Idiopathic ventral spinal cord herniation: a rare presentation of tethered cord

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Idiopathic ventral spinal cord herniation is a rare condition that has been increasingly reported in the last decade. The natural history and optimal management have yet to be defined. Therefore, debate exists regarding the pathogenesis and surgical management of this condition. The purpose of this review article is to further educate neurosurgeons about the surgical techniques and outcomes associated with treating this rare and often misdiagnosed condition.

KEY WORDS • idiopathic spinal cord herniation • tethered spinal cord • thoracic spine

Idiopathic ventral spinal cord herniation is a rare condition that primarily affects the thoracic spinal cord. Originally described by Wortzman et al. in 1974 after these authors discovered the spinal cord protruding through a dural defect when performing a thoracotomy for disc herniation, 128 cases have since been reported. With increased awareness among physicians and the widespread availability of MR imaging, more cases have been identified in the last decade. Despite this increase, many questions still exist regarding the pathogenesis, natural history, and surgical outcomes. Without a clear understanding of the natural history of this condition, the optimal management strategy for these patients remains controversial.

Although the origin of the dural defect is unknown, many theories have been postulated over the years, largely based on observations made during surgery. Variations in the type of dural defects include a full-thickness dural defect, a defect in the inner layer of duplicated dura mater, and an epidural cyst or pseudomeningocele. It has been proposed that the ventral dural defect causes a tethering of the spinal cord that progressively worsens as a result of persistent CSF pulsations and negative epidural pressures. This tethering results in progressive neurological symptoms, which are often misdiagnosed.

In this article, we review the clinical presentation, pathogenesis, radiographic imaging characteristics, and surgical results in patients with this rare condition.

Clinical Presentation

Patients usually present with progressive myelopathy and signs of either Brown-Séquard syndrome or spastic paraparesis months to years prior to surgical intervention. The interval between initial onset of symptoms and surgical intervention ranges from 6 months to 36 years, with a mean of 5.2 years. Approximately 66% of reported cases present with Brown-Séquard syndrome, 30% with symmetrical spastic paraparesis, 4% with pure sensory deficits, and 1% with isolated motor deficits. Bowel and bladder dysfunction are reported in 7%. The age of patients affected is between 21 and 78 years, with a mean age of 51 years. Twice as many women are affected as men.

Pathogenesis

Most idiopathic spinal cord herniations occur ventrally in the thoracic spine. Although many theories exist, the pathogenesis of this condition is unknown. It has been hypothesized that the thoracic spine is primarily involved, due to the normal kyphosis of the thoracic spine, the ventral position of the spinal cord within the midthoracic region, the spinal cord’s physiological ventral motion due to cardiac pulsations, and the biomechanical impact of flexion and extension movements on the thoracic spinal cord. Considering these factors, idiopathic herniations often develop through ventral or ventrolateral dural defects in the upper or midthoracic region. A review of the literature reveals that T4–5 is the most common level. A ventral dural defect, either congenital or acquired, is the proposed basis for this entity.

Congenital Defect

Some authors have proposed the possibility of a congenital dural defect that allows for gradual herniation of
the spinal cord over time. Theories about this congenital defect include a preexisting ventral pseudomeningocele, a meningeal diverticulum, or an extradural arachnoid cyst. Both Wortzman et al. and Masuzawa et al. suggested that herniation results from a congenital ventral meningocele. No other congenital bony spinal or neural deformities were reported in these cases. Some authors have questioned how a congenital defect can lead to symptoms in late adulthood.

Another theory advocated by some authors is congenital duplication of the ventral dura mater, with a defect in the inner layer. Nakazawa et al. suggested that a cavity caused by CSF pulsations forms between the layers of the dura, leading to herniation of the spinal cord and strangulation at the neck of the hernia. Opponents of this theory question the mechanism for disruption of this inner dural layer and the reason for fenestration.

