



IDIOPATHIC SPINAL CORD HERNIATION

İDİOPATİK SPİNAL KORD HERNİASYONU

Bahadır GÖKCEN¹,
Meriç ENERCAN²,
Mesut KILIÇ³,
Erden ERTÜRER⁴,
Çağatay ÖZTÜRK⁵

¹Assist. Prof., Surgeon of the Orthopaedics and Traumatology, İstanbul Science University Medical Faculty, Department of Orthopaedics and Traumatology, İstanbul.

²Surgeon of the Orthopaedics and Traumatology, Department of Orthopaedics and Traumatology, İstanbul Florance Nightingale Hospital, İstanbul.

³Assist. Prof., Surgeon of the Orthopaedics and Traumatology, Ondokuz Mayıs University Medical Faculty, Department of Orthopaedics and Traumatology, Samsun.

⁴Assoc. Prof., Surgeon of the Orthopaedics and Traumatology, İstanbul Science University Medical Faculty, Department of Orthopaedics and Traumatology, İstanbul.

⁵Assoc. Prof., Surgeon of the Orthopaedics and Traumatology, Department of Orthopaedics and Traumatology, Şişli Florance Nightingale Hospital, İstanbul.

Address: Op. Dr. Bahadır Gökçen, Surgeon of the Orthopaedics and Traumatology, İstanbul Science University Medical Faculty, Department of Orthopaedics and Traumatology, İstanbul.

Tel.: +90 533 574 21 04

E-Mail: bahadrgokcen@gmail.com

Received: 12th November, 2014

Accepted: 22th December, 2014

SUMMARY:

Idiopathic spinal cord herniation is a rare diagnosis. The differentiation of spinal cord herniation from arachnoid cyst is important. MRI (Magnetic Resonance Imaging) is helpful for the diagnosis. We report a patient with lumbar spinal stenosis and an idiopathic spinal cord herniation at the thoracic level.

Key words: Idiopathic spinal cord herniation, thoracic spine, surgical treatment

Level of evidence: Case report, Level IV

ÖZET:

İdiopatik spinal kord herniasyonu nadir görülen bir durumdur. Spinal kord herniasyonunu araknoid kistten ayırımını yapmak önemlidir. MRG (Manyetik Rezonans Görüntüleme) tanıda yardımcıdır. Biz bu çalışmamızda torasik bölgede idiopatik spinal kord herniasyonu ve lomber spinal stenozu olan bir hastayı bildirdik.

Anahtar kelimeler: İdiopatik spinal kord herniasyonu, torakal omurga, cerrahi tedavi

Kanıt düzeyi: Olgu sunumu, Düzey IV

INTRODUCTION:

Idiopathic spinal cord herniation is a rare diagnosis. The differentiation of spinal cord herniation from arachnoid cyst is important. MRI (Magnetic Resonance Imaging) is helpful for the diagnosis. We report a patient with lumbar spinal stenosis and an idiopathic spinal cord herniation at the thoracic level.

CASE REPORT:

A 43-year-old woman with a history of weakness and numbness in her right leg presented to the clinic. Neurological examination revealed as lightly decreased muscle strength in the right leg (clinical grade of motor power was 4/5 for ankle dorsiflexion) and full muscle strength in the left leg (motor power was 5/5). She had hypoesthesia at T5-T6 level with decreased position sensation bilaterally.

She reported no bowel or bladder complaints. The rest of the general and neurological examinations revealed no significant abnormalities. Based on the neurologic examination, a sacral L5-S1 root injury and thoracic medullary compression were considered. She had no significant history of trauma, surgery or infection. Her laboratory tests were all within normal limits. Plain films of the thoracic and lumbar spine were unremarkable. The patient underwent MR imaging of the thoracic and lumbar spine on 1.5T Sagittal T1- and T2-weighted MR sequences obtained on a 1.5-T imaging unit, which showed ventral displacement of the spinal cord at the T5-T6 level with prominence of the posterior subarachnoid space at this level, in addition to lumbar spinal stenosis at L4-L5 and L5-S1 levels (Figure-1,2). Sagittal T2-weighted MR image showed a focal anterior kink of the spinal cord at the T5-T6 level. The cord was adjacent with the posterior surface of the vertebral body. Additional 26 imaging studies were performed due to a possible posterior intradural arachnoid cyst.

Myelography and CT myelography confirmed the ventral displacement of the cord at the T5-T6 level. Widening of the dorsal subarachnoid space without any filling defect was shown on CT myelography (Figure-3). Calcified material from chronic disc herniation, and herniation of the left anterolateral portion of the cord through a dural defect were detected on axial CT myelogram (Figure-4).

