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Case Report

Surgical treatment of idiopathic transdural spinal cord herniation: a new technique to untether the spinal cord

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Summary

Idiopathic transdural spinal cord herniation is a rare but treatable cause of thoracic myelopathy caused by herniation of the spinal cord through a defect in the dura. The diagnosis is frequently missed or delayed, but the latest imaging techniques can document spinal cord herniation through a dural defect. Surgical treatment, consisting of reducing the herniation by closing the dural defect or widening the aperture to prevent spinal cord compression, is rather successful. We describe a new technique to untether the spinal cord by wrapping a dura graft around the myelum to prevent recurrent transdural herniation. Two patients and a review of the literature are discussed. We conclude that high-resolution T2 magnetic resonance imaging is the best imaging modality to detect the entity, and wrapping the myelum is an effective surgical technique to untether the spinal cord.

Keywords: Brown-Séquard syndrome; dural defect; herniation; myelopathy; spinal cord.

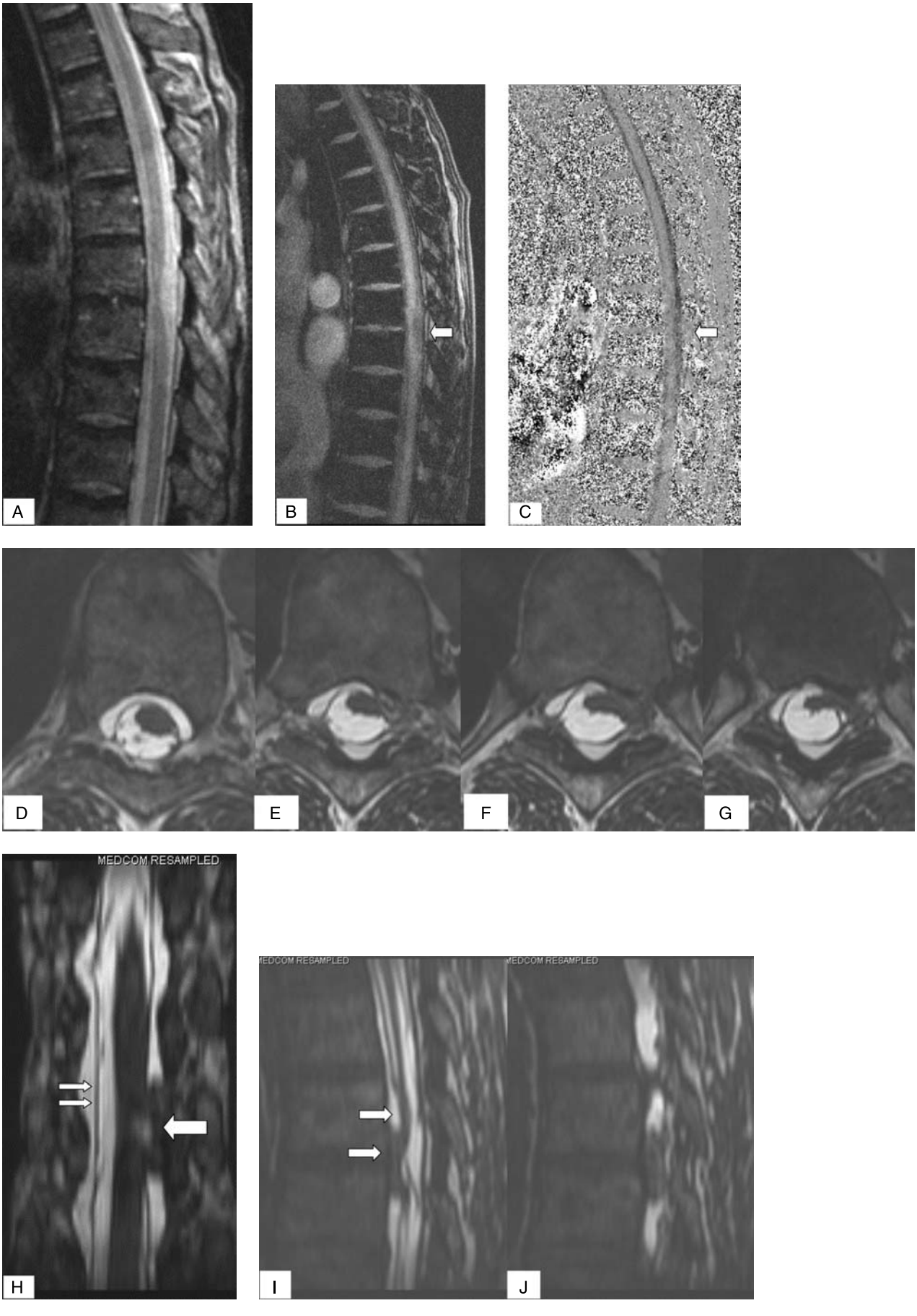
Introduction

Idiopathic transdural spinal cord herniation is a rare but treatable cause of myelopathy in which the spinal cord herniates through a defect in the dura. Wortzman *et al.* described the first patient with transdural spinal cord herniation in association with a thoracic herniated disc [24]. Presently, almost 80 patients have been described in the literature [14]. Patients typically present in middle age with a slowly progressive thoracic hemicord dysfunction. The exact cause of the defect in the dura is unknown but several theories have been proposed. Due to improved imaging techniques this condition is diagnosed more frequently. In this paper we present two patients with idiopathic transdural spinal cord herniation whose diagnosis was missed initially. The latest magnetic resonance imaging (MRI) techniques to document the entity, a new surgical technique to untether the spinal cord, and a review of the literature are discussed.

Case presentation

