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Editor
Nancy E. Epstein, MD
Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook

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Case Report

Thoracic ventral spinal cord herniation with progressive myelopathy – A case report and review of the literature

Taylor Anne Wilson[1]*, Ramachandran Pillai Promod Kumar[1], Emmanuel Omosor[2]*

[1]Department of Neurosurgery, Loma Linda University Medical Center, [2]Department of Neurosurgery, School of Medicine, Loma Linda University School of Medicine, Loma Linda, California, United States.

E-mail: Taylor Anne Wilson - taylor.wilson5@gmail.com; Ramachandran Pillai Promod Kumar - rpromodkumar@llu.edu; *Emmanuel Omosor - eomosor@llu.edu
*Both these authors contributed equally.

***Corresponding author:**
Emmanuel Omosor,
School of Medicine, Loma Linda University School of Medicine, 11175 Campus Street, Loma Linda-92350, California, United States.

eomosor@llu.edu

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ABSTRACT

Background: Idiopathic spinal cord herniation (ISCH) is a rare, underrecognized, and often misdiagnosed entity of unclear pathogenesis that typically presents as a slowly progressive thoracic myelopathy. There are less than 200 such cases reported in the literature. ISCH diagnosis and treatment are often delayed contributing to greater fixed neurological deficits, often leading to costly, unnecessary imaging studies, and inappropriate surgery.

Case Description: Here, a 48-year-old female presented with trauma-induced ISCH characterized by gradually worsening lower extremity myelopathy.

Conclusion: Idiopathic spinal cord herniation (ISCH) is rare, often underdiagnosed posttraumatic myelopathy that, when accurately diagnosed and treated, can result in good outcomes.

Keywords: Cerebral palsy, Myelopathy, Spinal cord herniation

INTRODUCTION

Idiopathic spinal cord herniation (ISCH), also called ventral spinal cord herniation (VSCH), is a rare and often underdiagnosed cause of slowly progressive thoracic myelopathy. It involves anterior or anterolateral displacement of spinal cord tissue through a small, ventral dural defect most typically arising in the upper/mid thoracic spine between the T2-T8 levels.[4,12] There are fewer than 200 such cases described in the literature. As the diagnosis of ISCH is often delayed, many patients undergo progressive irreversible neurological deterioration before diagnostic studies confirming the diagnosis and before definitive appropriate surgery.[5]

CASE DESCRIPTION

A 48-year-old female presented 3 years after being thrown against a wall with a sequela of traumatic brain injury (TBI) with cerebral palsy, worsening myelopathy, and gait ataxia. She initially utilized a walker for ambulation, but progressed to wheelchair bound by the time of presentation [Table 1].

Diagnostic studies

She underwent MRI studies of the brain and spine, plus EMG and NCV of the lower extremities. The thoracic MRI showed a ventral thecal sac irregularity with accompanying distortion of the ventral cord, consistent with "tethering;" cervical and lumbar studies were unremarkable [Figure 1]. The CT myelogram was suggestive of ventral dural tear and spinal cord herniation at the T7-8 levels and a diffuse ventral subdural fluid collection extending from T1 to T10 [Figure 2].

Surgery

We performed a T7-T8 laminectomy and unilateral facetectomy with removal of the right T7 pedicle and transverse process, exposing the T7-8 nerve roots [Figure 3]. After using ultrasounds to confirm the level, the right T7 nerve root was ligated. Using microsurgical technique, the dural defect was approached from the right side intra- and extradurally. Once the dural defect was identified, it was repaired with onlay and inlay DuraGen (Integra LifeSciences, Princeton, NJ, USA) and, after weighing the risk and benefits, we reinforced with DuraSeal (Integra LifeSciences, Princeton, NJ, USA) to minimize risk of postoperative CSF leak.

