CASE REPORT

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Amyotrophic lateral sclerosis mimicking radiculopathy: a case series

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ABSTRACT

The clinical findings of early stage amyotrophic lateral sclerosis (ALS) are similar to those of cervical or lumbar radiculopathy. In the current study, we describe 3 cases of ALS that were misdiagnosed as cervical or lumbar radiculopathy. Three patients (a 48-, a 52-, and an 80-year-old) visited our clinic due to motor weakness on either the upper or lower extremities. At other clinics or hospitals, they were diagnosed with radiculopathy due to herniated lumbar disc or cervical foraminal stenosis. The motor weakness in these patients was ipsilateral or bilateral but dominant on one side. Sensory deficits or bulbar symptoms were not observed. Of the 3 patients, 2 had neuropathic pain, but it was not dermatome related. We conducted an electrodiagnostic test and observed a low amplitude of compound motor action potential on the affected nerve and positive sharp waves on muscles of involved segments, including thoracic paraspinalis. All the patients were diagnosed with probable laboratory-supported ALS or possible ALS based on the established diagnostic criteria (El Escorial Criteria). In the present study, we showed three cases of ALS that were misdiagnosed as radiculopathy. Our study would be helpful for early and accurate diagnosis of patients with ALS.

Keywords: amyotrophic lateral sclerosis, mimic disorder, pain clinic, radiculopathy

Abbreviations:
ALS: amyotrophic lateral sclerosis
NRS: numeric rating scale
MRI: magnetic resonance imaging
CMAP: compound motor action potential
SNAP: sensory nerve action potentials
NCS: nerve conduction study

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INTRODUCTION

Amyotrophic lateral sclerosis (ALS), the most common motor neuron disease, is a progressive and devastating neurodegenerative disorder.[1] About 90% of ALS cases are sporadic, without a family history. The incidence of sporadic ALS is reported to be 1.89 per 100,000/year, and it is estimated that the general risk of ALS for a lifetime is 1:400 for women and 1:350 for men.[1] The risk of developing ALS peaks between the ages of 50 and 75 years.[2] It involves both the upper and lower motor neuron systems and has high morbidity and mortality.

ALS is diagnosed on the basis of the patients' clinical features and the results of an electrodiagnostic test. When patients present with bilateral motor weakness, the diagnostic accuracy is about 95%, but it reduces to 38% in patients with unilateral motor weakness.[3] Previous studies reported that ALS can be misdiagnosed because several neurological disorders have similar clinical presentations as ALS. Therefore, it is imperative to consider all possible disorders that can have similar manifestations before confirming the diagnosis of ALS.[4]

At pain clinics, pain physicians frequently encounter patients with motor weakness because it often accompanies neuropathic pain.[5] Although not frequent, ALS patients occasionally visit pain clinics without being diagnosed. In pain clinics, ALS can be easily missed or misdiagnosed as another disorder because pain physicians tend to consider only compressive neural disorders as differential diagnosis for motor weakness.

In the current study, we presented our experience from a pain clinic where ALS was misdiagnosed as cervical or lumbar radiculopathy. The study protocol was approved by the Institutional Review Board of a university hospital.

CASE REPORT

Case 1

A 48-year-old woman visited the pain clinic of our university hospital for motor weakness in the right lower extremity. Her weakness started 1 year ago and had slowly progressed. The weakness began at the right distal lower limb (ankle and toe dorsiflexors or plantar flexors) and progressed to the proximal muscles of the right lower limb. Additionally, she had slight pain of a tingling nature [numeric rating scale (NRS): 1] in the entire right lower extremity, which was not dermatome related. Prior to visiting our clinic, the patient had visited several clinics with the same symptoms and had been diagnosed with right L5 and S1 radiculopathy due to a herniated lumbar disc by clinicians. On lumbar spine magnetic resonance imaging (MRI), a central protrusion on L4-5 and a central extrusion on L5-S1 were observed (Fig. 1a). Cervical and thoracic spine MRI and brain MRI showed no abnormal lesions. The patient received repeated right L5 and S1 selective nerve root injection with corticosteroid, but her symptoms did not improve. Physical examination at our hospital revealed that her motor strength was 3/5 for her right hip flexor, knee extensor, and plantar flexor and 0/5 for her right ankle dorsiflexor and extensor hallucis longus on the Medical Research Council (MRC) scale for muscle strength. Moreover, we observed that the patient's left ankle dorsiflexor and extensor hallucis longus were mildly weakened (4/5 on MRC), but she had not noticed it. No sensory deficits or bulbar symptoms were observed. Bilateral biceps, triceps, knee, and ankle reflexes were normal. Mild atropy and fasciculation in the right calf muscles were obaserved. Compound motor action potential (CMAP) showed no response for the right peroneal nerve and decreased amplitudes for the right tibial and left peroneal nerves during the electrophysiological examination. The sensory nerve action potentials (SNAPs) of the bilateral superficial peroneal and sural nerves were normal. Nerve

