



## IDIOPATHIC SPINAL CORD HERNIATION: A CASE REPORT AND LITERATURE REVIEW

Stéphano Santos  
Belisário

Federal University of Espírito Santo – UFES.

Natália Santos  
Belisário

### ABSTRACT

Through a case report and review of the literature of idiopathic spinal cord herniation, we intend to discuss this rare condition, that seems to be increasing with the accessibility of magnetic resonance imaging. We present the clinical and radiological aspects, as well as the differential diagnoses of this progressive myelopathy that can be treated, and for this reason must be recognized. Case description: 61-year-old male patient with thoracolumbar pain, with Brown-Sequard syndrome, urinary retention and difficulty in walking. He was submitted to magnetic resonance imaging of the thoracic spine, which evidenced findings suggestive of idiopathic spinal cord herniation. The most common clinical presentation of this condition is Brown-Sequard syndrome. Magnetic resonance imaging is the diagnostic method of choice. In the investigation of patients with progressive myelopathy, this condition should be considered as one of the differential diagnoses, especially in those with Brown-Sequard syndrome, with no history of trauma or surgery.

**KEYWORDS** : magnetic resonance imaging, medullary herniation, spinal cord, thoracic myelopathy.

### INTRODUCTION:

First described in 1974, Idiopathic spinal cord herniation is a rare clinical condition, rarely diagnosed until the advent of magnetic resonance imaging. After the popularization of this diagnostic method, there was an increase in its incidence. (1)

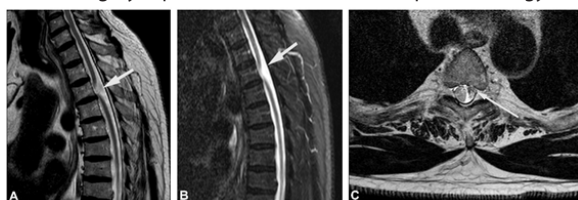
It is a progressive myelopathy that can be treated, and for this reason the recognition by the clinician is of great importance. (1,2) It is characterized by spinal cord herniation through a defect in the anterior portion of the dura mater of the thoracic segment, which may be the result of congenital dehiscence of the dura. (2)

### CASE REPORT:

A 61-year-old male, Caucasian, banker, attended the neurology clinic complaining of thoracolumbar pain that began 3 years ago, a constant, progressive worsening. He reported a decrease in muscle strength in the right lower limb, beginning 8 months ago, with progressive worsening. He complained of urinary retention and difficulty walking, which began 2 months ago.

It denied previous pathological antecedents, use of medications, trauma or surgeries.

Physical examination showed decreased muscle strength and deep sensitivity in the right lower limb, associated with hyperreflexia. In the lower left limb there was a decrease in the superficial sensitivity. He was submitted to a MRI of the thoracic spine (Figure 1), which evidenced left anterolateral deviation of the spinal cord at the level of the vertebral body of T3, with loss of the anterior cerebrospinal fluid at this level and presence of cerebrospinal fluid at the topography of the spinal cord. These findings were associated with an increase in the signal within the spinal cord at this level. No extradural masses or other structures were seen compressing the medulla later. The radiological evaluation was compatible with spinal cord herniation, which, within the context of absence of trauma, surgery or previous diseases, has an idiopathic etiology.



**Figure 1:** Magnetic resonance imaging of the thoracic spine. A - sagittal T2 image: anterior deviation of the spinal cord at the level of

T3, being noticed CSF in its usual topography. B - sagittal STIR image: anterior deviation of the spinal cord at the level of T3, being noticed CSF in its usual topography, emphasizing a slight alteration of intramedullary signal. C - axial T2 image at T3 level: absence of the spinal cord in the dural sac, with only a thin portion displaced anterolaterally to the left being visualized.

### DISCUSSION:

The primary pathophysiological factor for the development of idiopathic spinal cord herniation is the presence of a defect in the anterior portion of the dura, of unknown etiology, through which the spinal cord is insinuated (3). This failure may occur at more than one point and cause spinal cord herniation in more than one location (4). As a consequence, the herniated medullary segment undergoes secondary vascular injury, leading to progressive myelopathy (4).

