Idiopathic ventral thoracic spinal cord herniation

- A successful surgical management of spinal cord herniation in a patient with old thoracic spine fracture: a case report from Syria
- Endoscopically Assisted Release Surgery for Idiopathic Spinal Cord Herniation: Technical Case Instruction
- Atypical presentation of arachnoid web causing paraparesis with absence of sensory symptoms
- A case of conservatively managed idiopathic spinal cord herniation presenting with low-pressure headache
- Early detection of idiopathic thoracic ventral spinal cord herniation by using imaging: A case report
- Case report: A vertebral bone spur as an etiology for spinal cord herniation: case presentation, surgical technique, and review of the literature
- Idiopathic Ventral Spinal Cord Hernia-A Single-Center Case Series of 11 Patients
- Idiopathic spinal cord herniation at the cervicothoracic junction level presenting with unilateral sensory symptoms

Epidemiology

Idiopathic ventral thoracic spinal cord herniation is a rare condition that usually presents with progressive myelopathy or Brown Séquard syndrome.

Pathology

The underlying cause of ventral cord herniation is thought to be a dural defect allowing the subarachnoid space to communicate with the extradural space, which can be congenital or acquired.

The thoracic cord, naturally closely applied to the ventral dura due to the normal thoracic kyphosis then 'plugs' the hole and gradually herniates through the defect. It is this normal anatomical relationship between the ventral theca and anterior thoracic cord which accounts for this entity only being encountered in the anterior aspect of the mid to upper thoracic spine. The distortion of the cord parenchyma, formation of adhesions and possible vascular compromise in turn leads to myelopathy and neurological dysfunction [1].

Anterior displacement of the thoracic spinal cord should elicit consideration of herniation to prevent misdiagnosis and inadequate surgery. Surgical cord release and enlargement of the dural defect are safe and associated with good clinical outcomes [2].

Radiographic features

The key finding is focal distortion of the cord with a focal defect in the dura, and localised cord signal change. Although both CT and conventional myelography are able to visualise the distortion, MRI is

the modality of choice to assess cord signal change and to evaluate for the differential diagnoses of spinal arachnoid cyst and dorsal thoracic arachnoid web.

Ventral cord herniation is only encountered between T2 and T8 where the normal thoracic kyphosis leads to the thoracic cord being in close proximity to the ventral theca.

The key feature is focal distortion and rotation of the cord with no CSF seen between it and the ventral theca. In most instances the cord is seen to bulge beyond the confines of the theca and is associated with T2 signal abnormality at that level.

Small extradural CSF intensity collection may also be seen, thought to represent the bulging CSF-filled arachnoid layer.

Differential diagnosis

The key differential diagnoses include:

spinal arachnoid cyst

smooth indentation of the cord rather than a very focal distortion

altered CSF flow may be visible (on T2 or CSF flow studies)

myelography usually demonstrates cyst to fill slower than surrounding CSF

intact dura, possibly with CSF seen between the cord and the anterior theca

dorsal thoracic arachnoid web

scalpel sign due to focal compression from behind

intact dura, possibly with CSF seen between the cord and the anterior theca

Depending on the amount of signal abnormality within the cord, other causes of intramedullary cysts/abnormal signal should also be considered and contrast is important to exclude an intramedullary mass (e.g. ependymoma).

Treatment and prognosis

It requires a lateral or anterolateral approach to minimize spinal cord manipulation. The dural defect is widened which usually results in a reduction of the spinal cord herniation. A sling of dural substitute can then be slid anterior to the cord to prevent reherniation.

Surgery with division of adhesions and closure of the dural defect, which may require a dural graft/duroplasty, is curative. In most cases symptoms improve, however depending on the degree of pre-operative myelopathy complete recovery may not occur

Prominent T2 signal change within the cord may be a poor prognostic factor for full recovery.

Case series

Single-center data analysis of a case series of 11 consecutive patients who were diagnosed with ISCH and underwent surgery between 2009 and 2021.

All herniations were located in the thoracic spine between T2 and T9. In most cases, gait ataxia and dysesthesia led to further workup and subsequently to the diagnosis of ISCH. A “far-enough” posterior-lateral surgical approach, hemilaminectomy or laminectomy with a transdural approach, was performed under intraoperative neurophysiological monitoring which was followed by adhesiolysis, repositioning of the spinal cord and sealing using a dura patch. After surgery, clinical symptoms improved in 9 of 11 patients (81.8%), while only 1 patient experienced deterioration of symptoms (9.1%) and 1 patient remained equal (9.1%). The median preoperative McCormick grade was 3 (±0.70), while the median postoperative grade was 2 (±0.98) (P = .0047).

They found that in most patients, neurological deficits improved postoperatively. This indicates that surgery in ISCH should not be delayed in symptomatic patients [3].

More than 100 cases of Idiopathic ventral thoracic spinal cord herniation have been reported with significant variance in surgical treatment strategies and likewise, significant variance in patient outcomes. Although laminectomy has often been used, to date, there is no consensus regarding the optimal surgical approach or strategy for ventral dural repair.

Herring et al. report and illustrate a novel approach to repair the ventral dural defect with more than 2 yr of clinical follow-up. The specific approach and graft used are both detailed.

