Diagnosis and treatment of spinal cord herniation: a combined experience

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Object. Idiopathic spinal cord herniation (ISCH) is an uncommon clinical entity typically presenting with lower-extremity myelopathy. Despite the existence of 85 ISCH cases in the literature, misdiagnosis and delayed diagnosis remain a major concern.

Methods. The authors conducted a retrospective review of patients who underwent surgery for ISCH at their institutions between 1993 and 2004. Seven patients were treated for ISCH, five in New York and two in Buenos Aires. The patients’ ages ranged from 32 to 72 years. There were three men and four women. The interval between the onset of symptoms and surgery ranged from 12 to 84 months (mean 42.1 months).

Preoperatively, spinal cord function in four patients was categorized as American Spinal Injury Association (ASIA) Grade D, and that in the other three patients was ASIA Grade C. In all patients a diagnosis of posterior intradural arachnoid cyst had been rendered at other institutions, and three had undergone surgery for the treatment of this entity. In all cases, the herniation was reduced and the defect repaired with a dural patch. The follow-up period ranged from 10 to 147 months (mean 49.2 months). Clinical recovery following surgery varied; however, there was no functional deterioration compared with baseline status. Syringomyelia, accompanied by neurological deterioration, developed postoperatively in two patients at 2 and 10 years, respectively.

Conclusions. Patients presenting with a diagnosis of posterior intradural arachnoid cyst should be evaluated carefully for the presence of an anterior spinal cord herniation. Based on the authors’ literature review and their own experience, they recommend offering surgery to patients even when neurological compromise is advanced.

KEY WORDS • spinal cord herniation • arachnoid cyst • spinal lesion • thoracic spine

Diopathic spinal cord herniation is an uncommon clinical entity that presents with progressive sensorimotor dysfunction of the lower extremities. It occurs when the spinal cord herniates through an anterior dural defect in the upper to middle thoracic spine (Fig. 1). To date, more than 85 cases of ISCH have been reported in the English-language literature, the majority having been reported in the last 10 years. Current imaging modalities have enhanced our ability to diagnose this condition. Nevertheless, a review of recent literature reveals that a significant delay (range 2–14 years) exists between the onset of symptoms and surgical repair. This delay is probably due to both the rarity of the condition and the fact that ISCH can masquerade as an arachnoid cyst.

We describe our combined experience in the diagnosis and treatment of seven patients with thoracic ISCH. We illustrate one case in which some of the diagnostic pitfalls are detailed. Additionally, we discuss useful imaging guidelines in the diagnosis, and we present our surgical technique. The origin and pathogenesis of ISCH are reviewed.

Clinical Material and Methods

We retrospectively reviewed the charts, radiographic studies, and operative reports obtained in seven patients
treated for ISCH at our two institutions between 1993 and 2004. For comparison purposes, we used the ASIA Impairment Scale to determine levels of spinal cord dysfunction. All patients underwent preoperative MR imaging of the thoracic spine; two patients also underwent preoperative myelography. There were three men and four women who ranged in age from 32 to 72 years.

Surgical Technique

To facilitate intraoperative localization of the ISCH, all patients undergo preoperative radiographic evaluation of the thoracic spine, lumbar spine, and ribs. General anesthesia is induced and maintained with a total intravenous anesthesia technique. An arterial line is placed in the radial artery, and when feasible, SSEPs and MEPs are monitored continuously throughout the procedure. Patients are positioned prone on chest rolls on a radiolucent table. Both anteroposterior chest radiographs and lateral thoracic and lumbar radiographs are obtained, and the appropriate rib and vertebral levels are counted. In patients with no history of back surgery, a midline incision is made and a two-level laminectomy is performed. In patients with a history of surgery for posterior arachnoid cyst, we open the same incision and perform dissection carefully through the scar tissue to the dura mater.

