Neurosciences Quiz

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Amyotrophic lateral sclerosis. *Is it a sequela of surgical intervention?*

Case Presentation

A 64-year-old male with diabetes mellitus (DM), on diet, presented to the hospital with generalized body weakness, slurred speech, choking during meals, loss of appetite and body weight of 20 kg. His symptoms started after he had inguinal hernia repair surgery. After which he started to develop weakness on the right lower limb below the knee. This weakness progressed until it included the whole right lower limb. Then, he started to have weakness on his left lower limb. Subsequently the weakness involved all of his body for the last 6 months. He had history of on/off central abdominal pain, moderate to severe, which was relieved by passing stool. Sphincter control was intact. There was no history of fever, tuberculosis, loss of consciousness, and other neurological symptoms. On examination, the patient was conscious, oriented, and cachectic with generalized muscle wasting. Higher mental function and sensory examinations were normal. The motor exam showed fasciculation of bilateral upper limbs. The muscle tone of the upper limb was normal. The power was 4/5 distally, and 5/5 proximally. Reflexes were exaggerated (biceps 3+, triceps 3+, brachioradialis 2+). The muscle tone of the lower limbs was also normal. Power was 4/5 on the right, and 4/5 on the left proximally. The planter flexion was 3/5 on the right and 4/5 on the left, and dorsi-flexion was 1/5 on the right and 3/5 on the left. Gait was unsteady. Routine blood tests including erythrocyte sedimentation rate were normal. Nerve conduction study (NCS) sensory nerve action potentials (SNAPs) for all 4 limbs were normal in their distal latencies and amplitudes. The compound muscle action potentials (CMAPs) were absent for the lower limbs. The CMAPs for median, ulnar, radial, musculocutaneous, and axillary nerves were low in amplitude. The motor nerve conduction velocity (MNCV) was normal/mildly slow for the upper limbs. There was no evidence of motor conduction block for the proximal stimulation of the upper limb long peripheral nerves.

Electromyography (EMG). The first dorsal interosseous, pronator teres, extensor digitorum communis (EDC), biceps brachii, triceps, deltoid, supraspinatus, pectoralis major, tibialis anterior (TA), medial head of gastrocnemius, short and long head of biceps femoris, vastus lateralis, gluteus medius, mid-thoracic (T6 to T10) paraspinal muscles, and genioglossus were sampled. There were runs of fibrillation potentials, positive sharp waves (3+). There were frequent fasciculation potentials and complex repetitive discharges (CRDs). The recorded motor unit action potentials (MUAPs) were mostly polyphasic, for a longer duration and larger in amplitude. The firing rate for MUAPs was markedly reduced. There were no MUAPs recruitments for TA. The quantitative EMG (QEMG) using interference pattern analysis showed neurogenic clouds. Single fiber EMG (SFEMG) for EDC muscles showed an increased jitter.

Questions

1. What is the diagnosis?
2. What are the electrodiagnostic features of amyotrophic lateral sclerosis (ALS) in this patient?
3. How are the electrodiagnostic features of cervical spondylitic radiculomyelopathy different from ALS?
4. How are the electrodiagnostic features of cervical multifocal motor neuropathy with conduction block (MFMNCB) different from ALS?
5. What will the SFEMG results in ALS be and why?

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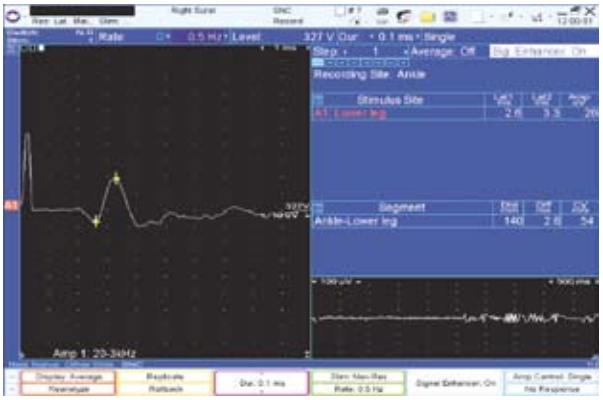

Figure 1 - Shows the recording of the right ulnar sensory nerve action potential (SNAP) (curve B) and right median SNAP (curve A).

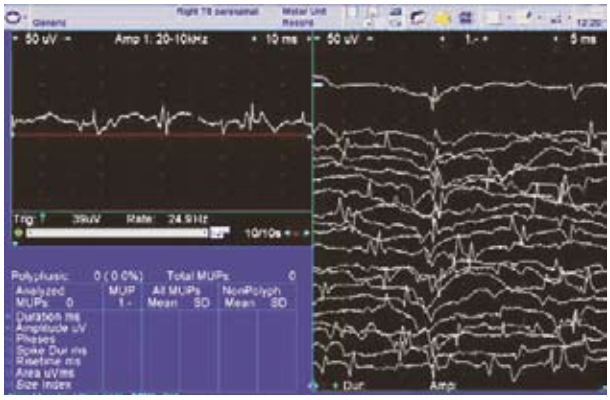

Figure 2 - Shows runs of fibrillation potentials and positive sharp waves, recorded from the T6 para-spinal muscle.

Answers & Discussion

1. Motor neuron disease/ALS, which are neurodegenerative diseases, characterized by progressive weakness and wasting of the muscles for speech, swallowing, respiration, and the extremities. The NCS showed normal SNAPs and sensory conduction of all the sensory nerves for upper and lower limbs (**Figure 1**).

2. Electrodiagnostic features: normal SNAPs and MNCV, low amplitude/absence CMAPs. There was no evidence of motor conduction block for the proximal stimulation of the upper limb long peripheral nerves. Active denervations and remodelled MUAPs with frequent fasciculation potentials seen in cervical, lumbosacral, thoracic, and bulbo cranial segments.[1-3] Hyperactive reflexes for both upper and lower limbs, marked muscle wasting and fasciculations. SFEMG: increased jitter, a non-specific finding. The above findings favor a diffuse process, possibly ALS rather than MFMNCB, and/or polyradiculopathy with spinal stenosis/myelopathy.

3. Sensory and motor nerve conduction studies were normal with the exception of low CMAPs and/or their absence especially in the lower limbs. Needle examination showed signs of active denervation, and chronic re-innervated MUAPs were also seen in the cervical paraspinal muscles and those peripheral muscles supplied by the cervical dermatomes. But, the presence of active denervation and chronic neurogenic MUAPs was unusual in T6 to T10 radiculopathy (**Figure 2**) as well as in bulbar muscles, as we have seen in our patient. However, radio-imaging studies for spondylotic changes in the lumbosacral regions are highly advisable for a clear association.

4. The electrodiagnostic features of MFMNCB mimic those for ALS. Therefore, the presence of proximal conduction block for motor nerve conduction should always be kept in mind, and that is why one should study motor conduction for the several proximal segments of all long peripheral nerves. One should also take area decrement of CMAPs for the definition of the conduction block. Needle EMG findings suggestive of diffuse denervation and re-innervation signs in paraspinal and bulbar muscles for MFMNCB are unusual.

5. The SFEMG showed increased jitter in ALS suggesting abnormalities in the neuromuscular junction. This may be due to inadequate neuromodulating mechanisms for the newly formed junctions after denervation. In our patient, the jitter was abnormal for the EDC muscle. We did not do fiber density studies because we did SFEMG studies using disposable concentric needle electrodes. However, SFEMG is sensitive for neuromuscular junction disorders, but is not specific for ALS.

References

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