

Idiopathic Spinal Cord Herniation

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ABSTRACT

History: A 54-year-old female presented with complaints of backache since 6 to 8 months, weakness in both lower limbs and difficulty in walking. She had past history of cervical and lumbar decompression surgery.

Imaging findings: On MRI, a focal kink was seen in the dorsal spinal cord at T8-T9 level. Anterior displacement of the spinal cord was also seen at this level with prominent posterior subarachnoid spaces. Along with other findings described, it was suggestive of idiopathic spinal cord herniation.

Management: Management strategy along with the etiopathogenesis and differential diagnosis is described in the text.

Conclusion: Despite the fact that MR imaging appearances of this entity are pathognomonic, missed diagnosis and misdiagnosis are common. Careful imaging study would avoid this.

Keywords: Idiopathic spinal cord herniation, Thoracic spine, MRI features.

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INTRODUCTION

Idiopathic spinal cord herniation (ISCH) is also known as spontaneous transdural spinal cord herniation. It is a rare cause of thoracic myelopathy. However, it has been diagnosed and reported with increasing frequency in recent years with improved availability of MR imaging and the increased awareness of this condition. Having it as a differential in mind can help in overlooking it as a possibility and also obviate the need of wrong surgery.

HISTORY

A 54-year-old female presented to the neurology clinic with complaints of backache since 6 to 8 months, weakness in both lower limbs and difficulty in walking. Rest of the neurological examination was normal. Patient had past history of cervical and lumbar decompression surgery.

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IMAGING FINDINGS

Multiphase plain and post contrast MRI of dorsal spine was performed using T1- and T2-weighted turbo spin echo sequences.

It showed a focal kink in the dorsal spinal cord at T8-T9 level. There was a focal anterior displacement of the spinal cord at this level with prominent posterior subarachnoid spaces. There was associated abnormal bright signal intensity within the spinal cord on T2-weighted images extending from T9-T11. There was no significant uptake of contrast. These imaging findings are suggestive of idiopathic spinal cord herniation (Figs 1 and 2).

DISCUSSION

Idiopathic spinal cord herniation is also known as spontaneous transdural spinal cord herniation. It is a rare cause of thoracic myelopathy. However, it has been diagnosed and reported with increasing frequency in recent years with improved availability of MR imaging and the increased awareness of this condition. Recognition of focal ventral displacement and angulation of the thoracic spinal cord, with or without cord thinning, on the images should lead one to consider this diagnosis (Fig. 3).

Najjar et al¹ reviewed the literature on this entity and identified 79 published reports of ISCH. The age range of patients with ISCH varies from 22 to 78 years, with a median age of 50 years.

There is a predilection for ISCH to occur in female patients (female-to-male ratio, 3 : 2).²



Fig. 1: T2-weighted sagittal image showing anterior displacement of the spinal cord at T8-T9 level with prominent posterior subarachnoid space



Fig. 2: T2-weighted axial image showing anterior displacement of the spinal cord at T8-T9 level with prominent posterior subarachnoid space

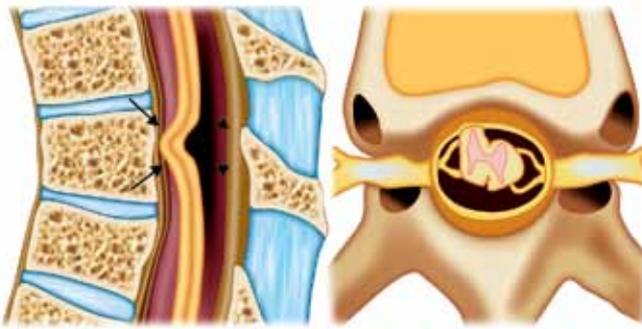


Fig. 3: Sagittal and axial images showing transdural herniation of spinal cord

Brown-Sequard syndrome is the most frequently described symptom of ISCH at presentation, occurring in more than half of the reported cases.³ Other symptoms at presentation include numbness and decreased temperature sensation in the legs, gait disturbances, pain and incontinence. The duration of symptoms is typically long and neurologic course is that of a slowly progressive dysfunction.

Imaging is a mainstay in the diagnosis of ISCH. On sagittal MR images, an anterior kink of the thoracic spinal cord is observed with an enlargement of the posterior subarachnoid space. Cord deviation is generally limited to less than two thoracic segments. The dural defect occurs in the thoracic spine, most commonly in between the T4 through T7 vertebral level. There may be associated cord atrophy at the level of herniation.

ETIOPATHOGENESIS

The cause of ISCH has not been firmly established, however a variety of mechanisms have been postulated. It has been suggested that close apposition of the thoracic cord to the anterior dura may be fundamental in the pathogenesis of this disease. Presence of clinically occult minor trauma or

a remote traumatic event has also been postulated as an inciting factor for dural tears. In a case report,⁴ intervertebral disk herniation was documented as a cause of dural defect. A few other authors have suggested an underlying congenital abnormality of the dura matter. Isu et al,⁵ in 1991, postulated that the ventral defect was caused by pressure transmitted from the cord from a dorsal arachnoid cyst and treated both patients reported by excising the arachnoid cyst with some sensory improvement.

DIFFERENTIAL DIAGNOSIS

Idiopathic spinal cord herniation has fairly characteristic imaging appearances, and a confident diagnosis often can be achieved on the basis of MR imaging alone. Recognition on MR images of focal ventral displacement and angulation of the thoracic spinal cord, with or without cord thinning, should lead to consideration of this diagnosis. However, it is important to exclude a dorsally located cystic lesion which may mimic a cord herniation. When a myelogram shows the uninterrupted flow of contrast material and the absence of a filling defect posterior to the herniated cord segment, an arachnoid cyst is excluded. Phase contrast MR imaging may be used to obtain similar information.⁶ Unfortunately despite the fact that MR imaging appearances of this entity are pathognomonic, missed diagnosis and misdiagnosis are common. The most commonly encountered erroneous radiologic diagnoses included intradural arachnoid cyst or extradural mass with cord compression, and disk herniation with cord tethering.³

MANAGEMENT

Most cases reported in the literature were treated surgically because of worsening symptoms. Surgical reduction of the herniated spinal cord may result in an improvement of symptoms even in patients with chronic neurologic dysfunction. However, in patients without a history of symptom progression, conservative management with regular monitoring may be more appropriate.⁷ Watanabe et al suggested that cord herniation occurs between the two layers of duplicated anterior dura; therefore, enlargement of the inner hiatus should be performed after surgical reduction of the cord has been accomplished. In eight of nine patients, neurologic symptoms resolved immediately after surgery.⁸

CONCLUSION

Despite the fact that MR imaging appearances of this entity are pathognomonic, missed diagnosis and misdiagnosis are common. Careful study and imaging in patients with spinal symptoms especially Brown Sequard syndrome might obviate many unnecessary surgeries in these patients.

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