Case 1, an otherwise healthy 58-year old woman, presented with slowly progressive muscle weakness of the left leg without pain. She did not complain of the right leg and both arms and micturition were normal. She had a long history of headache during the upright position which resolved spontaneously. Neurological examination revealed spastic paresis of the left leg, left-sided hypesthesia below T9 and impairment of pain and temperature sensation of right leg. Position and vibration sense was preserved in both legs. Deep tendon reflexes were markedly increased in the left leg and bilateral Babinski signs were present.

MRI of the spinal cord showed midthoracic atrophy and cervical spondylotic myelopathy. A multilevel cervical laminectomy was performed without neurological improvement. Two years later, the patient presented again with progressive weakness of the left leg and urinary incontinence. MRI of the cervical spinal cord showed adequate decompression. Anterior displacement of the spinal cord at T7–T8 with local atrophy was seen on thoracic MRI. A dorsal arachnoid cyst was suspected, but high resolution T2 imaging clearly showed transdural herniation of the spinal cord through an anterolateral defect in the dura (Fig. 1). Further, CSF flow imaging using phase contrast MRI, showed normal CSF flow posterior to the spinal cord, excluding an arachnoid cyst (Fig. 1). The patient underwent a thoracic laminectomy T6–T8. No spinal cord monitoring was used. The dura was opened under the microscope and an atrophic spinal cord displaced to the left was visible. The myelum was incarcerated through a 2 cm wide anterolateral dural defect and had an exophytic edematous appearance (Fig. 2). In order to perform an anterior untethering, the dentate ligament was transected and the nerve roots were preserved. The spinal cord was gently mobilised out of the dural defect. Primary suturing of the dura was not possible without causing massive torsion of the spinal cord. It was therefore decided to position a sheet of collegeous membrane (Tutopatch®) anterior to the spinal cord which was wrapped around the spinal cord and approximated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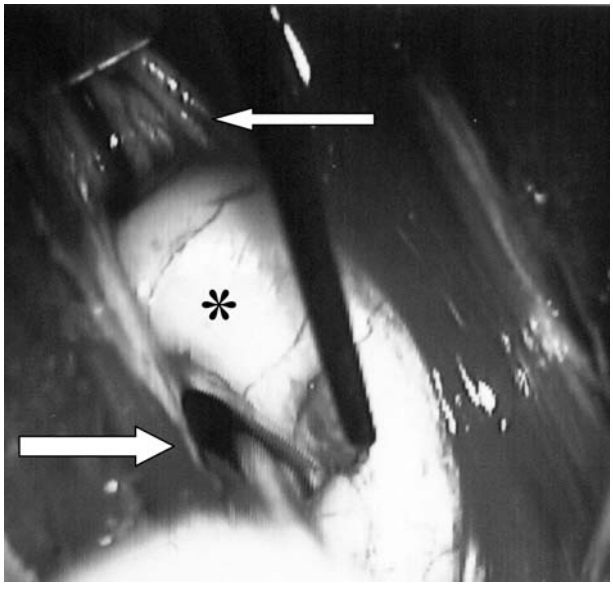