To provide a posterolateral approach to the anterior herniation, bone removal is extended further laterally on one side if the cord herniation is eccentric. The dura is opened in the posterior midline and the dural edges are sutured to the retracted paraspinal muscles. Two bipolar electrodes (PMT Corporation, Chanhassen, MN) are placed on the midline posterior epidural surface, rostral and caudal to the durotomy site. The rostral electrode is used to record SSEPs and the caudal electrode is used to record the MEP D-wave.

Using the operating microscope, the arachnoid is opened in the midline and its edges are clipped to the retracted edges of the dura. The spinal cord is noted to be adherent to the anterior dura with a large posterior subarachnoid space (Fig. 2A). Two adjacent dentate ligaments on one side are divided at their attachment to the dura, leaving a length of the ligaments attached to the pia mater and spinal cord to facilitate rotation of the cord. Using the dentate ligaments as handles, the spinal cord is gently rotated to the contralateral side. The dentate ligaments are then clipped to the edge of the cut dura (Fig. 2B). Partial rotation of the spinal cord facilitates the exposure of its anterior surface, allowing better visualization of the herniated area. Continuous feedback from the attending neurophysiologists is used to determine the feasibility of this rotation maneuver. If there is any change in the monitoring parameters, this rotation maneuver is aborted temporarily. We typically wait until the MEPs and SSEPs return to baseline levels and then attempt a limited second rotation maneuver. If the monitoring parameters change again, we stop this maneuver and further extend the osseous exposure laterally by drilling the entire facet joint and pedicle at this level.

The anteriorly located spinal cord herniation is then inspected both intra- and extradurally. The herniated spinal cord is usually tethered anteriorly to the epidural tissues and may or may not have a covering of arachnoid (Fig. 2A and B). Arachnoidal adhesions between the ring of the hernial orifice and the herniating spinal cord, as well as adhesions to the epidural tissues, are freed by blunt and sharp dissection, using operative magnification and microinstruments. If the reduction proves difficult, the dura...
adjacent to the hernial orifice is divided longitudinally to facilitate the reduction. Because the dentate ligaments are cut bilaterally, the surgeon can carefully alternate the rotation to achieve a complete view of the dural defect from both sides. After the herniated portion of the spinal cord in the extradural space is dissected free, this portion of cord is gently mobilized and brought into the intradural space. Once the cord is reduced, the hernial orifice is closed by duraplasty in which either AlloDerm (LifeCell Corporation, Woodlands, TX) or a piece of thoracolumbar fascia is used. The thoracolumbar fascia is harvested laterally, within the laminectomy incision. The graft is placed over the hernial orifice intradurally anterior to the spinal cord and sutured to the surrounding dura. We place four over the hernial orifice intradurally anterior to the spinal cord tissue. After the extradural herniated portion of the spinal cord was dissected free, this portion of cord was tethered anteriorly and to the left of the spinal canal. An incomplete membrane was noted around the herniated cord tissue. After the extradural herniated portion of the spinal cord was dissected free, this portion of cord was gently mobilized and brought into the intradural space. A segment of thoracolumbar fascia was harvested to serve as a sling, maintaining the reduced spinal cord within the intradural space. At this point in the procedure, the MEPs deteriorated in both amplitude and latency; 15 minutes later they recovered but did not return to the baseline values.

In all patients in our series a diagnosis of posterior in-
Combined experience in idiopathic spinal cord herniation