This theory is based on operative findings and was previously challenged for lack of radiological and pathological evidence. However, in 2008 Ishida et al. reported a case in which the duplicated dura ventral to the spinal cord was identified prior to surgery by using high-resolution 3D MR imaging. In a recent review of the literature, 16 of 89 cases reportedly had duplication of the dura. Duplication was always found to be ventral to the spinal cord, with the defect consistently in the inner layer. Pathological examination of the inner layer of dura mater was performed in 6 cases, showing similar histological findings to that of dorsal dura, without any evidence of inflammation.

Dorsal arachnoid cysts are also associated with ventral spinal cord herniation. Isu et al. hypothesized that the pressure of the dorsal arachnoid cyst causes thinning of the ventral dura until a tear occurs. The spinal cord then comes in contact with the dural defect, and CSF pulsations over time result in herniation. Resection of the cyst alone, however, does not result in spinal cord untethering or improvement of symptoms. Patients in whom cyst resection was performed initially have required additional surgery to untether the cord and to prevent further neurological decline.

### Acquired Defect

The proposed pathogenesis of acquired defects includes damage to the ventral dura mater by inflammation, remote spinal trauma, vertebral body defect, and thoracic disc herniation. Hausmann and Moseley theorized that in some cases, thoracic disc herniation may damage the ventral dura, allowing the cord to herniate through the defect. Several other authors have also described thoracic disc herniation producing an adjacent dural defect, with subsequent cord herniation at the level of the vertebral body and not at the disc level. These patients are thought to have a worse prognosis.

Erosion of the dura by a herniated and calcified disc has been observed just caudal to the level of spinal cord tethering. Although the theory of a thoracic disc causing ventral dural weakening with subsequent cord herniation is described often, the case reported by Miyaguchi et al. is the only one in the literature in which a herniated disc was visualized through the ventral dural defect at surgery. It is proposed that the thoracic disc injures the ventral dural surface with repeated flexion and extension, and predisposes it to further damage.

### Pathophysiological Features of Tethering

Borges et al. theorized that tethering of the spinal cord on the site of the herniation results in unilateral damage of the lateral funiculus, and that symptoms of spinal cord herniation are a result of tension on this structure. The lateral position of the lateral spinothalamic tract within the lateral funiculus predisposes its axons to dysfunction secondary to tension induced by the tethering. As the tension progresses, the corticospinal tracts become involved, resulting in progressive weakness and spasticity.

Another theory regarding the progressive nature of symptoms is ischemia due to distortion or involvement of anterior spinal vessels due to tethering. This vascular mechanism may also explain the lack of improvement in some patients following untethering of the spinal cord.

### Diagnostic Imaging

Magnetic resonance imaging often demonstrates ventral displacement of the thoracic spinal cord and enlargement of the dorsal subarachnoid space. A ventral C-shaped kink is typically seen on sagittal imaging (Fig. 1). However, this condition is often misdiagnosed and misinterpreted as a dorsal arachnoid cyst. Atrophy of the spinal cord with signal change can also be seen, potentially confusing this condition for astrocytoma, disc herniation, extradural compression, and transverse myelitis.

Misdiagnosis may lead to erroneous surgery for decompression and resection, with no clinical improvement. Several authors have reported clinical worsening following biopsy of the herniated spinal cord, which may appear abnormal compared with normal spinal cord tissue.

Computed tomography myelography has been used for better evaluation of the relationship of CSF to the spinal cord, particularly in differentiating the widened dorsal subarachnoid space with a dorsal arachnoid cyst. In cases of idiopathic ventral herniation, CT myelography demonstrates no filling defect dorsal to the spinal cord or retention of contrast agent ventral to the dura mater. In contrast, arachnoid cysts demonstrate an intradural filling defect (Fig. 2).

The MR imaging signal intensity of arachnoid cysts is similar to that of CSF on both T1- and T2-weighted images, and the cyst walls are usually not visible. Phase-contrast MR imaging may help establish the presence or absence of a dorsal arachnoid cyst by showing reduced CSF pulsations in the cyst. This modality is also valuable in evaluating absence of pulsatile flow ventral to the spinal cord at the level of the herniation. This absence is another valuable sign of tethered cord.