Fig. 2. Intra-operative view of patient 1 showing incarceration of the spinal cord through a 2 cm wide anterolateral dural defect. The myelum has an exophytic edematous appearance and is marked with an asterisk. Large arrow on the right shows the margin of the dural defect, the small arrow on the left shows the nerve roots

posteriorly. In this way the spinal cord was covered by a sleeve of dura graft to untether the spinal cord and prevent recurrent transdural herniation. The graft was sutured into the dura without causing constriction of the myelum. The dura was covered with Tissuecol® and the wound was closed in layers without external cerebrospinal fluid drainage. The initial post-operative neurological deficit was unchanged and there was no sign of cerebrospinal fluid leakage. Long-term observations are not available yet.

Case 2, a 43-year old man, presented with progressively ascending paresthesia and numbness of the right leg. The history regarding motor function and micturition was normal. Neurological examination revealed impairment of pain and temperature sensation below T7 at the right side. Position and vibration sense, motor function and reflexes were normal. MRI of the thoracic spine showed spinal cord atrophy at T4–T5 with anterior displacement and a dorsal cerebrospinal fluid collection. The radiological diagnosis was a dorsal arachnoid cyst. A laminectomy T4–T6 was performed and after opening the dura, a cerebrospinal fluid collection was discharged and no further exploration has been done. The postoperative neurological deficit was unchanged. One year later, however, he presented again with progressive weakness of the left leg, unchanged right-sided impairment of pain and temperature sense below T7, and bilateral Babinski signs. MRI showed unchanged anterolateral displacement of the spinal cord and, at this time, transdural spinal cord herniation was suspected. Moreover, the laminectomy had not been performed at the level of the displaced spinal cord. During surgical re-exploration the laminectomy was extended cranially. The dura was opened and the atrophic spinal cord was displaced to the left side. After transecting the dentate ligament, the spinal cord was mobilised gently out of the duradefect. As in case 1, the dura was not closed directly but a sling of Tutopatch® was placed anterior to the spinal cord encasing the myelum and sutured posteriorly with the autologeous dura. The postoperative MRI showed untethering of the spinal cord (Fig. 3) with a normal position in the spinal canal and the wrapped sheet of durapatch visible. The patient's neurological deficit was improved.

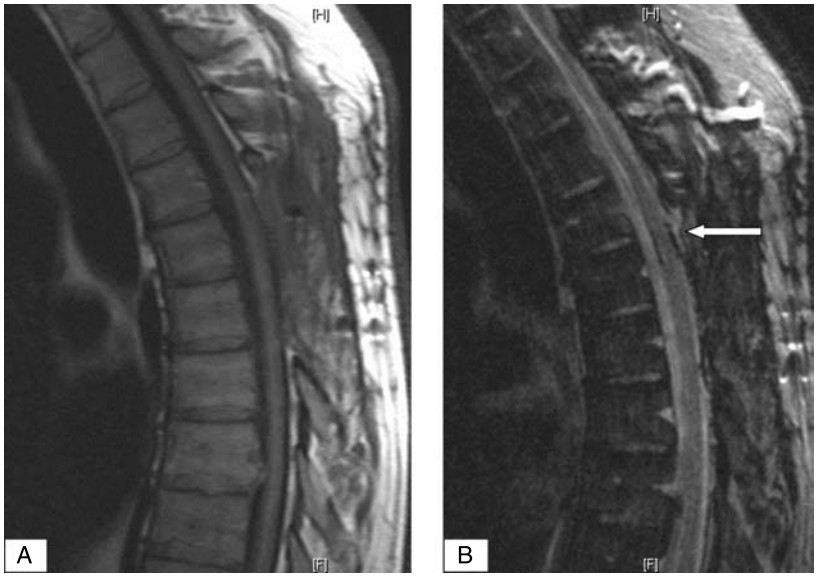

Fig. 3. (A) Post-operative sagittal T1 and (B) T2 weighted MR image show normal position of the spinal cord in the spinal canal. Note the hypointense signal around the cord (B, arrow), caused by the duroplasty

