
CASE REPORT

Idiopathic Spinal Cord Herniation

AS Tandon, D Saxena

Department of Radiology, St. John's Medical College Hospital, Sarjapur Road, Bangalore 560034, India

ABSTRACT

Idiopathic spinal cord herniation is a rare spinal cord disorder caused by spinal cord prolapse through a dural defect. Idiopathic spinal cord herniation usually occurs in the thoracic spine between the T4 and T7 vertebral levels. Neurological symptoms are slowly progressive and worsen over time, although timely diagnosis and treatment may enable reversal of symptoms. Clinical suspicion and magnetic resonance imaging lead to the diagnosis, with the latter demonstrating focal herniation of the spinal cord through an anterior dural defect. This report describes a patient with idiopathic spinal cord herniation diagnosed by magnetic resonance imaging and the literature about this rare entity.

Key Words: Hernia; Magnetic resonance imaging; Spinal cord diseases

中文摘要

特發性脊髓疝

AS Tandon, D Saxena

特發性脊髓疝是一種罕見的脊髓疾病。脊髓經硬膜缺損疝出，好發於胸椎T4至T7之間的節段。該病的神經系統症狀為緩慢進行性，隨時間推移惡化；及時的診斷和治療可使症狀逆轉。如磁共振成像顯示出經前方硬膜缺損而形成的脊髓疝病灶，則臨床高度懷疑加磁共振成像可明確診斷。本文報告一名磁共振成像診斷的特發性脊髓疝病例，並回顧及討論文獻所記載的病例。

INTRODUCTION

Idiopathic spinal cord herniation is a rare cause of slowly progressive myelopathy affecting middle-aged individuals. Since the first report by Wortzman et al in 1974,¹ 100 cases have been reported in the English-language literature.² Idiopathic spinal cord herniation usually affects the thoracic spinal cord between the T4 and T7 levels, and is caused by an anterior dural defect through which the thoracic spinal cord herniates. This leads to a thoracic cord syndrome, most commonly

the Brown-Séquard syndrome, which is curable if diagnosed early.³ A combination of clinical suspicion and magnetic resonance imaging (MRI) leads to the diagnosis. Awareness of the clinical syndrome and the characteristic imaging findings is essential for the diagnosis.

CASE REPORT

A 65-year-old man presented to the emergency department at St John's Medical College Hospital,

Correspondence: Dr Anisha S Tandon, Department of Radiology, St. John's Medical College Hospital, Sarjapur Road, Bangalore 560034, India.

Tel: (91) 080 2206 5196 / (91) 080 2206 5197 / (91) 080 2206 5198; Email: anisha.sawkar@gmail.com

Submitted: 2 Nov 2012; Accepted: 19 Dec 2012.

Bangalore, India, in October 2012, with an infected pressure ulcer on the plantar aspect of his right leg. Historical evaluation and clinical examination suggested a long-standing (3 years), slowly progressive lower-limb sensory deficit with paraparesis (power in the lower limbs was 3/5), and loss of pain and temperature sensations in his right leg. He had no previous history of trauma to explain the onset of symptoms, and no repeated neurological deficits or low back pain. Clinical examination excluded lumbar disc aetiology and there were no features to suggest the cauda equina syndrome. Laboratory tests were within normal limits. MRI of the spine was performed to assess for a compressive / non-compressive myelopathy. Sagittal T2-weighted images of the dorsal spine showed evidence of considerable focal anterior displacement of the thoracic spinal cord at the T2 level (Figure 1), with resultant widening of the posterior subarachnoid space (Figures 1 to 3). Axial T2-weighted images at the same level showed severe anterior displacement of the spinal cord, with the cord positioned in close proximity with the posterior aspect of the T2 vertebral body, which was worse on the left side (Figure 3). The posterior subarachnoid space at this

level and caudally appeared widened (Figures 3 and 4). There was no posterior disc bulge at the T4 level or above, and no intra- or extra-medullary lesions. A diagnosis of idiopathic spinal cord herniation was made and the patient was advised to undergo surgery. The patient refused surgical treatment and selected long-term physical rehabilitative therapy.



Figure 1. Sagittal T2-weighted magnetic resonance image shows ventral displacement of the spinal cord at the T2 level, with minimal bright signal in the cord (arrow).

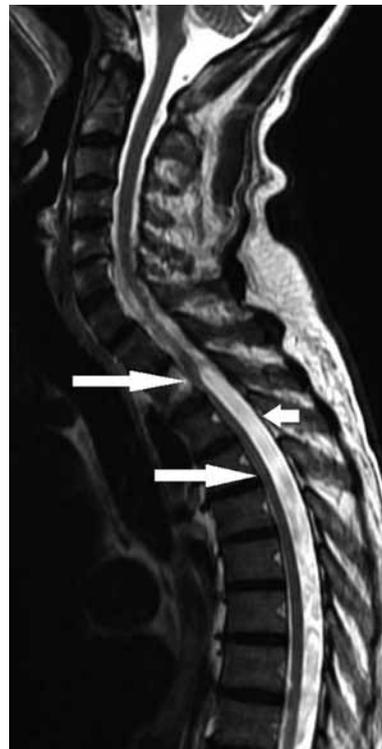


Figure 2. Sagittal T2-weighted magnetic resonance imaging shows the ventrally displaced spinal cord (long arrows) from the T2 level downwards, with a widened posterior subarachnoid space (short arrow).