Untethering of the spinal cord is clearly shown on postoperative imaging, with restoration of spinal cord
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Fig. 1. Preoperative MR imaging studies obtained in the thoracic spine of a 71-year-old woman who presented with Brown-Séquard syndrome and progressive myelopathy. Left: A sagittal T2-weighted image demonstrates a ventral spinal cord herniation at the level of T8–9. Right: An axial T2-weighted image shows ventral displacement of the thoracic spinal cord and a widened dorsal subarachnoid space.

Fig. 2. Preoperative CT myelography studies of the same patient showing no contrast ventral to the spinal cord at T8–9. Left: Sagittal image demonstrating contrast enhancement dorsal to the herniated spinal cord. This makes a diagnosis of arachnoid cyst less likely. Right: An axial image demonstrating the ventral herniation at the level of the disc space.

Fig. 3. Postoperative MR imaging study of the thoracic spine demonstrating realignment of the spinal cord within the canal and restoration of CSF ventral to the spinal cord. The patient showed neurological improvement at the 2-year follow-up.

alignment and evidence of CSF ventral to the cord (Fig. 3).

Surgery and Techniques

The optimal treatment strategy for idiopathic ventral spinal cord herniation has not yet been determined because the natural history of this rare condition is still unclear. In general, treatment consists of either conservative management or surgery. Conservative management has been proposed for patients without significant motor deficits or progressive myelopathy. Most reported series are surgical case discussions, and only a few authors have presented results of conservative management in these patients.

In one of the largest series, Massicotte et al. reported on 8 patients, 4 of whom did not have surgery. These 4 patients were observed for up to 8 years after presentation and none developed progressive neurological symptoms. In each of these nonsurgical patients, neither weakness nor spasticity were evident at presentation. Similarly, Ammar et al. reported on 2 patients who presented without weakness or spasticity and were followed for 15 months in one and 5 years in the other, without progression of symptoms. Senturk et al. followed a 38-year-old woman with progressive thoracic back pain without neurological deficits for 6 months without progression of symptoms. Hausmann and Moseley reported on 2 patients who presented with spasticity and weakness, but did not undergo surgery. No further follow-up information was provided. Given the lack of long-term follow-up, the classification of patients who would benefit from nonsurgical management is not yet known.

Surgery is typically recommended for patients with motor deficits or progressive neurological symptoms. Both dorsal and ventral surgical approaches have been used to treat ventral spinal cord herniations. Only Borges et al. and Wortzman et al. have used a ventral approach to treat these lesions. All other reports have used a strictly dorsal thoracic approach with laminectomy. Although the dorsal approach allows for a wide exposure of the spinal cord, it often does not provide adequate ventral exposure to untether the spinal cord and repair the dural defect safely. To avoid retracting the spinal cord, the dentate ligaments and arachnoid adhesions are usually released to mobilize it.

To obtain greater exposure of the ventral dura, costotransversectomy and transpedicular approaches have also been described. Gwinn and Henderson reported on 3 patients who underwent a unilateral posterolateral transpedicular approach for repair of the dural defect.
with bovine pericardia. All patients were clinically improved at 3 months, without recurrence of tethering. In a modification of the transpedicular approach, Chaichana et al.9 reported a bilateral transpedicular approach followed by unilateral removal of a pedicle and transverse process to provide a sufficiently wide bilateral exposure of the ventral dural defect. A Gore-Tex graft was sutured to the ventral dura for repair of the dural defect. At 6-week follow-up, the patient was improved clinically, with no radiographic evidence of ventral herniation.

Although techniques may differ among authors, release of the tethered spinal cord and repositioning of the cord to a normal anatomical position is generally agreed on as the main goal of surgery. To prevent reherniation, several different strategies have been advocated. These include enlargement of the ventral dural defect,2,25,30,31,39,44,45 primary closure of the defect with sutures,1,18,22 and insertion of a ventral patch for duraplasty.4,6,11,12,23

In a recent literature review, Saito et al.33 found that approximately 20% of patients with primary ventral dural closure worsened clinically after surgery, compared with 10% of patients with dural patching or widening of the dural defect. This difference is attributed to the difficulty of suturing the ventral dura from a strictly dorsal approach without excessively retracting the spinal cord. Many authors now prefer using a dural patch to seal the defect, because it minimizes spinal cord manipulation and requires less direct visualization.