Fig. 1. (A) Sagittal T2 weighted MR image in patient 1 showing focal atrophy of the thoracic spinal cord. (B) Sagittal cardiac gated 2D FLASH image in patient 1 without (B) and with velocity encoding (C) showing CSF flow posterior to the area of displaced and focally atrophic thoracic spinal cord (arrows), making an arachnoid cyst unlikely. (D–G) Consecutive axial heavily T2 weighted 3D 'constructive interference in steady state' (CISS) images (1 mm thin) in patient 1 showing herniation of the spinal cord through a dural defect towards the epidural space. (H) Coronal reformat of axially acquired CISS images in patient 1 showing transdural herniation of the spinal cord to the epidural space on the left (arrow). The images nicely show the intact dura on the right (small arrows). (I, J) Sagittal reformat of axially acquired CISS images showing transdural herniation of the spinal cord to the epidural space. The images nicely show the herniation through the dural defect (arrows)

Discussion

Idiopathic transdural herniation of the spinal cord through a dural defect is an unusual cause of myelopathy. Most patients present with slowly progressive spastic paraparesis, sensory disturbances or Brown-Séquard syndrome [2, 3, 5, 6, 10, 11, 13, 14, 18, 23]. The clinical picture is caused by anterolateral herniation of the spinal cord with consequent damaging of the lateral funiculus. The initial symptoms are related to the lateral spinothalamic tract and, as the disease advances, the corticospinal tract herniates through the defect as well. This explains why, as in our cases, a complete Brown-Séquard syndrome may not be present. Another known but rare symptom is cerebrospinal fluid hypotension related headache in the upright position, probably present in our first case [8]. Women are affected more often (men:women ratio is 1:2) and the median age at presentation is 46 years [19].

Idiopathic transdural herniation of the spinal cord seems exclusively located at the thoracic level. The explanation may be that, due to the physiological thoracic kyphosis with ventral positioning of the spinal cord, transdural spinal cord herniation occurs whenever an anterior dural defect is present.

The diagnosis of idiopathic transdural spinal cord herniation is often missed or delayed, as demonstrated by our cases. Typically, a C-shaped kinking of the spinal cord is visible on conventional sagittal MRI with disappearance of the ventral subarachnoid space and widening of the dorsal subarachnoid space (Fig. 1). At the level of the herniation, the spinal cord is often atrophic.

The differential diagnosis of this radiological image mainly consists of a dorsal arachnoid cyst with dorsal compression of the spinal cord. However, modern high-resolution T2 imaging is able to show the dural defect with transdural herniation of the spinal cord in the anterior epidural space (Fig. 1). Phase-contrast cine MRI can also distinguish a dorsal arachnoid cyst from transdural spinal cord herniation by showing normal pulsatile CSF flow posterior to the spinal cord [4, 5]. CT-myelography may be helpful but is not strictly necessary given the superior MRI possibilities. Other differential diagnoses are myelitis transversa and glioma. In a few cases a biopsy was performed while in retrospect transdural spinal cord herniation was visible.

Several theories regarding the etiology of the anterolateral dural defect have been formulated. Isu *et al.* postulated pressure transmission by a dorsal arachnoid cyst [9]. During surgery, however, a thickened arachnoid membrane with a widened dorsal cerebral spinal fluid space was found and not a true arachnoid cyst [19]. Another hypothesis is a congenital duplication of the ventral dura leading to herniation of the spinal cord through the inner dura [1, 15]. This theory is based on operative findings but not supported by radiological nor pathological evidence of dura duplication [19]. Najjar *et al.* proposed an inflammatory reaction caused by ventral adhesion of the spinal cord to the dura resulting in a dural defect [14]. A posttraumatic dural defect and transdural herniation of the spinal cord in relation to a thoracic herniated disc are described [20, 22, 24]. Inoue *et al.* postulated the presence of a spontaneous dural fistula which gradually increases into a larger dural defect. Consequent cerebrospinal fluid leakage from a dural tear could explain the history of postural headache [8]. Eventually, the spinal cord is herniated through the dural defect which stopped cerebrospinal fluid leakage and related postural headache, but led to progressive myelopathy. Our first patient had a similar history.

