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Idiopathic Spinal Cord Herniation: Case Report and Review of Literature

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ABSTRACT

Idiopathic Spinal cord herniation is a rare cause of myelopathy. However, this condition is been increasingly recognized. We describe a case report with a supplemental intraoperative video and review the pertinent literature.

Keywords: Spinal cord herniation, Brown-Sequard syndrome, Review

INTRODUCTION

Idiopathic spinal cord herniation (ISCH) is an extremely rare cause of myelopathy typically affecting the thoracic spinal cord. This condition is characterized by the herniation of spinal cord through an anterior or lateral dural defect. Wortzman first described this condition in 1974 [1]. It most commonly presents in middle aged women. Brown-Sequard syndrome is the most common mode of presentation [2]. The exact etiology of this disease is still unknown. Inspite of the published reports in the past, awareness of this condition is still low. We describe a case of spinal cord herniation and review the pertinent literature.

Case Report

A 53 year lady presented with history of tingling and numbness involving left lower limb, weakness of lower limbs which was more pronounced on the right side and gait instability. She reported a gradual worsening of symptoms since two years, which had become more conspicuous in last six months and needed a stick to ambulate. She also noticed that she was unable to perceive any temperature sensation in her left lower extremity but never had any bowel or bladder irregularity. She had no other co morbidity or prior surgery. On examination there was decreased tactile and pin prick sensation and reduced temperature perception in left lower limb. The joint position and vibration sense was impaired in bilateral lower limbs. Weakness in right lower limbs was more at distal extremity. The knee and ankle reflexes were diminished bilaterally. The neurological evaluation was suggestive of a Brown-Sequard syndrome at the level of T4. MRI of the dorsal spine suggested an anterior displacement of the spinal cord with a characteristic kink (**Figure 1**).

The herniated spinal cord was seen in the axial MRI. With the patient in prone position, using motor evoked potential (MEP) and somatosensory evoked potential (SSEP) guidance, a T3-5 laminectomy was done and a midline durotomy was performed and the dentate ligaments were sectioned. The anterior displacement of the spinal cord was visualized and the defect in the anterior dura was identified. The defect was increased in size to release the herniated spinal cord from the adhesions and the herniated cord was gently reduced(**Figure 2.**). All these maneuvers were guided by intraoperative electrophysiological monitoring. After ensuring complete reduction of the ventrally herniated cord at T4 level, the defect in anterior dura was carefully delineated and repaired with autologous fascia (onlay patch). The cord was slightly atrophied and the subdural space was capacious, hence the dura was closed primarily **(Video 1.)**. The postoperative course was uneventful and the patient improved symptomatically within 3 weeks of surgery. With continuing physiotherapy and gait training her walk improved significantly. At 3 months, she could walk around with very minimal support with significant improvement in her sensory perception. Her postop MRI showed complete reduction of the herniated cord (**Figure 1**).