Duraplasty was first described by Masuzawa et al.24 for a case in which a fascial flap was sutured to the ventral dural opening. Subsequently, a number of different grafting materials have been used to obliterate the defect, including muscle, fascia, fat, bovine pericardium, Teflon, and Gore-Tex.2,3,5,12,15,23,27,28,35,38,48

Because of the sporadic nature of this condition and limited long-term data, there is no evidence demonstrating superiority of one particular graft material or operative strategy in preventing the recurrence of tethering.

Clinical Outcomes

Because the number of surgically treated patients is limited, there is little evidence to predict which ones would benefit from surgery and when it would be most effective. Relying on case reports, which have significant variability in patient symptomatology and time to presentation, extensive reviews of the literature have only been able to make observations regarding the experience of surgeons to date.10,14,24 In general, surgery is usually followed by stabilization or improvement in neurological symptoms, but postoperative worsening has been reported. In one of the largest published series,16 10 patients were followed for 6–110 months after surgery. Four patients showed motor improvement, 2 had pain relief, and 1 experienced improvement in sensory function. Three patients were unchanged overall.

Although most patients improve neurologically after surgery, its long-term benefits are not clear. The range of follow-up is variable in reported cases, from weeks to years. In the longest reported follow-up, Selviaridis et al.26 reported a recurrence of ventral spinal cord herniation and clinical myelopathy after 10 years. The patient initially improved neurologically after the first surgery, then developed progressive paraparesis and urinary incontinence years later. During the first surgery, Surgicel was packed into the dural defect after untethering. On repeat surgery, a larger segment of the spinal cord was found to be herniated through a larger dural defect. Other cases of recurrence have been reported, ranging from 18 months to 3 years after surgery.2,29

Overall, patients tend to improve after surgery. Numerous case reports have demonstrated improvement in clinical outcomes for surgically treated patients. In a recent review of the literature, 73% of all patients improved, 20% experienced no change, and 7% deteriorated after surgery.24

When examined for differences based on presenting neurological signs, the outcomes for patients who initially presented with Brown-Séquard syndrome were better than for patients with spastic paraparesis. Improvement was reported in 56 (76.7%) of 73 patients with Brown-Séquard, and in 9 (47.3%) of 19 with spasticity.24

In the most comprehensive review of the literature, Groen et al.14 performed a meta-analysis to identify factors that affect postoperative outcome. In their analysis, Brown-Séquard syndrome and release of the herniated spinal cord were independent factors associated with favorable postoperative outcomes. With regard to management of the ventral dural defect, widening of the defect was associated with the highest prevalence of postoperative motor function improvement when compared with the application of a ventral dural patch. Persistent ventral displacement or realignment of the spinal cord on postoperative MR imaging was not found to correlate with outcome. However, only 56% of reported patients underwent postoperative imaging. Despite the meta-analysis, it is evident that the management of this condition needs to be individualized for each patient, because definitive conclusions cannot be made from a heterogeneous sampling of case reports.

Conclusions

Idiopathic ventral spinal cord herniation and tethering is a treatable cause of myelopathy that is often misdiagnosed. An MR imaging study that shows obliteration of the ventral CSF space with ventral focal deformity of the cord in the thoracic region raises a high index of suspicion for this condition. Surgical correction involves untying the spinal cord and repairing the dural defect. Surgery may lead to clinical improvement in select cases, despite a long history of myelopathy.

Disclosure

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

Author contributions to the study and manuscript preparation include the following. Conception and design: both authors. Acquisition of data: both authors. Analysis and interpretation of data: both authors.
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authors. Drafting the article: both authors. Critically revising the article: both authors. Reviewed final version of the manuscript and approved it for submission: both authors. Administrative/technical/material support: Krishnaney. Study supervision: Krishnaney.

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