Depending on the radiological findings, idiopathic herniation of the spinal cord and lumbar spinal stenosis were diagnosed. We operated on the patient for lumbar spinal stenosis and confirmed spinal cord herniation. The neurologic findings regressed after surgery. Right and the left leg muscle strengths were equal (clinical grade of motor power was 5/5).



Figure-1. Thoracic sagittal image of the MR of the patient.

Figure-2. Lumbar sagittal image of the MR of the patient with the spinal stenosis in the L4-5 disc level.

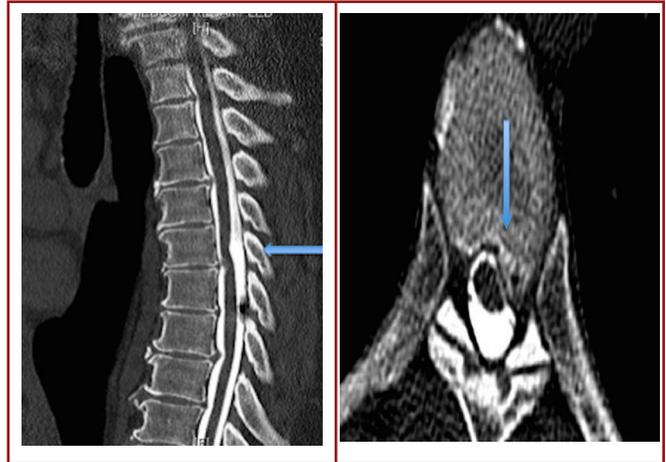


Figure-3. Thoracic sagittal image of the MR of the patient postoperatively

Figure-4. Thoracic axial image of the MR of the patient postoperatively

DISCUSSION:

Idiopathic thoracic spinal cord herniation is a rare and uncommon cause of myelopathy⁶. Brown-Séquad syndrome can be the initial presentation of idiopathic spinal cord herniation^{8,9,12,13}. In idiopathic spinal cord herniation, spinal cord herniated or prolapsed through an anterior or lateral defect in the dura mater⁶.

The presence of a dural defect can be considered as a precondition for the development of idiopathic spinal cord herniation. Entry of the cerebrospinal fluid into the extradural space through this defect forms a localized fluid collection which can be referred to by some as an extradural arachnoid cyst¹.

Cerebrospinal fluid pulsations force the spinal cord through this defect that can lead to adhesions, distortions and possible vascular compromise of the cord, causing progressive myelopathy³. Idiopathic spinal cord herniation has an unknown cause which is unrelated to trauma or a previous operation. Pressure erosion, thoracic disc extrusion, congenital disorder (preexisting ventral meningocele), duplication of the ventral duramater, congenital extradural arachnoid cyst, inflammatory process and congenital abnormal adhesions of the spinal cord to the anterior dura are possible underlying causes that have been postulated to explain the occurrence of this dural defect^{1,10}.

Idiopathic spinal cord herniation is probably an under diagnosed disorder that is being diagnosed more commonly due to increased use of MRI⁵. It is diagnosed by its characteristic imaging appearance. Imaging features of spinal cord herniations generally include a dural tear through which a portion of the cord protrudes.

Cerebrospinal fluid flows freely through the defect, causing increased turbulence in the fluid just dorsal to the site of herniation. The observation of this feature may allow the differentiation of spinal cord herniation from an arachnoid cyst. In addition, the calcification of nucleus pulposus leakage from a herniated disk may produce a linear area of hyperattenuation on computed tomography or signal hyperintensity on magnetic resonance imaging, the latter is an imaging feature known as the “nuclear trail” sign⁶.

The first report of an idiopathic spinal cord herniation was published by Wortzman et al¹⁴. Our review of the relevant literature revealed only limited number of reports, and most cases were published in the neurosurgical literature. The features of this condition in imaging need to be recognized. Several of the reported cases were initially misdiagnosed and resulted in a delay in the correct management⁵.

The most common misdiagnosis in the literature was misinterpretation of the expanded dorsal subarachnoid space for an arachnoid cyst². In previous reports, this condition is often seen in middle-aged people (range 21-78 years; mean 51 years) with a female predominance (male to female 1:1.8)⁵. In the 80% of the patients, the T2-T7 levels are affected with all other cases occurring in the thoracic spine (range T2 to T9). Negative pressure in the thoracic extradural space and proximity to the heart (high cerebrospinal fluid pressure) accounts for the thoracic distribution of spinal cord herniation. Most of the cases have been reported with Brown- Séquard syndrome which is the most frequent clinical presentation of spinal cord herniation and seen in approximately 80% of patients⁵.

The tract affected first by herniation is the lateral spinothalamic tract. This results in diminished pain and temperature sensation, and is frequently unilateral and ascending. As the

corticospinal tracts become involved, gradual, progressive weakness and spasticity of the leg(s) also occur. The resultant dissociated sensory deficit with asymmetric spastic paraparesis is known as Brown-Séquard syndrome¹.