Most patients are treated surgically although in some patients conservative management has merit. In the absence of progressive signs and symptoms, clinical observation and MRI surveillance is legitimate [2, 12]. The aim of surgical treatment is reduction of cord herniation by closing the dural defect, with or without a patch, or widening the defect to prevent spinal cord compression. Direct closure of the dural defect through an anterior approach has first been described by Wortzman *et al.* [24]. Most surgeons perform a posterior decompressive laminectomy with reduction of the spinal cord herniation and repair of the dural defect [1, 5, 12, 14, 16, 23]. In all cases, the dentate ligament is transected in order to mobilise the spinal cord. The dural defect is located anterolateral and therefore the ventral site of the spinal cord needs to be inspected in order to prevent misdiagnosis. The spinal cord is often incarcerated through the dural defect. After reducing the herniation, the spinal cord often has an edematous reddish appearance as in glioma. Some authors have performed a biopsy [10, 17]. Gwinn and Henderson described 3 patients in whom the dural defect has been closed through a posterolateral transpedicular approach [7]. Another technique mentioned by Watanabe *et al.* is enlarging the dural defect to prevent spinal cord incarceration [21].

The technique we used, placing a sheet of dural graft around the spinal cord, has not been described earlier. Primary suturing the dural defect with excessive spinal cord manipulation and consequent deterioration is prevented. However, retethering of the spinal cord due to

constriction of the graft might occur whenever the wrapping is too tight. In case of post-operative deterioration we postulate instant MRI to document this possibility, followed by surgical exploration and loosening the graft.

The outcome after surgical intervention is favorable in most patients with Brown-Séquard syndrome and less favorable in patients with spastic paraparesis [1–5, 8, 9, 11, 14–17, 19, 21, 22, 24]. Long-term postoperative follow-up reports are sparse, but retethering and development of a syrinx is documented in some cases [2]. Therefore, longer follow-up is needed to establish the natural course versus postoperative outcome of idiopathic transdural spinal cord herniation.

Conclusions

Idiopathic transdural spinal cord herniation is an unusual but increasingly recognised cause of progressive thoracic myelopathy. High-resolution T2 MRI is the method of choice to directly visualise the herniation of the spinal cord through a dural defect. The two main surgical strategies consist of closure of the dural defect with a graft, or simply widening the aperture to prevent incarceration. We add another surgical technique to untether the spinal cord by placing a sheet of dura graft around the myelum to prevent recurrent spinal cord herniation.

