characterized by rhythmic involuntary jerky movements of the soft palate of the throat and can be classified as ordinary (caused by a lesion in the triangle of Guillain Mollaret) or essential (with no identifiable lesion). Palatal tremor is one of the few movement disorders that persists during sleep. Improvement of palatal tremor is reported after administration of flunarizine and injection of botulinum toxin A into the tensor muscles of veli palatini.
8. **(C) Huntington's disease.** Huntington's disease is a degenerative, progressive, hereditary (autosomal dominant) disorder that is characterized by a movement disorder (usually chorea), dementia, and a personality disorder. The Huntington's disease gene is located on the short arm of chromosome 4 (4p16.3) and contains extra copies of trinucleotide repeats of cytosine-adenine-guanine. Imaging with MRI or computed tomography will show dilatation of the frontal horns of the lateral ventricles due to caudate nucleus atrophy. Treatment is based on the patient's symptoms. For treating depression and psychosis related to Huntington's disease, antidepressants and antipsychotic agents can be used, respectively. The choreiform movements can be controlled by typical neuroleptic agents, such as haloperidol and perphenazine, or presynaptic dopamine depleters, such as reserpine and tetrabenazine.
9. **(C) Dementia with Lewy bodies.** This patient has

dementia with Lewy bodies, which is a Parkinson plus syndrome. The central feature of dementia with Lewy bodies is progressive cognitive decline in addition to 3 defining features: pronounced "fluctuations" in alertness and attention, recurrent visual hallucinations, and parkinsonian motor symptoms.

10. **(D) Quetiapine.** Typical antipsychotics, such as haloperidol, and newer agents with dopamine D2 receptor affinity are avoided in the long-term treatment of visual hallucinations and agitation because of potential worsening of motor symptoms, cognitive decline, delirium, and features of neuroleptic malignant syndrome associated with dopamine receptor blockage.<sup>1</sup> Amitriptyline is not indicated for psychosis.
11. **(A) Biopsy of the liver.** Wilson's disease is a disease of copper metabolism that is associated with cirrhosis of the liver and degenerative changes in the basal ganglia. Decreased serum copper and ceruloplasmin and increased urinary copper can help point to a diagnosis, but the disease is most accurately diagnosed with liver biopsy. MRI may show atrophy of the cortex, cerebellum, and brainstem and ventricular dilatation, but these findings are nonspecific.<sup>2</sup> Patients with hepatic symptoms may have increased signal in the basal ganglia on T1-weighted images.<sup>2</sup>

#### REFERENCES

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