Case Report

Intermittent Compression In Cervical Dystonia: Rare Cause of Myelopathy

Tanvir RIZVI¹, Vipul GUPTA¹, Ajay GARG¹, Vinay GOYAL², Shailesh GAIKWAD¹, Nalinikant MISHRA¹

¹Department of Neuroradiology, AIIMS, New Delhi, INDIA
²Department of Neurology, AIIMS, New Delhi, INDIA

Abstract
The clinical and imaging features of two patients of cervical dystonia, who presented with signs & symptoms of myelopathy of recent onset, were studied. MRI documented atlantoaxial subluxation with cord compression and signal change at cervicomedullary junction in the first case, possibly due to overstretching or rupture of transverse ligament by the repeated neck movements. In the second case, there was C4/5 disc bulge. In spite of lack of cord compression, there was cord signal change from C4 to C6 level probably because of intermittent cord compression, ischaemia or ligament stretching due to extreme neck movements. Therefore, secondary myelopathy in patients suffering from cervical dystonia may result from unusual causes such as atlantoaxial instability or due to extreme neck movements superimposed upon degenerative changes.

Key Messages: Secondary myelopathy in patients suffering from cervical dystonia may result from unusual causes such as atlantoaxial instability or due to extreme neck movements superimposed upon degenerative changes

Keywords: Cervical spondylosis, Craniovertebral junction

Servikal Distonide İntemittan Baş; Myelopatinin Ender Bir Nedeni

Özet
Myelopati yakınıması ile başvuran servikal distonili iki hastanın klinik ve görüntüleme özelliklerini sunulmuştur. MRG’de ilk hastada, tekrarlayan boyun hareketleri sonucu transvers ligamanın ileri gerilmesine bağlı atlantoaksiyal subluksasyon, kord bası ve servikomedüller bileşkede sinal artımı saptandı. İkinci olguda ise C 4/5’ de bulging ve kord basısı saptanmamışına rağmen, ileri boyun hareketleri sonucu ligaman gerilmesine bağlı veya olası intermittent kord basısına bağlı C4 C6 arası sinal değişikliği görüldü. Bu nedenle, servikal distonili hastalarda dejeneratif değişiklikler ya da atlantoaksiyal insitabilitesi zemininde, ileri boyun hareketlerine bağlı olarak ikincil myelopati gelişebilir.

Anahtar Kelimeler: Cervical Dystonia, Atlantoaxial subluxation, Myelopathy
Introduction

Dystonia is characterized by abnormal posturing of body parts due to sustained or repetitive contraction of muscles. Dystonia may result in secondary complications, such as compression of the peripheral nerves, myeloradiculopathy and cervical spondylosis.\(^1\)\(^2\) We report two rare cases of cervical dystonia associated with atlantoaxial subluxation and cervical myelopathy, in which intermittent compression associated with neck movements played a role in myelopathy.

Case Presentation

Case 1:
A 34-year-old man presented with two months history of progressive weakness of upper limbs, frequent falls and difficulty in speaking in last one month. He had abnormal neck posturing, facial grimacing, forehead wrinkling, deviation of bilateral angle of mouth and stiffening of the neck muscles since six months of age. On examination, he had generalized dystonia with predominant involvement of neck, face & upper limb muscles. The movements involving neck were predominantly antero-posterior in direction. In upper limb there was wasting of small muscles of hand with hand grip weakness. Deep tendon reflexes were brisk. Muscle bulk and power were normal in lower limbs. Plantars were bilaterally flexor.

Craniovertebral junction MRI showed atlantoaxial subluxation, with distance between anterior arch of atlas and dens being greater than 5mm, resulting in canal narrowing & cord compression. The spinal cord showed focal hyperintensity at C1 level on T2WI, which was hypointense on T1WI (Fig.1 A-B). Rest of the cervical spine showed severe degenerative changes with osteophyte formation in vertebrae C4, 5 & 6 and disc bulges at C5/6 & 6/7 levels. MRI of brain was normal. Routine blood investigations including serum copper & ceruloplasmin were normal. Hence the diagnosis of idiopathic dystonia was kept. He was treated symptomatically with anticholinergics. He was referred to Neurosurgery for the possibility of surgical intervention, which he refused and was advised cervical collar. At six-month follow-up, he showed significant improvement in myelopathy, particularly the motor symptoms.

![Figure 1: Sagittal T1- (A) & T2- (B) weighted images reveal atlantoaxial dislocation, canal narrowing, cord compression & cord signal change at the cervicomedullary junction.](image)

Case 2:
A 31-year-old man had a history of birth asphyxia during twin vaginal delivery. He had dystonic cerebral palsy since birth. For one & half years prior to admission, he had history of increasing dystonia, along with increasing weakness and stiffness of both upper & lower limbs with frequent falls during walking. For last six to seven months, he had deviation of neck to left side on trying to speak. He had history of bowel & bladder incontinence for two months. On examination, he had spasmodic torticollis. He had blepharospasm, oromandibular dyskinesia with dysarthria. His power was 4/5 in upper limbs and 4+/5 in lower limbs. There...
was spasticity of all four limbs (LL>UL). Deep tendon reflexes were brisk, with ankle clonus and bilateral extensor plantars.