References

1. Aizawa T, Sato T, Tanaka Y *et al* (2001) Idiopathic herniation of the thoracic spinal cord: report of three cases. Spine 26: E488–E491
2. Ammar KN, Pritchard PR, Matz PG *et al* (2005) Spontaneous thoracic spinal cord herniation: three cases with long-term follow-up. Neurosurgery 57: E1067; discussion E1067
3. Borges LF, Zervas NT, Lehrich JR (1995) Idiopathic spinal cord herniation: a treatable cause of the Brown-Séquard syndrome – case report. Neurosurgery 36: 1028–1032; discussion 1032–1033
4. Brugieres P, Malapert D, Adle-Biassette H *et al* (1999) Idiopathic spinal cord herniation: value of MR phase-contrast imaging. AJNR Am J Neuroradiol 20: 935–939
5. Cellerini M, Bayon S, Scazzeri F *et al* (2002) Idiopatic spinal cord herniation: a treatable cause of Brown-Séquard syndrome. Acta Neurochir (Wien) 144: 321–325
6. Ewald C, Kuhne D, Hassler WE (2000) Progressive spontaneous herniation of the thoracic spinal cord: case report. Neurosurgery 46: 493–495; discussion 495–496
7. Gwinn R, Henderson F (2004) Transdural herniation of the thoracic spinal cord: untethering via a posterolateral transpedicular approach. Report of three cases. J Neurosurg Spine 1: 223–227
8. Inoue T, Cohen-Gadol AA, Krauss WE (2003) Low-pressure headaches and spinal cord herniation. Case report. J Neurosurg 98: 93–95
9. Isu T, Iizuka T, Iwasaki Y *et al* (1991) Spinal cord herniation associated with an intradural spinal arachnoid cyst diagnosed by magnetic resonance imaging. Neurosurgery 29: 137–139
10. Kumar R, Taha J, Greiner AL (1995) Herniation of the spinal cord. Case report. J Neurosurg 82: 131–136
11. Marshman LA, Hardwidge C, Ford-Dunn SC *et al* (1999) Idiopathic spinal cord herniation: case report and review of the literature. Neurosurgery 44: 1129–1133
12. Massicotte EM, Montanera W, Ross Fleming JF *et al* (2002) Idiopathic spinal cord herniation: report of eight cases and review of the literature. Spine 27: E233–E241
13. Masuzawa H, Nakayama H, Shitara N *et al* (1981) Spinal cord herniation into a congenital extradural arachnoid cyst causing Brown-Séquard syndrome. Case report. J Neurosurg 55: 983–986
14. Najjar MW, Baeesa SS, Lingawi SS (2004) Idiopathic spinal cord herniation: a new theory of pathogenesis. Surg Neurol 62: 161–170; discussion 170–171
15. Nakazawa H, Toyama Y, Satomi K *et al* (1993) Idiopathic spinal cord herniation. Report of two cases and review of the literature. Spine 18: 2138–2141
16. Pereira P, Duarte F, Lamas R *et al* (2001) Idiopathic spinal cord herniation: case report and literature review. Acta Neurochir (Wien) 143: 401–406
17. Sioutos P, Arbit E, Tsairis P *et al* (1996) Spontaneous thoracic spinal cord herniation. A case report. Spine 21: 1710–1713
18. Srinivasan A, Bourque P, Goyal M (2004) Spontaneous thoracic spinal cord herniation. Neurology 63: 2187
19. Tekkok IH (2000) Spontaneous spinal cord herniation: case report and review of the literature. Neurosurgery 46: 485–491; discussion 491–492
20. Urbach H, Kaden B, Pechstein U *et al* (1996) Herniation of the spinal cord 38 years after childhood trauma. Neuroradiology 38: 157–158
21. Watanabe M, Chiba K, Matsumoto M *et al* (2001) Surgical management of idiopathic spinal cord herniation: a review of nine cases treated by the enlargement of the dural defect. J Neurosurg Spine 95: 169–172
22. Watters MR, Stears JC, Osborn AG *et al* (1998) Transdural spinal cord herniation: imaging and clinical spectra. AJNR Am J Neuroradiol 19: 1337–1344
23. White BD, Tsegaye M (2004) Idiopathic anterior spinal cord hernia: under-recognized cause of thoracic myelopathy. Br J Neurosurg 18: 246–249
24. Wortzman G, Tasker RR, Rewcastle NB *et al* (1974) Spontaneous incarcerated herniation of the spinal cord into a vertebral body: a unique cause of paraplegia. Case report. J Neurosurg 41: 631–635

Comment

The authors report two cases of idiopathic spinal cord herniation and briefly sketch their surgical approach applied in those cases. They discuss clinical, imaging, and possible etiological features of this condition. The literature is reviewed with regard to surgical techniques applied. They also touch upon possible advantages of their own approach. They conclude that high-resolution T2 MRI is the best imaging modality, and they also seem to conclude that their new technique valuably adds to previous surgical approaches as found in the literature.

This is a valuable, well written case report and review of the literature about a rare condition which every neurosurgeon may nevertheless be confronted with some time, and which plays a role for differential diagnostic considerations in myelopathy cases. The cases are adequately presented, the figures are mainly instructive and well chosen. The discussion is very well structured and concise.

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