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Electrophysiologic Abnormalities in a Patient with Syringomyelia Referred for Asymmetrical Lower Limb Atrophy

Asimetrik Alt Ekstremite Atrofisi ile Başvuran Bir Siringomiyeli Olgusunda Elektrofizyolojik Bozukluklar

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Dear Editor,

Hydromyelia was first described by Olliver d'Angers as cystic dilatation of the central spinal cord (1). Chiari described the hydromyelic cavity, which was related with or not related with the enlarged central channel, as "syringohydromyelia" (1). Conventional nerve conduction studies and needle electromyography (EMG) is very important in evaluating patients with syringomyelia but the findings may not be specific (2). We report a patient presenting with asymmetric lower extremity atrophy who was diagnosed as having a spinal cord abnormality.

A 18-year-old male was admitted with asymmetric lower extremity atrophy and foot deformity. Symptoms were non-progressive and he did not have sphincter dysfunction. There was no severe infection, systemic disease or difficult birth in his medical history. Neurologic examination showed that his bilateral dorsiflexion strength was 4/5. He had hypoesthesia and impaired vibration and position senses in his right lower extremity. Right patella and Achilles reflexes were absent. Bilateral plantar reflexes were extensor. He had bilateral foot deformities and a skin lesion in the lumbosacral area. Bilateral atrophy predominantly effecting the right side was observed. In the sensory nerve conduction studies, a right peroneal superficial nerve response could not be recorded and the right sural nerve amplitude was low. Motor nerve conduction studies showed low amplitude left peroneal nerve response. Needle EMG revealed long-term and polyphasic motor unit potentials (MUPs) with high amplitudes and showing a sparse pattern in left tensor fascia lata, extensor digitorum longus, and extensor digitorum brevis muscles. There was no pathologic spontaneous activity. Electrophysiologic findings were considered as asymmetric multiradicular involvement affecting the dorsal root ganglion at the lumbosacral level. Lumbosacral magnetic resonance imaging showed syringohydromyelia reaching an anterior-posterior diameter of 2-3 mm and a split cord abnormality in the distal spinal cord at the level of L1 vertebra and spina bifida occulta at the level of L4-L5 and L5-S1 (Figure 1A, 1B). The asymmetric sensory-motor involvement, pyramidal signs, and lumbosacral skin lesion were thought to be caused by the multiradicular effects of syringomyelia involving the dorsal root ganglion at the lumbosacral level.

Syringomyelia is a liquid-filled cystic cavitation in the spinal cord parenchyma. It is localized between C2-C9 and it can expand up or down. Usually, it accompanies Chiari type 1 malformation. Other causes of syringomyelia are congenital malformations, post-infectious, post-inflammatory, post-traumatic pathologies, and spinal neoplasia. It is characterized by dissociated sensorial loss, distal weakness in the upper extremities and pyramidal signs in the lower extremities (2). Syrinx may be asymptomatic and it may be detected in spinal cord imaging, incidentally. Some patients may present with a progressive spinal cord deficit, which includes central pain syndrome showing segmental distribution (3). Patients may be sent to the EMG laboratory with a pre-diagnosis of mono/polyneuropathy or trap neuropathy, which may lead to misleading results (1).

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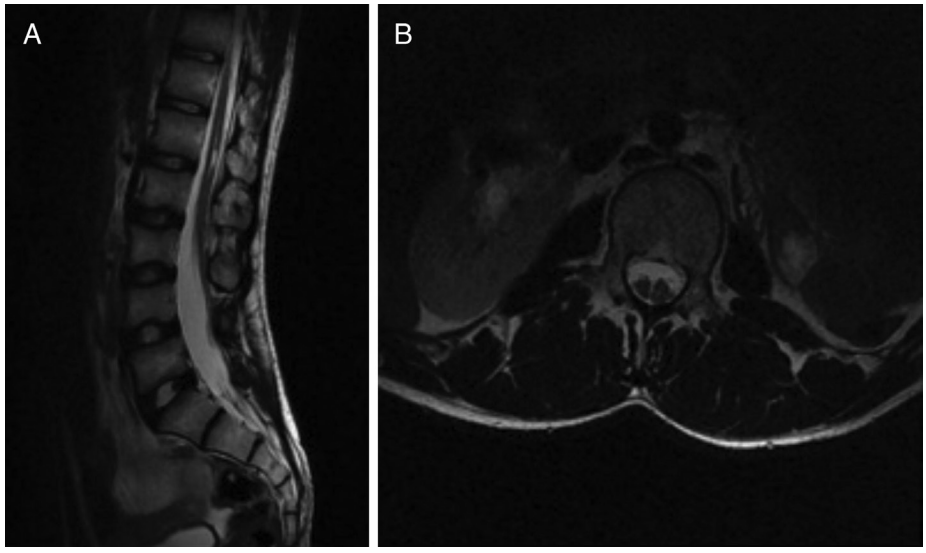

Figure 1. Diastematomyelia at the level of L2 vertebra corpus, syringomyelia cavity above and below this level, spina bifida occulta at the levels of L4-L5 and L5-S1 in sagittal (A) and transverse (B) lomber magnetic resonance images

Nerve conduction study abnormalities of syringomyelia include low amplitude compound muscle action potential, loss of F response or low amplitude F response with delayed latency. Sensory nerve action potentials (SNAPs) are usually normal (2). The right superficial nerve response could not be recorded and the right sural amplitude was recorded as low in sensory nerve conduction study in our patient. In cases where dorsal root ganglion cells and sensorial fibers extending to the periphery are intact and anatomic-physiologic integrity is protected, normal SNAPs are obtained, whereas in cases where there is damage in proximal axons reaching the spinal cord from dorsal root ganglion, dermatomal sensory deficit may occur (4). The sensory abnormality in our patient suggested that he had damage in the dorsal root ganglion. Low amplitude left peroneal motor nerve response could be caused by L5-S1 anterior horn damage due to syrinx. Asymmetric motor and sensory involvement findings could be suggestive of a polyneuropathy; however, normal sensory and motor nerve conduction responses in the upper extremities weaken the probability of polyneuropathy.

Needle EMG findings in syringomyelia are spontaneous activities including fibrillation, positive spikes and fasciculation, neurogenic MUPs, increased fiber intensity, jitter, and conduction blocks in varying degrees (5). However, these abnormalities sometimes cannot differentiate syringomyelia from motor neuron disease and poliomyelitis. In our patient, we recorded chronic neurogenic MUP changes in the muscles innervated by left L5 spinal root in needle EMG as reported in the literature. It was reported in the literature that fibrillation potentials could be seen in limited areas and especially in the muscles innervated by C8-T1 roots (1).

The distribution of muscle weakness and atrophy can guide in the differentiation of possible causes. Asymmetric lower extremity atrophy can be seen in patients with poliomyelitis sequela, but shortness of the extremity that is the most atrophic and a weak extremity draws attention in patients with poliomyelitis sequela. Foot deformities, muscle atrophy, weakness or absence of reflexes are suggestive of a hereditary polyneuropathy, but negative family history and nerve conduction studies guided us to exclude this diagnosis in our patient.

As a result, electrodiagnostic studies are very helpful in the detection and classification of peripheral nerve diseases. By presenting this case, we wanted to emphasize that EMG is a continuation of neurologic examination and it should be performed based on clinic findings.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: O.A., M.B.B., Concept: O.A., M.B.B., Design: O.A., M.B.B., Data Collection or Processing: O.A., Analysis or Interpretation: O.A., M.B.B., Literature Search: O.A., Writing: O.A.

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