Section Editor

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Clinical Reasoning:

A case of slowly progressive painful paraparesis

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SECTION 1

In 2003, a 50-year-old woman was admitted to the emergency department of a large hospital for a 2-month history of stabbing epigastric pain radiating backward to the right scapula. The pain was not relieved in any position and was not preceded by fever or trauma. Her history was unremarkable. General examination including temperature, pulse, and blood pressure was normal.

Acute epigastric pain may have different non-neurologic etiologies including gastric, biliary, and pancreatic disorders. To rule out gastric disorders, a gastroscopy was performed. Gastroscopy revealed mild erythema of the gastric mucosa, but the histologic examination and studies for *Helicobacter pylori* were negative. The complete blood count as well as

hepatic, pancreatic, and renal function were normal. Abdominal CT scan was normal. Ultrasonography of the right upper quadrant of the abdomen revealed no evidence of gallstones or cholecystitis. Dilation of the biliary and pancreatic ducts was ruled out by magnetic resonance cholangiopancreatography.

The patient was discharged with diagnosis of “gall-bladder hypokinesia” and pain slowly decreased over the following 2 years. In 2005, the patient demonstrated slowly progressive perineal and lumbar pain radiating to the left leg.

Question for consideration:

1. What investigations would you propose for this patient?

SECTION 2

Radicular pain along the leg can be related to disc herniation. Slipped discs occur more often in men aged between 30 and 55 years. The lumbar segment is more commonly affected followed by the cervical and thoracic segments. MRI is the gold standard in diagnosis of disc herniation.

The patient underwent lumbosacral MRI, which revealed disc herniation between the fourth and fifth lumbar vertebrae. In order to evaluate the involvement of the spinal roots at this level, an EMG study was performed. The test revealed minimal chronic neurogenic signs—namely, high-amplitude rapidly firing motor unit potentials without fibrillation potentials—in muscles supplied by the fourth lumbar root. Subsequently, the pain was complicated by left lower limb spasticity; because of these symptoms, L4–L5 laminectomy and flavotomy were performed in 2007. The surgical intervention did not produce any benefit. She progressively worsened over the course of the following years and

developed progressive gait disturbance with multiple falls, perineal and left lower limb hypoesthesia, and right lower limb spasticity. She also reported mild urinary retention. In 2012, neurologic examination revealed mild spastic paraparesis, hypoesthesia with upper level at T6, hyperreflexia in both lower limbs associated with bilateral clonus, and left Babinski sign. The cranial nerve and upper limb examination was normal.

Questions for consideration:

1. Low back pain radiating to the lower limbs is a typical manifestation of lumbar radiculopathy; however, could the association with perineal pain, gait disturbance, and lower limb spasticity suggest a different diagnosis?
2. What anatomic structure could be involved that would explain the clinical presentation?
3. What diagnoses are most likely in light of the above clinical findings?

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SECTION 3

In a patient with history of acute thoracic pain, sensory level, bladder dysfunction, hyperreflexia, clonus, and Babinski sign, we would worry about spinal cord disorders. These represent a heterogeneous group of syndromes characterized by spinal cord dysfunction resulting in paresis with a sensory level and autonomic (bladder, bowel, and sexual) impairment below the level of the lesion. Several causes could produce myelopathy: compressive pathologies (spondylotic changes or disc herniation); spinal cord vascular malformations; metabolic, nutritional, and toxic diseases; autoimmune or infective diseases; or paraneoplastic syndromes.

Myelopathies have distinct clinical and radiologic features and prognoses. High-resolution MRI revealed a typical appearance of transdural spinal cord herniation (SCH) at the T5–T6 level, with reduced sagittal diameter and asymmetric (right) transdural protrusion of the cord and widening of the dorsal subarachnoid space. The presence of an arachnoid cyst can be ruled out on the basis of the shape of the posterior surface of the herniated cord and because of evidence of flow artifacts in the posterior CSF. At the same level, a discrete disc herniation was present (figure 1).

MRI findings provide a reliable explanation for clinical features. Indeed, SCH causes a chronic slowly progressive myelopathy due to mechanical compression

and impaired blood flow in the stretched cord. We can speculate that in 2003, when the patient suddenly experienced epigastric pain with dorsal radiation, an acute dorsal disc herniation caused a dural defect with subsequent slowly progressive herniation of the cord manifesting with progressive sensory and motor symptoms caudal to the level of compression. The patient subsequently underwent surgery. SCH was confirmed and reduction was successfully performed (figure 2). Six months later, the patient showed a significant clinical improvement in gait and sensory deficits. Neurologic examination revealed left lower limb hypoesthesia, hyperreflexia in both lower limbs without clonus, and a left Babinski sign. She was able to return to her job at a bank.