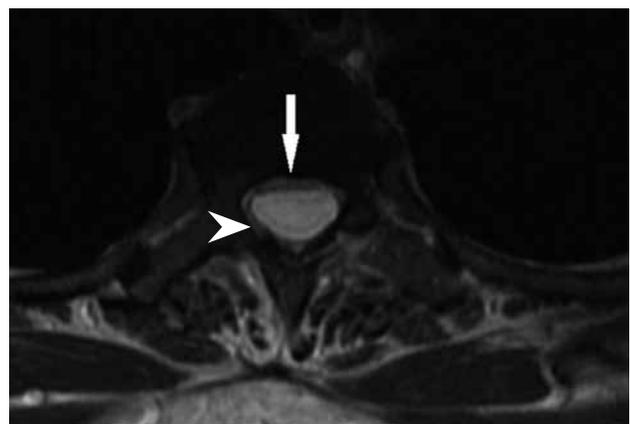


Figure 3. Axial T2-weighted magnetic resonance imaging at the T2 level shows the spinal cord displaced anteriorly and flattened against the posterior aspect of the vertebral body, slightly more to the left (arrow). A widened posterior subarachnoid space is noted (arrowhead).

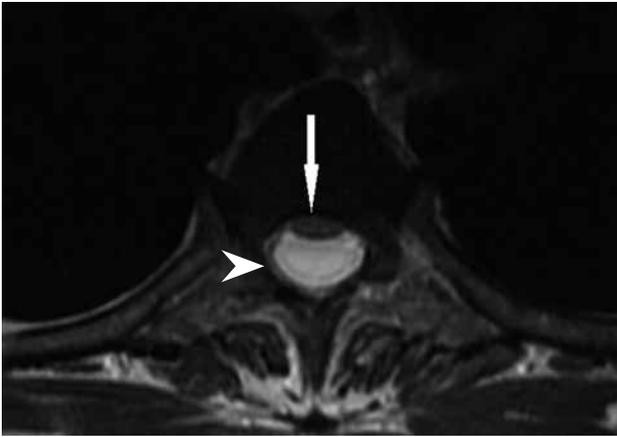


Figure 4. Axial T2-weighted magnetic resonance imaging at the T3 level shows the ventrally displaced spinal cord (arrow) with a widened posterior subarachnoid space (arrowhead).

DISCUSSION

Idiopathic spinal cord herniation is one of the unusual causes of thoracic myelopathy in which the thoracic spinal cord herniates through a dural defect leading to myelopathy due to kinking of the cord at the level of herniation.⁴ The age range of patients at presentation is 22 to 71 years (mean, 49 years).^{4,5} Thoracic spinal cord herniation is slightly more common among women than in men (male:female ratio = 2:3).^{1,4} The level of involvement is typically between the T4 and T7 vertebrae, a spinal segment that includes the area of physiological kyphosis.^{4,6}

As the name suggests, idiopathic spinal cord herniation has no definitive cause and, although the aetiopathogenesis of this disorder is unclear, various mechanisms have been postulated to explain the initial dural defect and subsequent cord herniation.^{4,6} The presence of clinically minor or occult trauma has been one of the mechanisms suggested for the occurrence of the dural defect.^{4,6} A thoracic level disc bulge which is calcified has been postulated to erode and tear the dura over a period of time and has been cited as being the aetiology.^{4,6} The presence of a calcific defect in the vertebral end plates adjacent to the site of spinal cord herniation has been described in some reports of idiopathic spinal cord herniation,⁶ and is probably related to posterior and interosseous disc herniation. The calcific defect produces an imaging appearance that has been described as a 'nuclear trail' sign.^{4,5,7} Some authors have postulated that a congenital weakness or anomaly of the dura mater may lead to cord herniation.^{4,8-10} Coughing has also been reported to have caused dural

tear in one patient.¹¹ The proposed theory is that after the dural defect has been established, cerebrospinal fluid (CSF) flows through the defect and enlarges it; the dentate ligaments anchoring the cord to the dura may tear and the ongoing CSF pulsations lead to herniation of the cord through the defect.^{4,6,7} The thoracic spine has certain unique features that predispose it to the occurrence of spinal cord herniation.^{4,6} These include limited range of motion of the thoracic spine and physiological kyphosis of the thoracic spine, which result in a wider thecal sac and more anterior positioning of the cord in the thoracic spine.^{4,12} Due to the anterior position of the cord, pulsatile CSF flow in the posterior subarachnoid space leads to greater anteroposterior kinetics of the spinal cord, thereby, pushing it into further contact with the ventral dural tear. This CSF pulsatility constitutes a likely mechanism for cord herniation.¹³

