

# pathology question

A 70-year-old man presents with loss of memory for recent events. He has forgotten his grandchildren's names, and he has been unable to manage his personal finances. Also, he has lost his way while driving to familiar locations. Which of the following is the most likely diagnosis?

- (A) Alzheimer disease
- (B) ALS
- (C) Creutzfeldt-Jakob disease
- (D) Huntington disease
- (E) Parkinson disease

The answer is A. Alzheimer disease is a major cause of dementia and is characterized by relatively slow, progressive memory loss followed in later stages by motor problems, contractures, and paralysis. Morphologic findings in Alzheimer disease include neurofibrillary tangles within neurons in the cerebral cortex, neuritic (senile) amyloid plaques, Hirano bodies, and generalized cerebral atrophy.

A 45-year-old man presents with involuntary facial grimaces and movements of the fingers. His mother had had similar symptoms beginning at about the same age. Her disorder had progressed to dancing movements, writhing of the arms and legs, and eventually coma and death. His maternal grandfather had had a similar disorder but at an age older than the mother. Which of the following is most characteristic of this disease?

- (A) Degeneration of upper and lower motor neurons
- (B) Dopamine depletion and depigmentation of the substantia nigra
- (C) Increased number of trinucleotide repeats in a gene on chromosome 4
- (D) Neurofibrillary tangles and amyloid plaques in the cerebral cortex

The answer is C. This is a case of Huntington disease, which is an autosomal dominant, fatal, progressive degeneration and atrophy of the striatum (caudate nucleus and putamen). The disorder is characterized by an increased number of trinucleotide (CAG) repeats in the HD (huntingtin) gene on the short arm of chromosome 4. Degeneration of the upper and lower motor neurons is characteristic of ALS. Dopamine depletion and depigmentation of the substantia nigra is characteristic of Parkinson disease. Neurofibrillary tangles and amyloid plaques are found in Alzheimer disease. Pick bodies can be found in Pick disease, which clinically resembles Alzheimer disease.

Several years ago, a 60-year-old woman had presented with bradykinesia, rigidity, a resting pill-rolling tremor in her right hand, and "mask-like," expressionless facies. She currently presents with gait problems, taking short, shuffling steps and losing her balance easily. Which of the following is the most likely diagnosis?

- (A) Alzheimer disease
- (B) ALS
- (C) Creutzfeldt-Jakob disease
- (D) Huntington disease
- (E) Parkinson disease

The answer is E. Parkinson disease, or parkinsonism, is characterized by a resting pillrolling tremor, masked facies, slowness of movements, muscular rigidity, and a festinating (shuffling) gait. Parkinsonism can be idiopathic or due to trauma, drugs, toxins (e.g., methyl-phenyl-tetrahydropyridine), or Shy-Drager syndrome. In the past, a major cause was von Economo encephalitis

A 56-year-old man with a past medical history of hypertension and diabetes mellitus is hospitalized after sustaining a mild concussion following a motor vehicle accident. His daily medications include aspirin, metformin, and lisinopril. He has smoked one pack of cigarettes daily for 20 years. He does not drink alcohol. He is married and works as an accountant. His family history is significant for coronary artery disease and diabetes mellitus.

His blood pressure is 143/95 mm Hg and pulse is 90/min with regular rhythm. His general physical and neurological examinations are unremarkable.

An MRI of the brain without contrast reveals a chronic infarct in the right basal ganglia without any evidence of intracranial bleeding.

This patient is at the highest risk for which of the following?

- A. Dementia [68%]
- B. Migraine headaches [7%]
- C. Multiple sclerosis [0%]
- D. Normal pressure hydrocephalus [3%]
- E. Seizure disorder [20%]

This patient's brain MRI shows a silent, chronic lacunar infarct in the right basal ganglia related to his hypertension and diabetes mellitus. Given his vascular risk factors, he is at risk of having additional ischemic events. These events can accumulate over time and cause significant damage to the surrounding white matter tracts and subcortical grey matter, which are essential for proper cognitive function. One study found that patients with silent infarcts with positive brain imaging have double the risk of developing dementia over the subsequent 5 years, compared to normal controls with normal brain imaging. Multiple silent infarcts were associated with an even higher risk than single infarcts. Therefore, this patient is at risk for developing vascular (Binswanger's) dementia in the future.



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