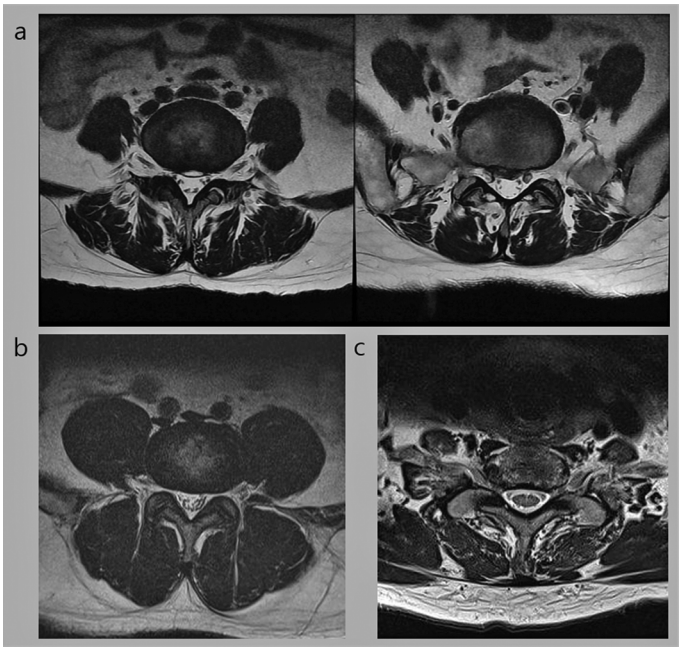

Fig. 1 Spine magnetic resonance imaging (MRI) findings of the 3 included patients
Fig. 1a (case1): On lumbar spine axial MRI, an L4-5 central protrusion (left) and L5-S1 central extrusion (right) are observed.
Fig. 1b (case2): On lumbar spine axial MRI, L4-5 diffuse bulging discs are observed.
Fig. 1c (case3): On cervical spine axial MRI, bilateral C7-T1 foraminal stenoses are observed.

conduction studies (NCSs) performed on the upper extremities were normal. On electromyography, positive sharp waves (2+~3+) were observed in the bilateral thoracic and lumbar paraspinalis and bilateral lower extremity muscles, including bilateral biceps femoris, tibialis anterior, peroneus longus, tensor fascia latae, gastrocnemius, and right vastus medialis. On follow-up at 6 months after the first visit to our department, the bilateral knee and ankle reflexes had increased. The patient was diagnosed with probable laboratory-supported ALS based on the established diagnostic criteria (El Escorial Criteria).[6] Her neuropathic pain in the entire right lower extremity was also considered as ALS-related.