The most frequent clinical presentation in this condition may be the Brown-Séquard syndrome.^{4,7,13,14} The other early neurological deficits described include numbness, pain, and gait disturbances due to sensory ataxia.^{4,6,7} The neurological course is slowly progressive and the symptoms are long-lasting.^{4,15,16} The mainstay of diagnosis remains MRI and the classical abnormality seen on sagittal MR images is an acute anterior kink of the thoracic cord leading to widening of the dorsal subarachnoid space, which follows the signal intensity expected of normal CSF.^{4,13} The deviated cord is generally limited to one or two thoracic segments.^{4,6} There may be associated cord atrophy and myelomalacic changes in the spinal cord at the level of the herniation.^{4,6,7} In most instances, MRI is sufficient for diagnosis. The common differential diagnoses for this condition are a dorsally located intradural arachnoid cyst and a disc herniation with ventral cord tethering.⁴ In equivocal cases, conventional myelography followed immediately by computed tomography myelography may be performed.⁴ Phase-contrast pulse cine MRI, which demonstrates CSF flow in the subarachnoid space posterior to the herniated spinal cord, may also be helpful for excluding an associated posterior arachnoid cyst.^{4,13}

CONCLUSION

Idiopathic spinal cord herniation is a rare cause of slowly progressive myelopathy that most commonly affects the thoracic spine. Many causes for this pathology have been proposed and a clinical scenario of long-standing, slowly progressive myelopathy is

usually encountered. The condition is usually diagnosed on MRI. A high index of suspicion, together with awareness of the clinical and imaging findings, will lead to an accurate diagnosis.

REFERENCES

1. 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;41:631-5. [cross ref](#)
2. Sasani M, Ozer AF, Vural M, Sarioglu AC. Idiopathic spinal cord herniation: case report and review of literature. *J Spinal Cord Med.* 2009;32:86-94.
3. Kim JM, Oh SH, Kim KJ, Park SH, Park KS. Idiopathic spinal cord herniation as treatable cause of progressive brown-sequard syndrome. *J Clin Neurol.* 2007;3:204-7. [cross ref](#)
4. Parmar H, Park P, Brahma B, Gandhi D. Imaging of idiopathic spinal cord herniation. *Radiographics.* 2008;28:511-8. [cross ref](#)
5. Spissu A, Peltz MT, Matta G, Cannas A. Traumatic transdural spinal cord herniation and the nuclear trail sign: case report. *Neurol Sci.* 2004;25:151-3. [cross ref](#)
6. Tekkök IH. Spontaneous spinal cord herniation: case report and review of the literature. *Neurosurgery.* 2000;46:485-91. [cross ref](#)
7. Massicotte EM, Montanera W, Ross Fleming JF, Tucker WS, Willinsky R, TerBrugge K, et al. Idiopathic spinal cord herniation: report of eight cases and review of the literature. *Spine (Phila Pa 1976).* 2002;27:E233-41.
8. Watanabe M, Chiba K, Matsumoto M, Maruiwa H, Fujimura Y, Toyama Y. Surgical management of idiopathic spinal cord herniation: a review of nine cases treated by enlargement of the dural defect. *J Neurosurg.* 2001;95(2 Suppl):169-72.
9. Nakazawa H, Toyama Y, Satomi K, Fujimura Y, Hirabayashi K. Idiopathic spinal cord herniation. Report of two cases and review of the literature. *Spine (Phila Pa 1976).* 1993;18:2138-41. [cross ref](#)
10. Sugimoto T, Kasai Y, Takegami K, Morimoto R, Maeda M, Uchida A. A case of idiopathic spinal cord herniation with duplicated dura mater. *J Spinal Disord Tech.* 2005;18:106-11. [cross ref](#)
11. Lasater GM. Primary intracranial hypotension. The low spinal fluid pressure syndrome. *Headache.* 1970;10:63-6. [cross ref](#)
12. Borges LF, Zervas NT, Lehigh JR. Idiopathic spinal cord herniation: a treatable cause of the Brown-Sequard syndrome — case report. *Neurosurgery.* 1995;36:1028-32. [cross ref](#)
13. Brugières P, Malapert D, Adle-Biasette H, Fuerxer F, Djindjian M, Gaston A. Idiopathic spinal cord herniation: value of MR phase-contrast imaging. *AJNR Am J Neuroradiol.* 1999;20:935-9.
14. Miyake S, Tamaki N, Nagashima T, Kurata H, Eguchi T, Kimura H. Idiopathic spinal cord herniation. Report of two cases and literature review. *J Neurosurg.* 1998;88:331-5. [cross ref](#)
15. Isu T, Iizuka T, Iwasaki Y, Nagashima M, Akino M, Abe H. Spinal cord herniation associated with an intradural arachnoid cyst diagnosed by magnetic resonance imaging. *Neurosurgery.* 1991;29:137-9. [cross ref](#)
16. White BD, Tsegaye M. Idiopathic anterior spinal cord hernia: under-recognized cause of thoracic myelopathy. *Br J Neurosurg.* 2004;18:246-9. [cross ref](#)