Other presentations of this condition were also reported, including included spastic paresis, bowel or bladder sphincter dysfunction, isolated motor or sensory disturbance, and chest pain^{2,11}.

Most of the previously cases were treated surgically, and published principally in the neurosurgical literature. The main purpose of surgical treatment is decreasing the pressure and reducing the spinal cord herniation. Surgery is usually indicated in patients with progressive myelopathy and Brown-Séquard syndrome. It is believed that the outcomes of surgery are more successful than cases with spastic paraparesis^{4,5}. In our patient we did not plan an operation for idiopathic spinal cord herniation due to the reported complications of surgery². We have planned the surgery for lumbar spinal stenosis, we have done posterior instrumentation and posterior laminectomy.

After the operation there was a progressive improvement in the neurologic status of patient. Similar to our management, there are other authors who also did not consider surgery in idiopathic spinal cord herniation⁷. If the patient has no progressive myopathy and Brown-Séquard syndrome, observation in this condition alone can be helpful for preventing the reported complications of surgery. Our case emphasized the importance of neurologic examination and also pointed the importance of rare conditions that all physicians have to keep in mind. We believe that increasing use of the spinal MRI will decrease the rates of misdiagnosis in idiopathic spinal cord herniation.

REFERENCES:

1. Blasel S, Hattingen E, Baas H, Zanella F, Weidauer S. Spontaneous spinal cord: MR imaging and clinical features in six cases. *Clin Neuroradiol* 2008; 18: 224-230.
2. Darbar A, Krishnamurthy S, Holsapple JW, Hodge CJ. Ventral thoracic spinal cord herniation: frequently misdiagnosed entity. *Spine* 2006; 31: 600-605.
3. Dix JE, Griffitt W, Yates C, Johnson B. Spontaneous thoracic spinal cord herniation through an anterior dural defect. *AJNR Am J Neuroradiol* 1998; 19: 1345-1348.
4. Groan RJ, Middel B, Meilof JF, JB de Vos-van de Biezenbos, Enting RH, Coppes MH, Journée LH. Operative treatment of anterior thoracic spinal cord herniation: three new cases and an individual patient data meta-analysis of 126 case reports. *Neurosurgery* 2009; 64: 145-160.

-
5. Ghostine S, Baron EM, Perri B, Jacopson P, Morsette D, Hsu FP. Thoracic cord herniation through a dural defect: description of a case and review of the literature. *Surg Neurol* 2009; 71: 362-366.
 6. Hemant P, Paul P, Barunashish B, Dheeraj G. Imaging of idiopathic spinal cord herniation. *Radiographics* 2008; 28: 511-518.
 7. Kwong Y, Jakanani G, Rao N, Fang CSJ. MRI findings in herniation of spinal cord. *J Radiol Case Rep* 2010; 4: 1-5.
 8. Massicotte EM, Montanera W, Ross Fleming JF, Tucker WS, Willinsky R, TerBrugge K, Fehlings MG. Idiopathic spinal cord herniation: report of eight cases and review of the literature. *Spine* 2002; 27: 233-241.
 9. Miyaguchi M, Nakamura H, Shakudo M, Inoue Y, Yamano Y. Idiopathic spinal cord herniation associated with intervertebral disc extrusion. *Spine* 2001; 26: 1090-1094.
 10. Najjar MW, Baesa SS, Lingawi SS. Idiopathic spinal cord herniation: A new theory of pathogenesis. *Surg Neurol* 2004; 62: 161-171.
 11. Prasad A, Brar R, Sinha S, Rana S. Idiopathic spinal cord herniation. *Singapore Med J* 2013; 54: 43-45.
 12. Vallée B, Mercier P, Menei P, BouhourF, Fischer C, Fournier D, Bougeard R, Diabira S, Mahla K. Ventral transdural herniation of the thoracic spinal cord: surgical treatment in four cases and review of literature. *Acta Neurochir (Wien)* 1999; 141: 907-913.
 13. Wada E, Yonebu K, Kang J. Idiopathic spinal cord herniation: report of three cases and review of the literature. *Spine* 2000; 25: 1984-1988.
 14. White BD, Tsegage M. Idiopathic anterior spinal cord hernia: under-recognized cause of thoracic myelopathy. *Br J Neurosurg* 2004; 18: 246-249.
 15. Wortzman G, Tasker RR, Rewcastle NB, Richardson JC, Pearson FG. Spontaneous incarcerated herniation of the spinal cord into a vertebral body: a unique cause of paraplegia. Case report. *J Neurosurg* 1974; 41: 631-635.