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GRAND ROUNDS

Idiopathic spinal cord herniation: consideration of its pathogenesis based on the histopathology of the dura mater

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Abstract

Introduction We present a patient with idiopathic spinal cord herniation (ISCH) whose dura mater was histopathologically examined to elucidate its pathogenesis.
Case report A 33-year-old previously healthy man presented with progressive walking difficulty, spasticity of the right lower leg, and hyperesthesia below the right chest. Neuroimaging revealed right ventral displacement of the spinal cord at T5–6. The diagnosis was ISCH and he underwent release of the herniation from the ventral dural opening. Dural biopsy at the edge of the ventral opening and in the dorsal durotomy was performed. Postoperatively, his gait was improved. Histopathological examination of the ventral dural specimen showed non-specific degeneration, i.e., loose arrangements of collagen fibers, edematous changes, minor inflammatory cell infiltration, and angiogenesis. The specimen from the dorsal durotomy was normal.
Conclusion It is unclear whether the observed degeneration besides the ventral opening was the primary cause of ISCH or reflected secondary changes resulting from cumulative damage due to pulsation of the herniated spinal cord. However, the degeneration limited to the ventral opening suggests that ISCH was a local event in an individual with a normal dural theca.

Keywords Idiopathic spinal cord herniation · Dura mater · Histopathology · Pathogenesis

Case presentation

This 33-year-old man with no apparent relevant medical history including trauma experienced progressive walking difficulty for 6 months prior to visiting our institution. Physical examination revealed spasticity without weakness in the right lower leg. His patellar and Achilles tendon reflexes were hyperactive on the right; pathological reflexes were negative. There was hyperesthesia below the right T10 dermatome; his pain-, vibration-, and position sensations were normal. No bladder or bowel disturbances were found.

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Diagnostic imaging findings

Magnetic resonance imaging (MRI) revealed right ventral displacement of the spinal cord with an acute angle at the T5–6 intervertebral level; there were no findings suggesting a cystic lesion dorsal to the cord (Fig. 1a). On computed tomographic myelogram (CTM) just after contrast injection, there was no opacification in the subarachnoid space ventral to the displaced cord; the space dorsal to the cord was well opacified (Fig. 1b).