TABLE 1
Summary of demographic, pre- and postoperative data obtained in seven patients with ISCH*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age (yrs), Sex</th>
<th>ASIA Grade/Symptom</th>
<th>Preop</th>
<th>Postop</th>
<th>ISCH Level</th>
<th>Interval (mos)</th>
<th>FU (mos)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>65, F</td>
<td>ASIA C w/ BSS; urinary dysfunction</td>
<td>ASIA C w/ BSS; no improvement of urinary symptoms</td>
<td>T4–5</td>
<td>36</td>
<td>60</td>
<td></td>
</tr>
<tr>
<td>2†</td>
<td>32, M</td>
<td>ASIA C; urinary dysfunction</td>
<td>ASIA D; incomplete improvement of urinary symptoms</td>
<td>T7–8</td>
<td>12</td>
<td>147</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>54, F</td>
<td>ASIA C w/ BSS; urinary dysfunction; dyspareunia</td>
<td>ASIA C w/ BSS; incomplete improvement of urinary &amp; bowel symptoms</td>
<td>T2–3</td>
<td>84</td>
<td>16</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>60, F</td>
<td>ASIA D w/ BSS; bowel dysfunction</td>
<td>ASIA D w/ BSS; objective improvement of motor strength &amp; gait</td>
<td>T2–3</td>
<td>27</td>
<td>42</td>
<td></td>
</tr>
<tr>
<td>5†</td>
<td>59, F</td>
<td>ASIA D w/ BSS; urinary dysfunction</td>
<td>ASIA E; incomplete improvement of urinary symptoms</td>
<td>T5–6</td>
<td>16</td>
<td>40</td>
<td></td>
</tr>
<tr>
<td>6</td>
<td>34, M</td>
<td>ASIA D</td>
<td>ASIA D</td>
<td>T7–8</td>
<td>60</td>
<td>30</td>
<td></td>
</tr>
<tr>
<td>7</td>
<td>72, M</td>
<td>ASIA D w/ BSS</td>
<td>ASIA D w/ BSS</td>
<td>T4–5</td>
<td>60</td>
<td>10</td>
<td></td>
</tr>
<tr>
<td>mean</td>
<td>53.7</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>42.1</td>
<td>49.2</td>
</tr>
</tbody>
</table>

* BSS = Brown–Séquard syndrome; FU = follow up; Interval = time gap between the onset of symptoms and surgery.
† Following an initial improvement of 141 (Case 2) and 30 (Case 5) months, a thoracic syrinx and clinical deterioration developed (ASIA Grade D to C [Case 2], and ASIA Grade E to D with BSS [Case 5]).

Illustrative Case

Case 7

**Presentation.** This 73-year-old man presented in October 2003 with a 5-year history of gait difficulty; a diagnosis of lumbar stenosis had been previously made. He reported a feeling of fatigue in the legs and numbness in the right thigh. He was unable to walk more than two blocks without stopping. He also complained of a loss of perception of temperature sensation in his right leg, especially while in the shower. Additionally, he complained of numbness in the left side of his anterior chest wall, as well as frequency of micturition.

**Examination.** This patient was a mildly obese individual with a spastic gait. On motor examination, left hip flexion was Grade 4/5, whereas his remaining motor status was unremarkable. On sensory examination, he was found to have a pinprick sensory level at T6–7 on the right side. Left-sided sensation was normal. Deep tendon reflexes were Grade 2 on the right side and Grade 3 on the left. He had an upgoing plantar reflex on the left side. Spinal cord function was ASIA Grade D and Brown–Séquard syndrome was present.

Despite the severe stenosis noted on the lumbar MR imaging study, the patient was referred for thoracic MR imaging, which revealed a T4–5 abnormality, suspicious for anterior spinal cord herniation (Fig. 4A and C). The patient was advised to undergo surgery for the thoracic spinal cord herniation in November 2003 but refused. In September 2004 he presented with left leg weakness and increasing anterior chest wall numbness that extended further laterally, but he again refused surgery. In December 2004 he presented with worsening of his gait and was walking with a cane, and he experienced greater left hip weakness (Grade 3/5) and left knee flexion weakness (Grade 4/5). In May 2005, with his condition continuing to deteriorate, he agreed to the surgery.

tradural arachnoid cyst had previously been established at other institutions, and three patients had undergone surgery for this lesion. These three patients (Cases 1–3) did not improve symptomatically following surgery. One patient (Case 1) underwent craniotomy for a meningioma when she had presented with progressive myelopathy. One patient (Case 5; Fig. 3) had a thoracic disc herniation causing spinal cord compression. She underwent a thoracoscopic discectomy (Fig. 3C) before being brought back for the treatment of the anterior ISCH (Fig. 3D).