DISCUSSION

SCH was first reported by Wortzman et al.¹ in 1974 as a treatable cause of myelopathy. It is often misdiagnosed and because of this, it progressively gained importance, as shown by the increasing number of cases described in the literature: 10 cases in 2000, 30 cases in 2004, and about 150 to date. SCH is prevalent in middle-aged women. To date, there is only one pediatric case of SCH described in the literature.² Idiopathic SCH (ISCH), which results from a dural defect of unknown origin, is distinguished from herniation due to surgery or documented traumatic cause. Usually SCH occurs between the

Figure 1 Preoperative spine MRI findings

Sagittal T1- and T2-weighted images (A, B) demonstrate the focal adhesion of the spinal cord to the anterior wall of the spinal canal and the presence of a discrete disc herniation at the T5–T6 level. The axial T2-weighted images (C, D) show the right anterior displacement of the cord that protrudes beyond the dura (long arrow). The posterior convex contour of the spinal cord and the evidence of low signal (due to turbulent CSF flow, short arrows) in the posterior subarachnoid space rule out the possibility of arachnoid cyst.

Figure 2 Postoperative spine MRI findings

Postoperative sagittal (A) and axial (B) T2-weighted images demonstrate the regular shape, size, and intradural location of the spinal cord, with normal evidence of perimedullary subarachnoid space; there is minimal intramedullary spinal cord signal alteration.

T4 and T7 levels through a dural defect located ventrally, ventrolaterally,³ or, more rarely, dorsally.⁴

Tethered cord syndrome, consequent to SCH, is a stretch-induced functional disorder of the spinal cord due to mechanical compression and to the impaired blood flow in stretched cord. This can cause back pain radiating to the lower limbs as well as sensory and motor deficits.

Patients commonly present with symptoms of spinal cord compression indistinguishable from those produced by other causes of extradural or intradural extramedullary cord compression.⁵ Local or radicular pain is usually the presenting symptom, followed by progressive sensory and motor deficits below the level of compression and disturbance in sphincter control. While local or radicular pain is commonly the presenting symptom of compressive spinal cord disorders, it tends to appear in more advanced phases of the disease in intramedullary spinal cord disorders, in which symptom progression is also more insidious.⁵

In more than 50% of cases the clinical presentation is the Brown-Sequard syndrome,⁶ characterized by unilateral motor deficit due to the involvement of the anterolateral funiculi⁷ and ipsilateral proprioception loss due to dorsal column involvement together with contralateral pain and temperature sensation deficits due to the involvement of the spinothalamic tract, which crosses over immediately after the entry into the spinal cord. In ISCH, Brown-Sequard syndrome can progress to spastic paraparesis. Other symptoms include bladder dysfunction, intractable leg pain, and, more rarely, headache that could be related to an alteration in CSF circulation. In our patient, local pain was the presenting symptom, followed for several years by unilateral lower limb sensory and motor deficits; this presentation might be caused by a hemicord lesion, although the presentation was not that of the Brown-Sequard syndrome.

Several hypotheses have been advocated to explain ISCH. Wortzman et al.¹ postulated that the rupture of a dorsal disc produces the dural defect responsible for SCH. More recently, it has been suggested that a focal inflammatory process could induce adhesion between the spinal cord and ventral dura and then CSF pulsation, posterior arachnoid cyst pressure,⁸ or abnormal movements could produce erosion of the dura and cord herniation.

In the present case, SCH might be related to a disc disease according to the theory proposed by Wortzman et al.¹ In our patient, a dorsal median disc herniation at T5–T6 that occurred in 2003 (when she first experienced pain) caused a dural defect through which the spinal cord progressively herniated during the following years with the development and worsening of the neurologic clinical presentation.

MRI is the gold standard for diagnosis of this condition. A ventrally displaced and thinned cord in the upper or mid thoracic region represents the classical finding; the availability of high-resolution T2 images provides the direct demonstration of the herniation of the spinal cord beyond the anterior dura mater and excludes a posterior arachnoid cyst. Indeed, a convex contour of the posterior surface of the cord is more consistent with SCH, while a concave contour is suggestive of an arachnoid cyst. Moreover, phase-contrast MRI could be useful to differentiate SCH from arachnoid cyst by demonstrating an absence of pulsatile CSF flow at the ventral side of the spinal cord at the level of the herniation.⁹ In select cases, CT myelogram may be required. MRI can also have prognostic significance; indeed, T2 signal change in the herniated cord suggests progression toward irreversible spinal cord damage, associated with a worse prognosis.

Early surgery is the gold standard treatment in this condition¹⁰; the surgeon can reduce the spinal cord and repair the dural defect or can widely open it in order to avoid strangulation.

AUTHOR CONTRIBUTIONS

M.C., F.C., and V.D.L. drafted/revised the manuscript for content, including medical writing for content. M.C. and V.D.L. were involved in study concept and design. V.D., C.Q. C.C., and G.M. participated in analysis and interpretation of data.

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DISCLOSURE

The authors report no disclosures relevant to the manuscript. Go to Neurology.org for full disclosures.

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