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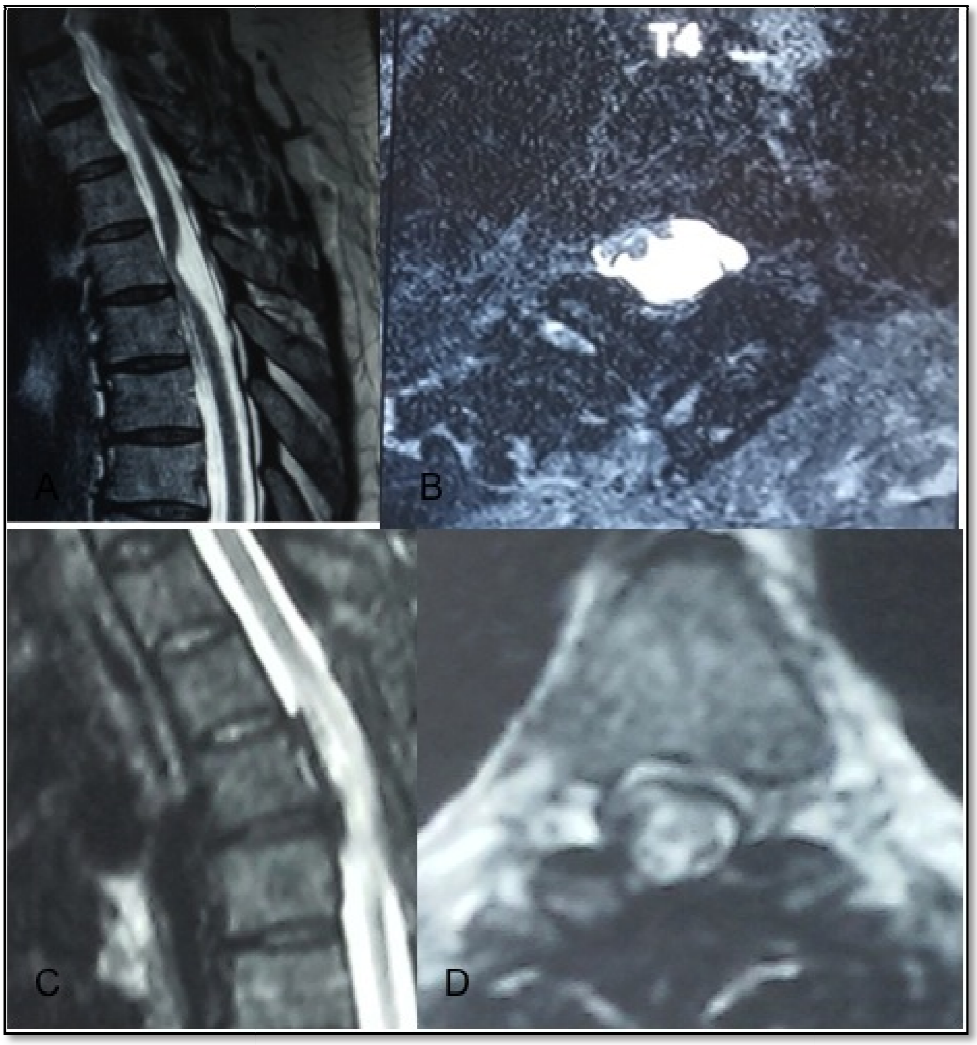


Figure 1 . Sagittal MRI (A) showing the characteristic ventral kink with enlarged dorsal subarachnoid space. Axial MRI (B) showing herniation of the spinal cord anterolaterally and complete obliteration of anterior subarachnoid space. Postoperative MRI (C) and (D) at 3 months shows the graft in situ with complete reduction of the previously herniated spinal cord.

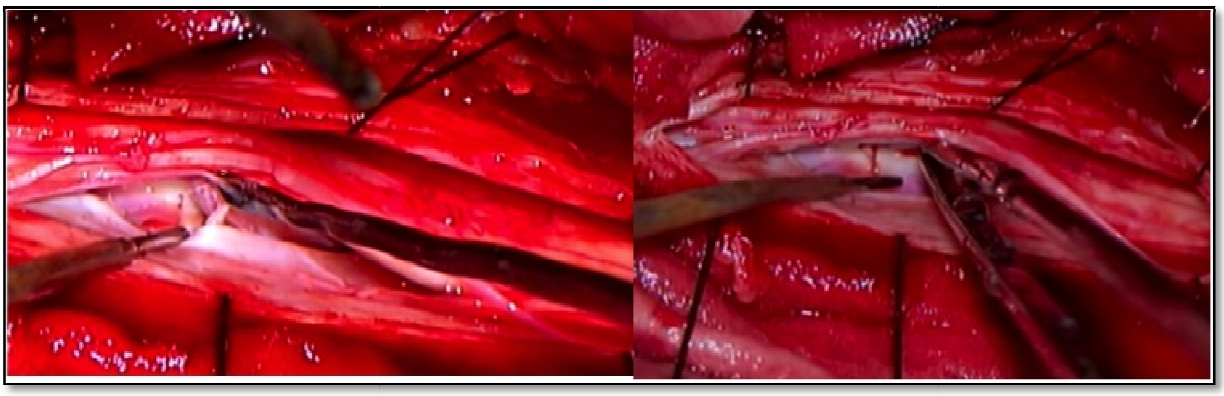


Figure 2. Intraoperative images show the initial arachnoid dissection of the herniated spinal cord and dural defect (A), followed by complete reduction of the herniated cord and delineation of the entire dural defect (B).