Postoperative Clinical Course

The follow-up period ranged from 10 to 147 months (mean 49.2 months). All of our patients presented with significant neurological deficits. Neurological recovery after surgery varied; however, at the time of the last follow-up examination, no deficit was worse than it had been at baseline preoperatively. Status in three patients initially improved (Cases 2, 4, and 5), with the ASIA grade improving in two (Cases 2 and 5). The status in these two patients, after initial clinical improvement, deteriorated at 141 and 30 months, respectively, when a syrinx developed rostral and caudal to the area of the laminectomy (Fig. 3E). We recommended syrinx drainage and duraplasty to both patients. The patient in Case 4 experienced a definite neurological improvement 3 years after the surgery; however, she still has an up-turning toe on the right side and hypesthesia with a T4–5 level over the left side of her body (her spinal cord function is ASIA Grade D). Of the four remaining patients, one (Case 6) suffered a transient postoperative worsening of motor function, which slowly improved to preoperative baseline status. The status in three patients (Cases 1, 3, and 7) remains at the preoperative baseline, with none having exhibited additional neurological deterioration. No postoperative CSF leak or wound infection developed in any of the patients.
Operation. A T3–5 laminectomy was performed. A widened subarachnoid space was identified posteriorly with the spinal cord plastered to the anterior dura. The spinal cord was noted to herniate out of an anterior dural defect and was adherent to the VB anteriorly. A small nipple of the cord was noted poking out of the piaarachnoid, whereas the rest of the herniation was contained within the arachnoid membrane. After reduction of the herniated cord, a piece of Alloderm was placed as a sling beneath the herniation overlying the hole in the dura, and this was tacked to the adjacent dura in several places.

Postoperative Course. The patient’s left leg weakness and gait improved. He was started on Coumadin for atrial fibrillation 10 days after surgery. One month later he was admitted to an outside hospital with a subacute subdural hematoma and underwent drainage via a bur hole. Since this event, his condition has deteriorated, and he is ambulating with a walker.

Discussion

Idiopathic spinal cord herniation is an uncommon entity occurring in the thoracic spine; it presents with progressive neurological symptoms in the lower extremities. Symptoms include progressive weakness, numbness, and myelopathy. A delay in accurate diagnosis of ISCH often leads to significant neurological dysfunction and probably exacerbates the poor outcomes observed following operative repair. Consistent with this observation, three patients in our series had undergone excision of a presumed posterior intradural arachnoid cyst. Unfortunately without a clear-cut origin for ISCH, the mechanism behind the formation of this ventral dural...
defect and subsequent spinal cord herniation remains speculative. Nevertheless, a number of theories have been advanced to explain its pathogenesis: 1) trauma in newborn and young children resulting in injury to the spinal cord with laceration of the dura without radiological evidence of fracture or dislocation; 2) pressure erosion of the anterior dura in patients thought to harbor intradural arachnoid cysts; 3) congenital defects, with the cord herniating into a preexisting anterior meningocele; and 4) duplication of the anterior dura, with the spinal cord herniating through the outer dural layer. To date, there is no radiological/imaging or pathological evidence to validate this concept. Closure-related defects of the neural tube are commonly posterior and occur at the rostral and caudal regions of the spine. They are associated with dysraphic syndromes and are unlikely to cause the anterior dural defects noted in patients with ISCH.

If an anterior dural defect is found to be a consistent anatomical precursor to the development of ISCH, the mechanical dynamics that result in eventual herniation through this defect become somewhat easier to explain. Arachnoid herniation may occur through the anterior dural defect into the extradural space and CSF would flow freely in and out. The thoracic spinal cord is anteriorly located and lies against this anterior dural defect. As long as CSF flows freely in and out of the anterior defect, no herniation will occur. In time, as the spinal cord develops adhesions to the edges of the dural defect, CSF flow becomes impeded. Cardiac pulsations and respiratory movements now push the cord into the defect and cause the observed herniation. The constant pounding of the CSF pulsations forces the cord further through the defect. The spinal cord becomes strangulated as it is forced through the narrow neck of the hernial orifice, resulting in compression of the neural elements. Neurological deficits may be due to spinal cord compression, ischemia, tethering of the cord at the site of the herniation with traction and distortion, or a combination of these factors.