This condition is most frequently described in the T4 and T7 segments, with adults more frequent in the fifth decade of life, especially women, in a ratio of 2: 1 (1). Clinical symptoms are progressively progressive, and the natural history of the disease is slow. In most cases, the diagnosis usually occurs in later stages (5). The most common clinical presentation is Brown-Sequard syndrome, occurring in about half of the cases. The involvement of the spinothalamic tract is the main cause of the initial clinical manifestations. There are also common in the evolution of the disease, gait disturbances and impairment of sphincter control. (5,6) Magnetic resonance imaging of the thoracic spine is the diagnostic method of choice (1,2). The evaluation of the images occurs mainly through those acquired in the sagittal and axial imaging planes. The sagittal images allow visualization of the anterior or anterolateral displacement of the medulla, represented by an anterior curvature of this in the form of "C" or "S", that establishes contact with the anterior dural surface. Also in the images obtained in this plane, it is also possible to observe the enlargement of the dorsal CSF, in the topography where the displaced marrow should be. In cases of longer evolution, there is still atrophy of the medullary segment involved (1,2). In T2-weighted MRI sequences, signal changes in the marrow can be observed, characterized by increased signal in the affected area. This finding is associated with a worse prognosis, since patients with this alteration tend not to show improvement of symptoms when submitted to surgical intervention (1,7).

As the main differential diagnosis, the posterior arachnoid cyst should be considered (2). This condition causes anterior marrow deviation, compressing it. In this way, it presents a similar aspect in

the evaluation through magnetic resonance. Extradural medullary compressions and disc herniations with dural retraction should also be considered as differential diagnoses (1).

The treatment is given through a surgical approach, as a way to reverse myelopathy (6). Even in patients with long-term neurological deficit, in most cases, neurosurgical intervention provides significant clinical improvement (8).

The purpose of surgery is to restore the anatomy, releasing the imprisoned spinal segment and closing the dura mater, either by direct suture or using a dura mater analogue (8).

#### **CONCLUSION:**

In the investigation of patients with progressive myelopathy, one of the differential diagnoses is idiopathic spinal cord herniation, especially in those with Brown-Sequard syndrome, with no history of trauma or surgery. Because it is a cause of reversible myelopathy, early recognition is extremely important in the prognosis of affected patients, since the appropriate surgical treatment may reverse the clinical course. Magnetic resonance imaging is the diagnostic method of choice and should be sought in patients with clinical suspicion.

#### **REFERENCES:**

1. Parmar H, Park P, Brahma B, Gandhi D. Imaging of idiopathic spinal cord herniation. *Radiographics*. 2008;28:511-8.
2. Watters MR, Stears JC, Osborn AG, et al. Transdural spinal cord herniation: imaging and clinical spectra. *AJNR Am J Neuroradiol* 1998;19:1345-8.
3. Darbar A, Krishnamurthy S, Holssaple J, Hodge C. Ventral thoracic spinal cord herniation: frequently misdiagnosed entity. *Spine* 2006;17:600-5.
4. Aydin AL, Sasani M, Erhan B, Sasani H, Ozcan S, Ozer AF. Idiopathic spinal cord herniation at two separate zones of the thoracic spine: the first reported case and literature review. *Spine J* 2011;11(8):e9-e14.
5. Berg-Johnsen J, Ildstad E, Kolstad F, Züchner M, Sundseth J. Idiopathic ventral spinal cord herniation: an increasingly recognized cause of thoracic myelopathy. *J Cent Nerv Syst Dis* 2014;6(6):85-91.
6. White BD, Tsegaye M. Idiopathic anterior spinal cord hernia: under-recognized cause of thoracic myelopathy. *Br J Neurosurg* 2004;18(3):246-249.
7. Imagama S, Matsuyama Y, Sakai Y, et al. Image classification of idiopathic spinal cord herniation based on symptom severity and surgical outcome: a multicenter study. *J Neurosurg Spine* 2009;11(3):310-319.
8. Chaichana KL, Sciubba DM, Li KW, Gokaslan ZL. Surgical management of thoracic spinal cord herniation: Technical considerations. *J Spinal Disord Tech* 2009;22:67-71.