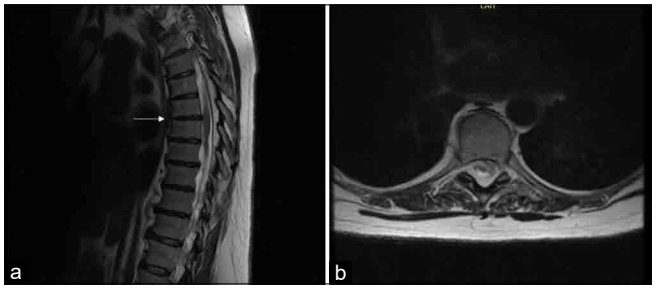

Figure 1: Preoperative MRI thoracic spine. (a) Sagittal view; arrow pointing to the level of the spinal cord herniation at T7-8, (b) Axial view at the level of the spinal cord herniation T7-8. MRI thoracic spine, T2-weighted sequence.

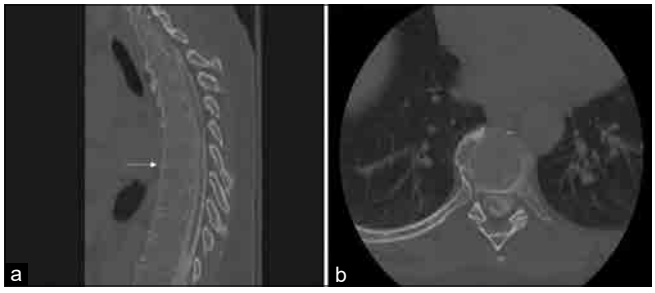

Figure 2: CT myelogram. (a) Sagittal view; arrow pointing to the level of the spinal cord herniation at T7-8, (b) Axial view at the level of the spinal cord herniation at T7-8. CT myelogram.

Follow-up

Postoperatively, the patient went to rehab and within 2 months, she was ambulating with assistance using a walker. Her strength continued to improve at her 4-month and 1-year neurosurgical follow-up visits [Table 1]. She is now ambulating independently with a walker. On repeat, MRI at 4 months postoperatively demonstrates postsurgical changes with a well-circumscribed right paraspinal fluid collection with minimal mass effect on the spinal cord [Figure 4]. There was no clinical or radiographic evidence of recurrent herniation, and the patient did not experience clinical signs or symptoms of CSF leak.

DISCUSSION

ISCH is a rare cause of progressive thoracic myelopathy. Females are affected nearly twice as frequently as males.[5,9]

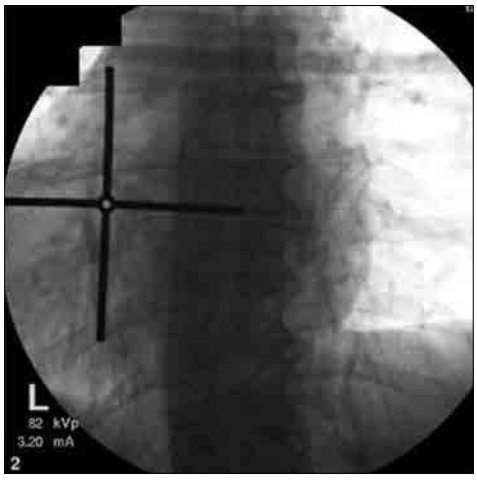

Figure 3: Intraoperative localization of T7-8 with fluoroscopy. Intraoperative localization of the level of the spinal cord herniation at T7-8 with fluoroscopy.

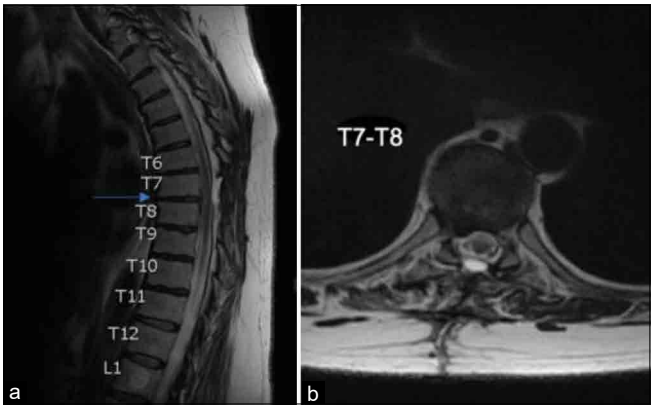

Figure 4: Postoperative MRI thoracic spine. (a) Sagittal view; arrow pointing to the level of the prior spinal cord herniation at T7-8, (b) Axial view at the level of the spinal cord herniation T7-8.

