

## Cervical Spondylotic Myelopathy Mimicking ALS

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### Abstract

**Cervical spondylotic myelopathy presenting with muscle wasting in upper extremities and insignificant sensory loss has been termed as cervical spondylotic amyotrophy. This condition has to be differentiated from Amyotrophic lateral sclerosis which also has similar presentation. Here we present a case study of cervical spondylotic myelopathy resembling Amyotrophic lateral sclerosis clinically.**

**Key words:** Cervical spondylotic myelopathy, Cervical spondylotic amyotrophy, Amyotrophic lateral sclerosis

### Introduction

In cervical spondylotic myelopathy, it is usual to see hand presenting with spastic dysfunction and deficient pain sensation which has been termed "Myelopathy hand",<sup>1</sup> and occasionally a different type of myelopathy hand is seen which is characterized by muscle wasting and motor dysfunction in patients with cervical spondylosis. This type of myelopathy hand has been termed "Amyotrophic type of myelopathy hand".<sup>2</sup> Due to absent or insignificant sensory deficits this syndrome is sometimes confused with motor neuron disease, especially Amyotrophic Lateral Sclerosis (ALS).<sup>3</sup> We describe a case of cervical spondylotic myelopathy which mimics ALS clinically.

### Case Report

A 56 year old male came to our OPD with 8 months history of motor weakness in all four limbs. On physical examination, severe atrophy in both shoulder girdle including deltoid, supraspinatus, infraspinatus, scapular muscles, biceps, triceps, forearm muscles and small muscles of hands (Fig 1) were noted. Neurological examination showed decreased tone and depressed biceps and triceps reflexes. The muscle tone in lower limbs was increased with hyperactive deep tendon reflexes both knee and ankle and bilateral plantar extensor. Sensory changes showed decreased pinprick and vibration sensation both lower limbs without any significant sensory loss in upper limbs.

The motor nerve conduction velocity (MNCV) showed slowing of conduction velocities of both Median, Ulnar, Peroneal and Tibial nerves. CMAPs and distal latencies

were within normal limits. Sensory nerve conduction velocity (SNCV) showed slowing of conduction velocities of Median, Ulnar, and Sural nerves. SNAP amplitude diminished in both Sural, Right Ulnar and Left Median nerve,"F" latency marginally delayed in Left Ulnar and normal in others. EMG findings of the sampled muscles left and right Abductor Digiti Minimi, bilateral Abductor Pollicis Brevis, bilateral Extensor Digitorum Brevis, left Tibialis Anterior showed silence at rest, no abnormal resting potential, on volition-high amplitude long duration polyphasic potential were reduced, interference seen which were suggestive of chronic partial denervation both upper limbs and lower limbs.

The MRI of cervical spine was suggestive of C3-C4 posterocentral disc prolapse with central canal stenosis with canal anteroposterior (AP) APD 10mm, C4-C5 posterocentral disc prolapse with central canal stenosis with canal anteroposterior (AP) APD 6mm with cord compression and atrophy (Fig 2), C5-C6 posterocentral disc prolapse with central canal stenosis with canal anteroposterior (AP) APD 7mm with cord compression, C6-C7 posterocentral disc prolapse with central canal stenosis with canal anteroposterior (AP) APD 8mm C7-D1 posterocentral disc prolapse with central canal stenosis with canal anteroposterior (AP) APD 11mm. The patient was operated a week later, C4 corpectomy with adjacent disectomy and screw plate fixation C3-C5 done.

One month into the post operative period, patient was referred to us and admitted to the rehabilitation ward of our hospital. By now, he developed pain right side of neck, poor voice quality and required effort for speech. On examination of oral peripheral mechanism, lips showed mild weakness in terms of strength and sealing was inadequate at times. Jaw drooped when fatigued and

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Figure 1. Hand muscle wasting

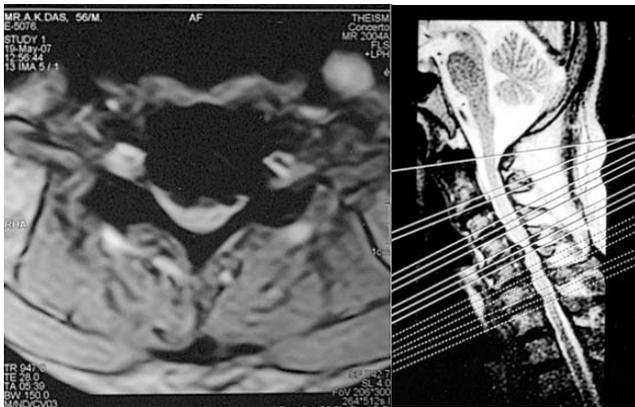


Figure 2. Axial T2 weighted MRI at C4-5 level showing reduced transectional area of spinal cord and extradural cord compression by calcified herniated disc. Sagittal T2-weighted MRI showing cord compression from C-3-4 to C-6-7 most notable at C-4 to C-5, with linear region of increased signal intensity is extending from C-4-6.

phonation time shorter with uneven changes in laryngeal volume (from Frenchay Dysarthria assessment). Cranial nerve examination revealed no marked impairment. Indirect laryngoscopy showed left vocal cord moving less and bowing during phonation (phonatory gap), right vocal cord compensating. Diagnosis of flaccid Dysarthria secondary to spinal nerve lesion was made. Patient was put to extensive physiotherapy, occupational therapy and speech therapy and is showing improvement.

## Discussion

Muscular atrophy in upper extremities with insignificant sensory loss is occasionally caused by cervical spondylosis, <sup>4, 5</sup> usually named as cervical spondylotic amyotrophy. In the age group of 50-70 years both cervical spondylotic myelopathy and ALS are common and has to be differentiated from each other.<sup>6</sup>

In the present case, along with severe muscular atrophy with lower motor neuron findings in both upper limbs and upper motor neuron findings in both lower limbs, patient also developed dysarthria which were characteristic of ALS. Based on clinical presentation, radio and electro diagnostic tests and post treatment results, we were able to diagnose it as a case of cervical spondylotic amyotrophy and rule out ALS.

It has been reported that for accurate diagnosis of cervical spondylotic amyotrophy, attention should be paid to the narrow anteroposterior (AP) canal diameter of the cervical spine (less than 13mm), multisegmental spondylosis in C5-C6 and C6-C7 discs levels and reduced transectional area of the spinal cord at the C7,C8 and T1 spinal cord segments. <sup>2</sup> In this case also there was narrowing of anteroposterior (AP) canal diameter of the cervical spine (less than 13mm) at multisegmental levels (Fig 3) and electro diagnostic tests were consistent with cervical spondylotic myelopathy. Disc protrusions, posterior osteophytes, retrolisthesis, ligamentous entrapments are well known etiologic factors for myelopathy <sup>5</sup> but the exact patho-physiology of this particular syndrome (cervical spondylotic amyotrophy) has not been well understood, Kameyama et al report that this syndrome may be multisegmental damage to the anterior horns caused by dynamic cord compression, possibly through circulatory insufficiency.<sup>7</sup>

## Conclusion

Critical analysis is necessary to differentiate cervical spondylotic amyotrophy a type of cervical spondylotic myelopathy from ALS.

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