



Case Report

Treatment of progressive paralysis associated with cervical myelopathy and suspected amyotrophic lateral sclerosis: A case report

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ABSTRACT

Background: Amyotrophic lateral sclerosis (ALS) is an intractable progressive disease, with an incidence of 2.2–2.3 per 100,000 individuals, which is not extremely low. ALS symptoms are accompanied by spinal myeloradicular motor deficit; its differential diagnosis is must because progressive paralysis needs emergency surgery.

Case Description: A 64-year-old man with suspected ALS showing progressive paralysis with cervical myelopathy was diagnosed as normal after performing a nerve conduction study preoperatively. Postoperative diffuse fasciculation after posterior decompression allowed the diagnosis of ALS through needle electromyography (EMG). Thereafter, the patient's condition slowly deteriorated and he died after 16 months.

Conclusion: Surgery might aggravate ALS symptoms; however, surgery for progressive paralysis in patients with suspected ALS is required for distinguishing patients with non-ALS paralysis. Approximately 70% of cases have spinal-onset ALS lacking typical cranial nerve symptoms; thus, to prevent unnecessary surgery, surgeons should at least know the characteristic features of ALS and should be aware that early diagnosis requires needle EMG for definitive diagnosis of ALS.

Keywords: Amyotrophic lateral sclerosis, Cervical myelopathy, Needle electromyography, Progressive paralysis, Treatment

INTRODUCTION

Amyotrophic lateral sclerosis (ALS) is an intractable progressive disease, with an incidence of 2.2–2.3 per 100,000 individuals,^[3] which is not extremely low. ALS symptoms are accompanied by spinal myeloradicular motor deficit with 67.4% of cases showing no cranial nerve symptoms.^[6] Conversely, progressive paralysis without sensory disturbance might be related to ventral root and spinal anterior horn compression. Thus, differential diagnosis is required because progressive paralysis needs emergency surgery. However, canal narrowing with spinal cord compression does not always exhibit neurological symptoms. The incidence of canal narrowing with spinal cord compression was significantly higher in patients with ALS than that in normal controls (ALS: 72.0%, controls: 29.5%).^[5] Moreover, surgical treatment at the onset of paralysis with ALS is suspected to be a risk factor for developing or worsening of

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ALS.^[5,6] Conversely, Yoshor *et al.*^[8] controversially reported that patients who had spinal surgery tended to have a longer interval between symptom onset and ALS diagnosis; 86% cases showed no improvement, 9% showed minor improvement and only 5% showed significant benefits.

A patient with spinal cord compression and suspected ALS lacking typical sensory deficit, who consulted a neurologist preoperatively, presented with progressive arm drop and difficulty in standing. Postoperative diffuse fasciculation was noted on a video, which led to the definitive diagnosis of ALS with needle electromyography (EMG). Here, the postoperative course of a missed-definitive diagnosis of ALS paralysis is presented and ways to prevent unnecessary surgical treatment for such ALS patients were retrospectively reflected.

CASE REPORT

A 64-year-old man, with no significant history except lumbar surgery with foot drop and sciatica, noted difficulty in right shoulder elevation, walking, and using chopsticks for a month. Physical examination revealed hyperreflexia and motor weakness in the upper limbs (shoulder elevation 3/5, elbow flexion 4/5, and finger extension 2-/3) and slow stream of urination without sensory disturbance. Bilateral Hoffman sign and Babinski sign were positive. Despite previous lumbar surgery, foot drop progressed bilaterally without numbness or sensory disturbance, implying the presence of motor neuron disease (MND). Imaging studies revealed canal narrowing at multiple locations with ossification of the posterior longitudinal ligament and spinal cord compression with T2 signal change at C4–C5 [Figures 1a and b; 2a and b]. Paralysis progressed to shoulder elevation 2/5- and standing difficulty in a few weeks, which required emergent spinal cord decompression as MND was excluded. He had no sensory disturbance but had a previous history of lumbar surgery for progressive foot drops. Although there were no cranial nerve symptoms, patient was referred to our acquaintance neurologist to exclude spinal-onset ALS. Motor and sensory nerve conduction velocity was examined, and the neurologist diagnosed the patient to be normal and recommended reconsultation if the patient showed dysphagia or dysarthria. We were unaware that at least needle EMG was required to exclude early-stage ALS.^[2] Moreover, ALS is reportedly associated with a higher incidence of spondylosis compared with normal control.^[5] Unfortunately, the patient could not stand at all and required prompt decompression. After the patient was explained in detail, the patient selected surgery to prevent progression of paralysis; informed consent was obtained. He underwent C3–C7 laminoplasty and right C4–C5 foraminotomy [Figures 3a and b]. Diffuse fasciculation appeared on the upper arms postoperatively [Video 1] with transient deterioration of

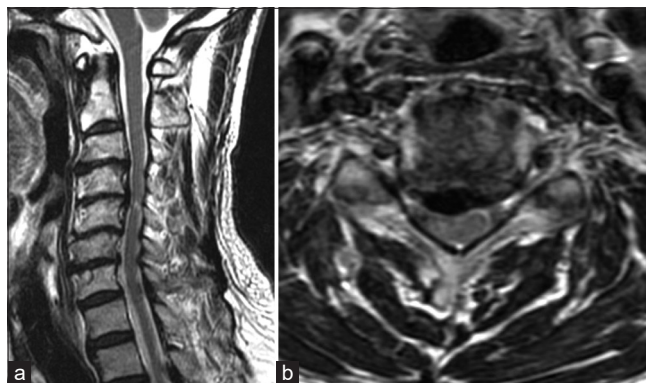


Figure 1: Sagittal magnetic resonance imaging (a) showing the multilevel spinal cord compression with T2 high signal intensity change at C4–C5. Axial image (b) showing foraminal stenosis at right C5 and spinal cord compression dominantly on the right side.