Table 1: Physical Exam at Presentation and 4 Month Follow-Up

Focused Neurologic Exam	Pre-Operatively	Post-Operatively (4 months)	Post-Operatively (1 year)
Motor			
-Hip flexion	Right 2/5, Left 3/5	Right 4-/5, Left 4/5	Right 4/5, Left 4+/5
-Knee extension	Right 2/5, Left 3/5	Right 4-/5, Left 4/5	Right 4/5, Left 4+/5
-Dorsiflexion	Right 2/5, Left 3/5	Right 4-/5, Left 4/5	Right 4/5, Left 4+/5
-Extensor hallucis longus	Right 2/5, Left 3/5	Right 4-/5, Left 4/5	Right 4/5, Left 4+/5
-Planter flexion	Right 2/5, Left 3/5	Right 4-/5, Left 4/5	Right 4/5, Left 4+/5
Sensory			
-Light touch	Diminished T10 distally	Baseline, no deficit	Baseline, no deficit
-Pinprick	Diminished T10 distally	Baseline, no deficit	Baseline, no deficit
-Vibration	Diminished T10 distally	Baseline, no deficit	Baseline, no deficit
-Proprioception	Diminished T10 distally	Baseline, no deficit	Baseline, no deficit
Reflexes			
-Patellar	3+	3+	3+
-Achilles	3+	3+	3+
-Plantar (Babinski)	Downgoing	Downgoing	Downgoing
-Clonus	Absent	Absent	Absent
Tone	Increased – spastic	Increased – spastic	Increased – spastic
Ambulatory status	Wheelchair bound	Ambulating with walker	Ambulating with walker

Patients are typically in their fifties (range 12–80 years of age).[5,6] With less than 200 cases described in the literature, ISCH is often misdiagnosed, leading to delays in patient care and increasing the risk of further neurological decline.[5,11]

Etiology

Several hypotheses exist regarding the pathogenesis of ISC. In 1974, Wortzman *et al.* described a patient with a gradually worsening spastic paraplegia due to a congenital myelocele.[12] Other theories purport that the dural defect is acquired, occurring secondary to trauma, disc protrusion, ventral pressure from a dorsal arachnoid cyst, and disc herniation.[3,11] In the thoracic spine, anterior dural erosion generates a CSF leak that pushes adjacent spinal tissue with the cord herniation tamponading the leak and producing progressive myelopathy.[5,9]

Variable neurological presentation

Characteristically, ISCH leads to gradual onset of lower extremity myelopathy and spastic paresis; however, 60–75% of patients present with Brown-Sequard syndrome, while others develop asymmetric/symmetric lower extremity spastic paresis. Other symptoms/signs include 70% incidence of neck/upper back pain and 10% frequency of bladder dysfunction.[2]

Diagnostic MR or Myelo-CT studies

Thin cut thoracic spine MRI is the gold standard for diagnosing ISCH. These studies typically show a C-shaped ventral kink or displacement of the thoracic cord adjacent to the dural defect.[11] MRI with and without contrast should be performed to rule out a dorsal arachnoid cyst versus intradural extramedullary mass.[7] The CT myelogram may more directly demonstrate the ventral dural defect.[5,8]

Outcomes with early surgery

Earlier operative intervention has better prognoses/outcomes.[3] The failure to consider ISCH in the differential diagnosis exposes patients to redundant, unnecessary imaging studies, and delayed surgery.[9]

Operative intervention requires direct repair of the dural defect, reduction of the cord into the canal, and detethering adhesions.[5] The standard operative approach is laminectomy with release of ligaments and lysis of adhesions or costotransversectomy with unilateral pedicle removal.[1,5,7].

Dural closure methods

Dural repair techniques for ISCH include primary repair versus duraplasty. Primary repair is often considered more secure; however, it may contribute to thecal sac stenosis and greater postoperative neurological deficits due to pressure around the cord. Conversely, duraplasty minimizes the risk of inadvertent spinal cord manipulation and the related postoperative neurological dysfunction, but introduces a foreign material into the patient.[4,10,12]

CONCLUSION

ISCH is rare, often underdiagnosed posttraumatic myelopathy that, when accurately diagnosed and treated, can result in good outcomes.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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