MRI cervical spine (Fig.2 A-D) revealed degenerative changes & diffuse disc bulge at C4/5 level, not causing cord compression. However, spinal cord showed signal alteration in the form of hyperintense signal on T2WI & hypointense signal on T1WI and mild cord expansion extending from C4 to C6 level. Craniovertebral junction & brain were normal. He was treated with anticholinergics and baclofen. He was also advised cervical collar, but was lost to follow-up.

**Discussion**

Cervical dystonia may result in orthopaedic and neurological complications, including cervical spine degeneration, spondylosis, disk herniation, vertebral subluxations and fractures, radiculopathies and myelopathies. Awareness of frequent occurrence of complications and screening for symptoms of radiculomyelopathy in patients with dystonia is essential to avoid irreversible spinal cord damage.

Rarely patients can present with atlantoaxial dislocation. Atlantoaxial dislocation usually occurs due to congenital causes (occipitalization of atlas, os odontoideum, aplasia &/or dysplasia of dens, Down’s syndrome), trauma, rheumatoid arthritis, ankylosing spondylitis, and infections of pharynx and upper neck. Rarely, instability of the upper cervical spine sometimes results from dysfunction of the ligaments without fractures. Because the ligaments consist mainly of collagen fibers, repeated trauma to the cervical spine could lead to irreversible overstretching, or even rupture. Dystonia results in repetitive antero-posterior movements, which can rarely cause irreversible overstretching or rupture of the transverse ligament leading to atlantoaxial subluxation, as was seen in our first case. There is only one similar case reported in literature by Adel Al-Jishi, et al, who found atlantoaxial subluxation and cervical spondylosis with cord compression at C5-6 and C6-7 levels on MRI in a 46-year-old male patient of primary dystonia. However, there was no cord compression or signal change at cervicomedullary junction. Hence, ours is the first case in

![Figure2: Sagittal T1- (A) & T2- (B) weighted images reveal spondylotic changes with a small disc at C4-C5 level without any evidence of significant cord compression (Axial T1WI-C). The cord shows increased signal from C4-C6 level on T2WI (B, D).](image-url)
literature showing atlantoaxial subluxation with secondary myelopathy in a patient of dystonia.

In our second case, the degenerated disc was not causing significant compression, but the cord showed signal change at that level. Although, compression is thought to be the main factor causing myelopathy in cervical spondylosis, ischemia may also contribute to cord damage\(^7\). The cord may be rendered ischaemic during neck flexion and extension, when the cross-sectional area of the canal decreases in extension by up to 16\(^\%\)\(^8\). The normal canal lengthens in flexion and shortens in extension, but if cord movement is restricted by root sleeve and dural fibrosis or osteophytes, then traction forces may develop, which may result in myelopathy\(^9\). Myelopathy may also result from repeated injury to the cord during flexion-extension movements of the neck, when the cord is squeezed between the ligamentum flavum and an osteophyte\(^1\). Probably, the mild degenerative changes along with excessive neck movements were resulting in intermittent cord ischemia, stretching or compression and thereby causing myelopathy in our case. Thus factors other than degenerative changes may play a role in myelopathy in these cases and should be considered during clinical decision-making. This may imply that immobilization may suffice in some of these cases by preventing cord injury due to the excessive neck movements. MRI should be done in such cases to document the cord injury, even in patients with mild degenerative changes.

In our second case, C4/5 disc was involved. Nishihara & colleagues\(^2\) in their study of nine athetoid patients with cervical spondylotic myelopathy also found that C3-C4 and C4-C5 were the most frequently affected sites, as opposed to C5-6 involvement in older patients with cervical degeneration. They attributed this difference to the different patterns of movement at the cervical spine. Rotation and lateral bending, which are greater in persons of torticollis and athetosis occur at C3 to C4 and C4 to C5, predisposing to degenerative changes at these levels. Therefore, as seen in our cases, analysis of the type of neck movements may guide towards the likely site of injury in these patients.

The management of this complication is controversial. Nishihara & colleagues suggested anterior fusion to be the better surgical procedure for patients with cervical spondylotic myelopathy with movement disorder rather than cervical laminectomy\(^2\). However, immobilization of cervical spine postoperatively is extremely difficult, because of the involuntary movements\(^10\). Symptomatic support with cervical collar may add to the necessary stability and limitation of movement, as in our first case. Focal dystonia patients can also benefit from local injection of botulinum toxin\(^11\).

**Conclusion**

To conclude, cervical dystonia may be rarely associated with cervical myelopathy. The flexion-extension movements can cause cervical myelopathy at C-V junction while rotation and lateral bending are likely to cause myelopathy at C3-4 or C4-5 levels. Potentially dangerous complication like atlantoaxial subluxation, may be responsible for cord compression in dystonia. Cervical myelopathy may occur due to repetitive trauma caused by dystonia, which may be responsible for secondary deterioration in such patients. Such cases may go undiagnosed in the absence of imaging.

**References**


Correspondence
Tanvir RIZVI
Department of Neuroradiology, AIIMS, New Delhi, INDIA

E-mail: rizvi_tanvir@hotmail.com

Received by: June 14.2005
Revised by: Aug 12.2005
Accepted: Sept 05.2005

The Online Journal of Neurological Sciences (Turkish) 1984-2005
This e-journal is run by Ege University Faculty of Medicine, Dept. of Neurological Surgery, Bornova, Izmir-35100TR as part of the Ege Neurological Surgery World Wide Web service.
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URL: http://www.jns.dergisi.org
Journal of Neurological Sciences (Turkish) ISSN 1302-1664