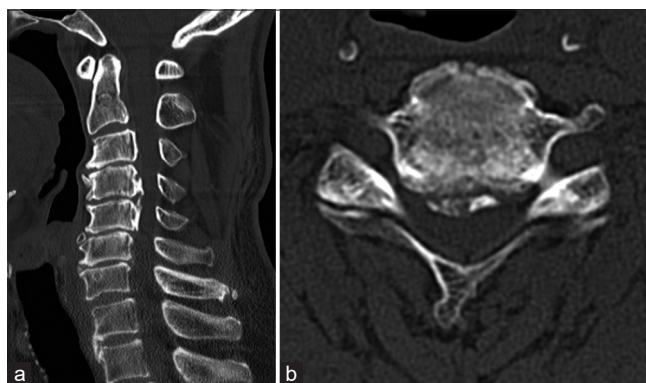


Figure 2: Computed tomography showing segmental ossification of posterior longitudinal ligaments at C4–C5 in sagittal view (a). Axial image at C4–C5 showing central to right dominant ossification and foraminal stenosis (b).



Figure 3: The patient underwent C3–C7 laminoplasty (C3, 7 dome osteotomy and C4–C6, laminoplasty) and right C4–C5 foraminotomy (a and b).

shoulder elevation 2-/5-. Intravenous administration of methylprednisolone 500 mg on the first day, which was gradually reduced to 125 mg on the third day, improved shoulder elevation to 2/5-. Clumsiness in both hands persisted, but he could walk with a T-cane. The right shoulder showed improvement in elevation to 100° during hospitalization. Based on information on fasciculation and reconsultation even without cranial nerve symptoms, the acquaintance neurologist diagnosed the patient with ALS through needle EMG. The neurologist informed us that the upper limbs' strength of the patient slowly deteriorated subsequently and he became totally debilitated at postoperative 8 months, with dysphagia being noted at 10 months. However, he refused artificial ventilation and died at 16 months.

DISCUSSION

The percentage of motor cells decrease to 30% at 1 year, and the mean duration for artificial ventilation is 3 years after ALS onset.^[1] ALS symptoms never improve and continue to progress but vary as rapid, intermediate, rapid on chronic, and slow.^[7] The modified El Escorial criteria and Awaji recommendations with the modified adaptation of needle EMG are recognized to prevent diagnostic delay and enable earlier diagnosis by mean of 6 months.^[2]

Avoiding surgery is better; however, treatment of progressive severe paralysis for suspected case without definitive diagnosis is unknown. Our emergent surgical indication for cervical spinal cord decompression is acute progressive paralysis accompanied by inability to stand or move extremities against gravity. In case with symptoms that do not clearly fit the criteria for cervical myelopathy, we consult a neurologist preoperatively. Spinal-onset ALS is noted in 67.4% of individuals without typical cranial or bulbar symptoms;^[6] our patient was suspected to have these symptoms; therefore, we referred him to a neurologist. Surgeons should recognize that definitive early diagnosis requires needle EMG at least. Pino *et al.*^[6] reported that surgery aggravates symptom progression; however, Yoshor *et al.*^[8] controversially reported no differences in age at symptom onset, severity of impairment at the time of diagnosis, or rate of disease progression between patients with ALS having spinal decompression and other patients with ALS. False-positive diagnosis and nonsurgical treatment of patients with suspected ALS who actually do not have ALS result in adverse outcomes such as complete paralysis. Slow progression is common but ALS symptoms neither show improvement nor remission. Previous report shows that even ALS cases exhibit symptoms of spondylotic myelopathy simultaneously.^[5] Moreover, sudden onset with cervical myelopathy in patients with ALS was reported.^[4] Considering foot drop, our case showed rapid on chronic progression but the early diagnosis was missed. It is not clear whether our surgery delayed or accelerated the progression

of ALS, but our patient also had simultaneous symptoms considering the postoperative recovery of shoulder elevation and walking ability. At the early stage of ALS even without muscle atrophy, denervations with needle EMG can be useful for diagnosis.^[2] Nerve conduction velocity is only useful in the differential diagnosis of other MNDs. Spine surgeons should refer and request neurologist to perform needle EMG in case of suspected spinal-onset ALS. Other characteristics such as positive tendon reflex with atrophic muscle, weight loss, wide fasciculation, tongue atrophy, and weakness of cervical flexion are late symptoms but should be considered. Negative symptoms such as disability in eye movement, sensory disturbance, and bladder and rectal disorders should be carefully examined.

Only 10% of patients have familial ALS with hereditary cause; thus, genetic diagnosis is not always required. Indeed, several genes lead to better understanding of ALS, but remission is impossible. ALS symptoms continue to deteriorate and never improve; however, simultaneous myeloradiculopathy might show improvement with decompression surgery. Such information should be examined in eligible patients preoperatively. Some drugs have been reported to delay progression; therefore, only early definitive ALS diagnosis can lead to a better quality of life in patients with ALS.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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