Fortunately, the diagnosis of ISCH has become easier with MR imaging units in which higher-resolution magnets are used. Closed MR imaging in a high-field (1.5-tesla) magnet, when programmed for thinner slices, enhances the ability to diagnose this entity. For an accurate MR imaging diagnosis of thoracic cord herniation, certain classic features should be present. The sagittal neuroimages should show displacement of the spinal cord anteriorly, with an S- or C-shaped kink to the cord associated with widening of the posterior subarachnoid space (Figs. 3A and 4A). Sagittal images may also demonstrate atrophy of the cord with possible signal changes. Axial images will show herniation of the cord with the spinal cord plastered to the back of the VB, leaving a large posterior arachnoid space (Figs. 3B and 4C). Importantly, we have noticed that, with appropriately thin and stacked axial MR images, it is possible to visualize the nerve roots traversing the widened posterior subarachnoid space on their way to the neural foramina (Fig. 4C). This characteristic is in contrast with that observed on MR images obtained in patients with a posterior intradural arachnoid cyst, in which the nerve roots skirt the periphery of the cyst to reach the neural foramina (Fig. 4D). Cine-phase MR imaging has also been shown to be useful, especially in the diagnosis of arachnoid cysts.

Primary closure of the anterior dural defect, closure of the defect with a duraplasty, or enlargement of the dural defect in an attempt to prevent the spinal cord from repeated herniation and strangulation has been recommended in the different series. We performed
a duraplasty in all our cases. Enlargement of the dural defect, a technique described in 2001 by Watanabe, et al.,

3 is an interesting procedure with good results. Although duraplasty is technically more demanding, it prevents re-
tethering of the spinal cord.

As stated by Massicotte, et al.,

9 “understanding the natural history of this entity is made difficult by its rare oc-
currence.” In their thorough literature review, the authors found that the status in 33 of 50 surgically treated patients improved after surgery. Some authors have recommended undertaking clinical follow-up observation without sur-
gery, because the signs and symptoms attributable to ISCH are nonspecific and may not be progressive. In our experience, neurological signs attributable to ISCH do progress in time. As with any condition presenting with myelopathy, the aim of surgery is to stop neurological de-
terioration and possibly reverse existing deficits. We have not encountered any patients with an incidentally discov-
ered ISCH. In the present series all of the patients had neu-
rological deficits attributable to the ISCH. A possible ap-
proach in cases with incidental ISCH would be to undertake follow up until the patient exhibits neurologic dysfunc-
tion. We believe the best results and recovery of function are achieved with early intervention.

The long-term development of syringomyelia was an unexpected complication that has not been previously re-
ported after the repair of ISCH. Ammar, et al.,

5 have reported the observation of a signal change in the spinal cord 6 months after the reduction of a spinal cord hernia-
tion. Although it was interpreted as a syringomyelia pre-
operatively, the surgeons found no syrinx cavity intraop-
eratively. Syrinx formation may be related to iatrogenic obstruc-
tion of the subarachnoid pathways by the patch graft and possible post-surgical local arachnoiditis. Based

on our observation of syrinx formation, we are inclined to recommend that patients undergoing surgery for spinal cord herniation should, in addition to the repair of the an-
terior hernial orifice, also undergo a posterior expansive duraplasty to widen the subarachnoid space and prevent syrinx formation.

Conclusions

Idiopathic spinal cord herniation is a rare cause of pro-
gressive myelopathy. Current imaging modalities have

made earlier diagnosis of ISCH possible, thus limiting preoperative neurological deterioration and improving the chances for resolution of the neurological dysfunction. Pa-

tients presenting with a diagnosis of posterior intradural arachnoid cyst should be evaluated carefully for the pres-
ence of an anterior spinal cord herniation. Surgical treat-
ment should include repair of the anterior dural defect and posterior duraplasty, thereby possibly preventing syrinx formation in the longer term.

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