Multiple Choice Questions Section

The Neurosciences Journal introduces this new section on multiple choice questions as part of its commitment to continuous education and learning in Neurosciences. Experts in various neuroscience specialties are invited to participate with their knowledge and expertise in this section.

Neurology, neurosurgery, and other board residents are encouraged to read this section to improve their knowledge and direct their reading for written examinations.

Electrodiagnosis in neuromuscular disorders

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Choose the most appropriate single answer.

1. For which one of the following neuromuscular diseases, is nerve conduction study (NCS)/electromyography (EMG) the first choice as a diagnostic test?
   a. Duchenne/Becker muscular dystrophy
   b. Mitochondrial myopathy
   c. Spinal muscular atrophy (SMA)
   d. Hereditary motor sensory neuropathies (HMSNs)
   e. Acute inflammatory demyelinating polyneuropathy (AIDP/Guillain-Barré syndrome [GBS])

2. The normal sensori-motor nerve conduction with absent F-waves and tendon jerks of a moderately desaturated semicomatosed patient in the intensive care unit may be seen in:
   a. A classical pattern of critical illness myopathy involving both proximal and distal muscles
   b. Post-synaptic neuromuscular disorder
   c. The early stages of motor neuron disease (MND) (axonal loss) before the period of Wallerian degeneration
   d. Acute inflammatory demyelinating disorder like GBS
   e. A classic pattern found in presynaptic disorders such as botulism

3. Which one of the following statements is true for needle EMG in neuromuscular disorders?
   a. Decreased recruitment in severe botulism
   b. Fibrillation potential and positive sharp waves are absent after 14 days of myasthenic crisis
   c. Motor unit potentials (MUPs) remain stable and unchanged
   d. The early recruitment pattern of Lambert-Eaton myasthenic syndrome (LEMS) differs from that of botulism
   e. Single fiber EMG (SFEMG) is a diagnostic test for myasthenia gravis (MG)

4. Which of the following statements is not true for hereditary motor sensory neuropathy (HMSN) type I?
   a. Slowing of nerve conduction is uniform in all nerves
   b. Sensory studies are usually abnormal
   c. Conduction block and evidence of temporal dispersion are diagnostic hallmarks
   d. Compound muscle action potential (CMAP) amplitude may be very low in the lower extremities
   e. The EMG shows evidence of distal re-innervation

5. Which one of the following protocols is involved in the electrodiagnostic test approach for amyotrophic lateral sclerosis (ALS)?
   a. Sample at least one limb for proximal and distal muscles with different nerve innervation
   b. Cervical paraspinal muscles should be sampled for denervation
   c. A study for active denervation and reinnervation in 3 of the 4 body segments
   d. Bulbar muscle sampling is optional
   e. Sensory motor conduction for peripheral nerve showed significantly reduced velocity
Answers:

1. **d**
The HMSN are inherited polyneuropathies of variable course. In certain individuals it may progress very slowly (lifetime), therefore, family history and NCS/EMG as a first choice of investigation may help in determining the underlying cause (pg. 393). Duchenne/Becker muscular dystrophy, mitochondrial myopathy, and SMA are related to molecular genetics where DNA and other genetic analysis are the first choice of test. Acute inflammatory demyelinating disorder (AIDP/GBS) is an acquired inflammatory demyelinating process, and CSF analysis is the first choice (pg. 634).

2. **d**
Guillain-Barré syndrome typically begins at the root level as a demyelinating process, thereby attributing absent F-waves. However, absent F-waves in heavily sedated or comatose patients are normal findings (pg. 619). In critical illness myopathy, the sensory-motor conduction is normal including F-Waves, but the CMAP amplitude may be low. The needle EMG showed early recruitment with low amplitude and shorter duration polyphasic motor units. The interference pattern analysis showed horizontal clouds. Critical illness myopathy can be differentiated from critical illness neuropathy by direct muscle stimulation (pg. 623). In post-synaptic neuromuscular disorders like MG, the NCS and F-waves are normal. Repetitive nerve stimulation (RNS) at a slow rate shows decrement and immediate post-exercise facilitation (a repair of decrement). Needle examination may show unstable myopathic units (pg. 556). In the early stage of MND (axonal loss), before the period of Wallerian degeneration, sensori-motor conduction including F-wave and needle examination is usually normal. However, at a slow rate, the RNS may show decrement. In a classic pattern found in presynaptic disorders such as botulism, the CMAP amplitude is low with normal sensory conduction and F-wave latency. The EMG may show myopathic changes with an early recruitment and active denervation. However, the RNS gives decrement at a slow rate and increment at a high rate (pg. 620-621).

3. **a**
The recruitment of motor units is normal or reduced in the early stage, but if almost every muscle fiber of every motor unit is blocked by the toxin, there will be reduced recruitment similar to motor unit loss in axonal neuropathy (pg. 563). Fibrillation potentials and positive sharp waves are often seen after 4-5 days of severe myasthenic crisis, especially in botulism. Motor units are unstable, namely, the motor unit action potential (MUAP) may change from impulse to impulse, or may drop from time to time from the same motor unit. The morphology of the MUAP usually appears myopathic (pg. 564). Denervation sign, and decremental response may also be seen in severed denervating and myotonic disorders. In this situation, the unstable MUAPs favor neuromuscular disorders (pg. 565). The early recruitment may be seen in both LEMS and botulism, but it is the clinical findings that can differentiate LEMS from botulism. The SFEMG measures the variation in the time between the firing interval (jitter) of the 2 adjacent muscle fibers of the same motor unit (pg. 557). The SFEMG is very sensitive (95% to 99% in generalized MG), but non-specific (pg. 559). Any neuropathic or myopathic conditions with signs of denervation and reinnervation resulting from newly formed or dying neuromuscular junctions (NMJs) lead to increased jitter. In differentiating NMJ disorders from myopathy, an increased jitter of a normal NCS and EMG study of a patient with weak muscles offers a diagnostic clue.

4. **c**
The HMSN is a progressive disease with markedly slow nerve conduction velocity. The slowing is uniform without temporal dispersion or conduction block (pg. 399), which is a diagnostic hallmark for acquired demyelinating polyneuropathy with the exception of hereditary neuropathy with liability to pressure palsy (pg. 417). The sensory study usually shows low amplitude sensory nerve action potential, or absence of responses. As HMSN is a demyelinating disease, there is some secondary axonal loss leading to reduction of CMAP amplitude (pg. 399), signs of distal reinnervation, and signs of spontaneous activity.

5. **c**
The study of different proximal and distal muscles supplied by different nerves and roots of at least 3 of the 4 body segments is a proof that ALS is a diffuse MND, and is a completely distinct entity and different from mononeuropathies and radiculopathies (pg. 431). Thoracic paraspinal muscles should be sampled to avoid the coexistence of cervical spine disease. Bulbar muscle sampling for denervation/re-innervation signs is also helpful in differentiating ALS from polyradiculopathies. The nerve conduction study in ALS is essential to establish MND, where sensory conduction is normal as well as to exclude conduction block due to a demyelinating lesion such as multifocal motor neuropathy with conduction blocks (proximal stimulation should be accurately secured) (pg. 435).

References