Case 2

A 52-year-old man visited our pain clinic for weakness and pain in the entire right lower extremity, which was not dermatome related (dull nature, NRS: 2). His symptoms began 6 months ago and slowly became aggravated. At another university hospital, he was diagnosed with right L5 and S1 radiculopathy. On lumbar MRI, diffuse bulging discs were observed on L4-5 (Fig. 1b). Cervical and thoracic MRI and brain MRI showed no abnormality related to the patient's symptoms. On physical examination in our clinic, the motor strength of the right ankle dorsiflexor, ankle plantarflexor, and extensor hallucis longus was MRC 2/5. No sensory deficits and bulbar symptoms were observed. Bilateral biceps, triceps, knee, and ankle reflexes were normal. Also, mild atrophy was observed in his right calf muscles, but fasciculation was not found. On the NCS performed in our clinic, CMAP of the right peroneal nerve showed no response, and that of the right tibial nerve showed low amplitude. The NCS studies of the bilateral upper extremities

and the left lower extremity were normal. On electromyography, positive sharp waves (2+~3+) were observed in the bilateral lumbar and thoracic paraspinalis, tibialis anterior, and peroneus longus. On follow-up at 6 months after the first visit to our department, the muscle tones of the bilateral lower limbs had increased. Bialteral biceps and triceps reflexes were normal, but bilateral knee and ankle reflexes had increased. Based on the established diagnostic criteria, we diagnosed him with probable laboratory-supported ALS. Additionally, his pain in the entire right lower extremity was considered as ALS-related.

Case 3

An 80-year-old man visited our clinic for slowly progressive left distal upper extremity pain for 8 months without pain. In the previous hospital, he was diagnosed with left C8 radiculopathy due to C7-T1 foraminal stenosis. On the cervical MRI, foraminal stenosis on bilateral C3-4, C4-5, and C7-T1 was observed (Fig. 1c). On physical examination, his left finger flexor and wrist extensor were MRC 4/5, and sensory deficit was not observed. Bulbar symptoms were also absent. Bilateral biceps, triceps, knee, and ankle reflexes were normal. Slight muscle atrophy was observed in the left intrinsic hand muscles. On the other hand, fasciculation and split hand sign were not observed. On the NCS in our hospital, CMAP of the left median and ulnar nerves showed low amplitude but that of the right upper and bilateral lower extremities were normal. On electromyography, positive sharp waves (1+~2+) were observed in the left cervical paraspinalis, flexor carpi radialis, abductor pollicis brevis, abductor digiti minimi, 1st dorsal interossei, and bilateral thoracic paraspinalis and genioglossus. On follow-up at 6 months after the first visit to our department, the deep tendon reflexes of the bilateral upper limbs and the muscle tones of the bilateral upper limbs and trunk had increased. Based on the established diagnostic criteria, we diagnosed him with possible ALS.

DISCUSSION

In the current study, we presented 3 cases of ALS that were misdiagnosed as cervical or lumbar radiculopathies.

Early stage ALS and cervical or lumbar radiculopathy have similar clinical presentations. ALS is characterized by the loss of anterior horn cells.[1] When anterior horn cells are involved only in one or two segments in the early stage of ALS, it can be misdiagnosed as a radiculopathy due to herniated disc or spinal stenosis, which occurred in our patients who were misdiagnosed with lumbar or cervical radiculopathies.[4,7] Electrodiagnostic examination has an important role in the diagnoses of ALS and radiculopathy, but its results cannot distinguish between them in the early stage of ALS, where it can present as a radiculopathy.[7]

However, in the case of radiculopathy, the nerve roots are involved, which can produce both motor and sensory symptoms, and neuropathic pain can develop along the distribution of the corresponding dermatomes.[8,9] In contrast, ALS involves the anterior horn cells, affecting only motor fibers; therefore, sensory symptoms are not presented and neuropathic pain does not develop along the dermatome.[1] Hence, when pain physicians observe a patient who has motor weakness likely induced by radiculopathy without sensory symptoms, dermatomal neuropathic pain, or corresponding imaging findings, they should consider the possibility of ALS. Also, The incidence of asymptomatic MRI findings of spinal stenosis and herniated discs is high.[10] Accordingly, if the patients' radiculopathy symptoms are atypical, it is necessary to distinguish ALS through detailed examination and observation, although related lesions may be found on imaging studies.

CONCLUSION

In summary, by presenting 3 cases of ALS mimicking cervical or lumbar radiculopathy, we showed that the diagnosis of early stage ALS can be confused with radiculopathy. Our study would be helpful for early and accurate diagnosis of patients with ALS.

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CONFLICT OF INTEREST STATEMENT

The authors declare no conflict of interest.

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