A retrospective chart review of all known cases of idiopathic spinal cord herniation at the Cleveland Clinic over the last 15 yr was performed. Postoperative outcome scores (including the Japanese Orthopedic Association score (JOA), European Myelopathy score, and Nurick) were calculated preoperatively and postoperatively.

A total of 5 patients were identified. Four of five patients improved clinically after surgery and 1 patient remained unchanged at last follow-up (average 23.2 mo, range 12-60 mo). There were no complications. All patients had postoperative magnetic resonance imaging demonstrating realignment of the spinal cord and no recurrence of tethering.

A unilateral dorsolateral, transpedicular approach combined with laminectomy provides excellent exposure for ventral or ventrolateral dural defects associated with idiopathic spinal cord herniation and minimizes spinal cord manipulation. A collagen matrix graft used as an onlay between the spinal cord and ventral dural defect is a safe and effective option for ventral dural repair [4].

Among 1519 patients with spinal space-occupying lesions, 66 patients demonstrated spinal dura mater pathologies. Neuroradiological and surgical features were reviewed and clinical data analyzed.

Saccular dural diverticula (type I, n = 28) caused by defects of both dural layers, dissections between dural layers (type II, n = 29) due to defects of the inner layer, and dural ectasias (type III, n = 9) related to structural changes of the dura were distinguished. For all types, symptoms consisted of local pain followed by signs of radiculopathy or myelopathy, while one patient with dural ectasia

presented a low-pressure syndrome and 10 patients with dural dissections additional spinal cord herniation. Type I and type II pathologies required occlusion of their dural defects via extradural (type I) or intradural (type II) approaches. For type III pathologies of the dural sac no surgery was recommended. Favorable results were obtained in all 14 patients with type I and 13 of 15 patients with type II pathologies undergoing surgery.

The majority of dural pathologies involving nerve root sleeves remain asymptomatic, while those of the dural sac commonly lead to pain and neurological symptoms. Saccular dural diverticula (type I) and dissections between dural layers (type II) pathologies were treated with good long-term results occluding their dural defects, while dural ectasias (type III) were managed conservatively [5].

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92 cases were reported in the literature till 2006 [6].

It is a frequently misdiagnosed condition. It preferentially affects women and causes progressive thoracic myelopathy that presents as a Brown Séquard syndrome or as spastic paraparesis. Although its etiology and pathogenesis are controversial, ISCH is characterized by the presence of an anterior dural defect that allows the incarceration of a segment of the cord. Typically, a C-shaped ventral displacement and kinking of the cord are visible on sagittal MRI. Surgery aimed at stopping or reversing myelopathic symptoms is usually recommended for symptomatic patients. Surgical options include reduction of the hernia and direct suturing, or enlargement of the dural defect, with or without patching. Suturing under the cord in a very tight space can be troublesome and may lead to neurological deterioration. The authors present the case of a symptomatic ISCH in which nonpenetrating titanium microstaples were used to close the dural defect after cord reduction. The patient experienced a good outcome, and the follow-up MRI study showed adequate cord repositioning and stability of the suture. The use of microstaples, which allows for an easier and faster dural closure than conventional suturing, is a novel technical adjunct that has not been previously reported for this condition. In addition, microstaples produce minimal metallic artifact that does not hinder the quality of follow-up MR images [7].

see Sadek AR, Nader-Sepahi A. Idiopathic thoracic intravertebral spinal cord herniation. Br J Neurosurg. 2016 Dec 14:1-2. [Epub ahead of print] PubMed PMID: 27967246 from the Department of Neurosurgery, Wessex Neurological Centre , University Hospital Southampton , Southampton , UK.

Thoracic spinal cord herniation (TSCH) is underdiagnosed.

Case reports

A patient underwent surgery using the dural graft sling technique for repair of the dural defect and restoration of normal spinal cord position within the thecal sac. A review of the literature revealed a total of 171 patients supplemented by our 1 patient, which were then analyzed.

The majority of patients, treated with a variety of surgical techniques, experienced improvements in symptomatology. Our patient experienced significant improvement in symptomatology.

Although ISCH is a rare clinical condition that causes myelopathy, patients managed with surgery generally, though not universally, have a favorable neurological outcome. The associated surgical

technique video demonstrates the dural sling technique for the treatment of this rare disorder [8].

A 61-year-old man with a history of progressive myelopathy causing left lower-extremity weakness with associated numbness, impaired gait, foot drop, incontinence, and sexual impotence was referred without any previous treatment. Computed tomographic myelography and magnetic resonance imaging of the thoracic spine showed ventral spinal cord herniation at T3-T4. Neurological monitoring was recorded preoperatively and intraoperatively. The patient underwent left-sided posterolateral exploration via T3-T4 laminectomies and costotransversectomy for intradural cord release/detethering of the spinal cord with additional superior and inferior extension and repair of the dural defect. Arthrodesis was not considered necessary. After cord release, motor evoked potentials showed immediate improvement from baseline. Dural duplication was considered the cause of TSCH in this case. Total reduction of herniation was evident in postoperative images. The postoperative course was uneventful, and at the last follow-up, the patient had regained ambulation and sphincter control [9].

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