DISCUSSION

1. Pathogenesis

Presence of a dural defect is the pathognomic feature of this condition. There are three theories regarding the pathogenesis of this dural defect. (**Table 1.)** Aizawa and coworkers described 3 types of ventral dural defect: (a) a defect in the inner layer of the duplicated ventral dura; herniated spinal cord is covered by an outer layer of dura (b) direct epidural spinal cord herniation through a full-thickness dural defect and (c) an epidural cyst or pseudomeningocele [11].

Irrespective of the reason of dural defect, it is believed that the spinal cord attempts to plug the defect initially and eventually spinal cord herniation occurs due to the CSF pulsations. This is followed by adhesions at the site of defect, which "tethers" the cord and causes vascular compromise and subsequent myelopathy. The limited motion in the thoracic spine, the anterior position of the thoracic cord, the physiological thoracic kyphosis and the physiological anterior motion of the spinal cord with cardiac and pulmonary movement may facilitate this process [12].

Table 1. Proposed causes of ISCH

Underlying cause	Mechanism
1. Congenital	Congenital defect of dura matter. Some cases have reported spinal cord herniation between the two layers of dura, probably due to duplication of dura. Absence or defect of one of the dentate ligaments has also been explained as facilitatory for this condition. [3-6]
2. Trauma, Inflammation	Remote injuries may be a possible cause. Disc herniation and calcification is another postulated cause of the dural defect. [7,8,9]
3. Dorsal arachnoid or pseudocyst	Dorsal mass may push the cord anteriorly. (Least accepted) [10]

2. Imaging

MRI is the gold standard technique for diagnosis of ISCH. However misdiagnosis is extremely common and has been reported in 21% of cases in literature [13]. The most common "cause" of misdiagnosis was a dorsal arachnoid cyst.

In MRI, typical features of ISCH [14] are as follows

1. Ventral displacement,

2. Sharp ventral angulation of thoracic spinal cord,

3. Enlargement of dorsal subarachnoid space

4. Attachment of the herniated spinal cord to the ventral dura with complete obliteration of intervening CSF space.

ISCH has been classified based on sagittal imaging into three types according to the severity of herniation and displacement: Type K (kink), showing a ventral spinal cord kink; Type D (discontinuous), showing complete spinal cord disappearance at the herniated site; and Type P (protrusion), showing obliteration of anterior subarachnoid space no or minimal kink [15]. These types may represent different stages in the development of ISCH. Phase-contrast MRI can be crucial in excluding a posterior compressing arachnoid cyst and may replace computed tomographic myelography [14]. In ISCH, the spinal cord is frequently shifted ventrolaterally and sometimes rotated toward the side of tethering. Tethering of the spinal cord at the side of the herniation results in unilateral damage of the lateral funiculus and might cause symptoms of Brown-Sequard syndrome. Occasionally bone defect due to the ISCH has also been reported [15]. Some of the other atypical signs include the nuclear trail sign, presence of T2 hyperintensity in the cord or the presence of a syrinx [16].

3. Management

Surgery is the treatment of choice for reversing or stopping the progression of serious neurologic deficits. Intraoperative electrophysiology is an important adjunct. A combination of MEP and SSEP is usually preferred for these cases [2]. Laminectomy with sectioning of the dentate ligaments and gentle rotation of the cord is usually adequate to visualize the herniation. Occasionally these maneuvers may not suffice and in those cases, posterolateral bone removal (facetectomy or costotransversectomy or transpedicular approach)may be performed to performed to gain access anteriorly or anterolaterally [12]. This additional bone removal may also be necessary in cases with persistent intraoperative electrophysiological changes.

There are 3 main treatment strategies to manage the dural defect:

(a) Primary closure of dural defect

Wortzman first performed primary suturing of the dural defect in 1974 [1]. However there is not enough space to pass the needle for primary suture. This procedure increases the risk of spinal cord damage.

(b) Duraplasty after reducing the spinal cord

Duraplasty is a more commonly used technique first described by Masuzawa and coworkers in 1981 [17].Some of dural substitutes which were used are muscle fascial flap, fat, lyophilized dura, and gortex membrane [18]. Some authors have performed duraplasty, even when there is evidence of dural duplication [19]. Arts et al. have proposed the use of a dural graft to wrap the spinal cord and thus to prevent future re-herniation [20].

(c) Enlargement of the dural defect to prevent cord strangulation.

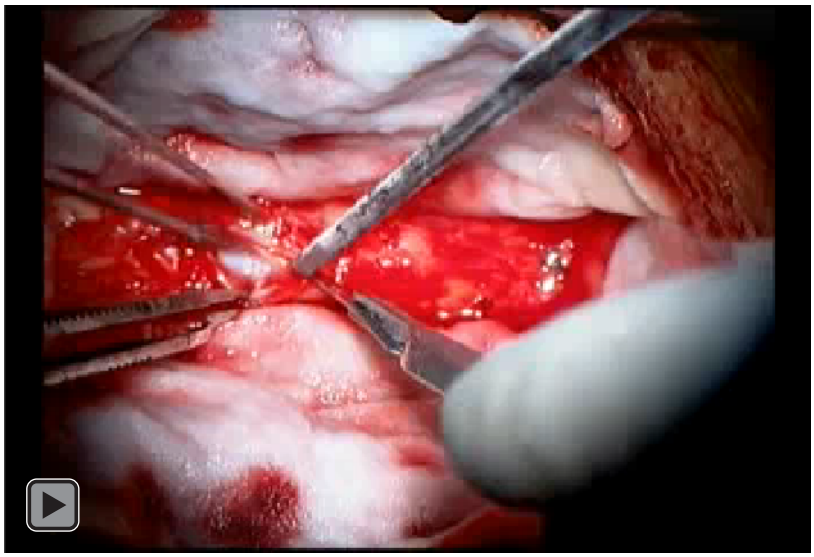
The technique was first described by Nakazawa and was performed by the authors, who proposed "dural duplication" as the cause of spinal cord herniation [3]. It must be emphasized that reduction of the spinal cord should be performed first before enlargement of dural defect is carried out to prevent neurological deterioration [6]. This technique is easier and requires less traction of the spinal cord than repair of the dural defect. However, anterior cerebral spinal fluid collection is a complication after this procedure, though they are rarely symptomatic.

Some authors in the past have also performed a biopsy of the herniated cord with a suspicion of a malignancy due to the reddish appearance of the cord [21, 22]. Postoperative syrinx has also been reported in two patients out of seven patients in a caseseries. These authors have therefore recommended an expansile posterior duraplasty to ensure circulation of the CSF [12]. Recurrence of herniation has also been reported infrequently [23].

There are a total of 15 cases reported in literature where the patients have been treated in a non-operative manner [13]. Conservative management may be possible in patients with mild or non-progressive neurological deficits. These cases have shown stabilization of their neurological deficits without any improvement or worsening [13].

4. Predictors of outcome

The outcomes for patients who initially presented with Brown-Séquard syndrome were better than for patients with spastic disorders (paralysis, paraparesis, and monoparesis) [2]. Groen et al in their meta analysis also reported that the patients undergoing release of the spinal cord and enlargement of the dural defect had the best outcome in terms of motor function [2]. Barbagallo et al stated that outcome was less favorable for spinal cord herniation at a vertebral body level than for disk-level herniation [24]. Presence of bony defects and T2 hyperintensities in the preop imaging has also been suggested as a predictive factor for severe preoperative symptoms as well as poor postoperative outcomes [21].In a review of 159 patients, it was reported that 74% improved after surgery, 18% had stabilization of their neurological deficits and 8% deteriorated after surgery [13]. Recurrence of herniation has been reported in a patient even after 10 years of surgery and hence long-term surveillance is recommended [25].



Video 1. Idiopathic Spinal Cord Herniation: Intra-operative Video

CONCLUSION

ISCH is a rare cause of myelopathy, which is being increasingly recognized with advances in imaging and improved awareness. Reduction of herniation and duraplasty to repair the dural defect is a safe and effective method to improve the neurological status in patients with Brown-Sequard syndrome.

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