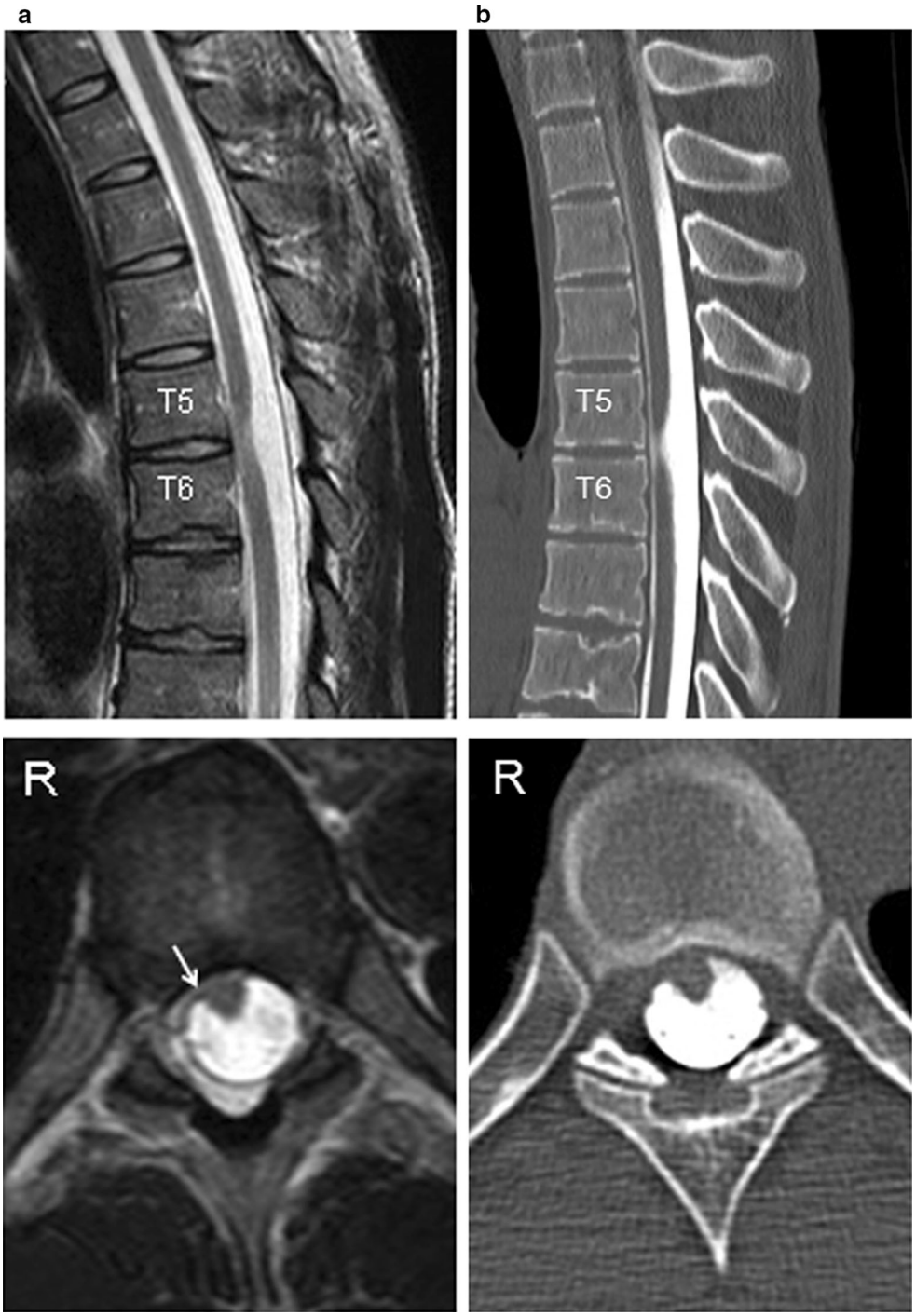

Fig. 1 Preoperative 3-tesla *T*2-weighted MRI (**a**), and CTM obtained just after contrast injection (**b**). Sagittal- (*upper row*) and axial images at the T5–6 intervertebral level (*lower row*) demonstrate ventral displacement of the spinal cord with an acute angle at the *T5–6* intervertebral level. Axial images show right ventral displacement of the cord, obstruction of the subarachnoid space ventral to the cord, and a thin ventral epidural space (*arrow*). There are no findings suggesting a cystic lesion dorsal to the cord

The diagnosis was idiopathic spinal cord herniation (ISCH).

Historical review of the condition, epidemiology, diagnosis, pathology, and differential diagnosis

ISCH, first reported by Wortzman et al. in 1974 [1], is characterized by ventral displacement of the spinal cord through an opening in the dura mater. Advances in imaging technology have led to the identification of ISCH in almost 200 adults [2–4]; their age ranged from 21 to 78 years (mean 51 years). Its incidence (64%) is higher in women than men [2]. ISCH is localized at the thoracic spine, dominantly at T3–T7 (80%), and at the level of intervertebral discs (68%) [2]. In general, the dural defects are seen as a ventral single lesion at 1 or 2 segments. Dorsal- [5], separated multiple- [6], and lesions extending over three segments [2] are extremely rare. The progressive lower extremity symptoms of ISCH mainly present as Brown-Séquard syndrome (66%) and spastic paraparesis (30%). Motor or sensory disturbance alone in the unilateral lower extremity [2, 3] is rare.

At present, a preoperative diagnosis of ISCH is made when focal ventral displacement of the spinal cord and obstruction of the subarachnoid space ventral to the displaced cord are observed on *T*2-weighted MRI scans and CTM [3]. On sagittal images, the cord was classified as type K (ventral spinal cord kinking 66.7%), type D (spinal cord disappearance at the herniated site 25%), and type P (protrusion of the ventral aspect of the spinal cord without kinking at the dorsal aspect 8.3%) [7]. On axial images, the location of the herniation was classified as type C (central, i.e., midline 50%) and type L (lateral 50%). A spinal cord appearance of type P was correlated with good- and type C herniation with poor postoperative recovery.

The dural opening in ISCH is longitudinal and oval, and internal partial- or whole-layer penetration of the membrane is seen [2, 3]. Mechanisms underlying the development of a dural opening and associated spinal cord herniation have been proposed. They are a history of trauma [1], pressure erosion by a dorsal arachnoid cyst compressing the spinal cord ventrally [8], thoracic disc herniation [1], and the spread of systemic inflammation [9] that result in shearing and weakening of the dural fibers. Congenital pathologies (ventral meningocele [1], dural duplication [10], and extradural arachnoid cysts [11]) have also been suggested. The physiological proximity of the ventral dura mater in the kyphotic thoracic spine to the spinal cord may be an anatomic contributing factor [3]. Direct contact between the spinal cord and the pre-existing dural opening leads to arachnoid adhesions. The combination of pulsation and negative pressure in the epidural space wedges the spinal cord and results in cerebrospinal fluid leakage into the opening and its widening over time [12].

The spinal dura mater is a possible key structure implicated in the occurrence of ISCH. Histological and biomechanical studies of the spinal dura mater may shed light on the features and background of the dural opening in ISCH. As the collagen fibers, i.e., the main supporting tissue, are dominantly oriented longitudinally, the dura mater has higher resistance in this than the transverse direction [13]. Consequently, if shearing force is a causative factor in ISCH, the dural opening can be expected to be longitudinally oval. The looser arrangement of the dural collagen fibers near the intradural side than the epidural side [14], i.e., the potential fragility near the intradural side, may be a background for the formation of a dural opening by some increased intradural force. Depending on the depth of the opening, a duplicated dura mater (intramural pouch) or a penetrated dura mater is formed. Less elastin, a connective tissue that helps to retain elasticity, is present in the ventral part than the dorsal part of the spinal dura mater [15]. This may explain why the site of ISCH is primarily ventral. However, due to the equality of the elastin content on the spinal levels [15] and the lack of data on its content in individuals of different ages and genders, we cannot explain the predominant occurrence of ISCH in the thoracic spine and in middle-aged women.

Histopathological findings on the dura mater in ISCH showed that specimens harvested from the inner- or the inner and outer layers of duplicated membranes at the ventral opening were compatible with the dura mater [7, 10, 16–18]. In most instances, the arrangement of fibers comprising the dura was normal; wavy fibers were seen occasionally [16]. No patients presented with evidence of inflammation and there were no specific pathological findings to identify the pathogenesis of ISCH.

A preoperative differential diagnosis between ISCH and other pathologies with ventral displacement of the spinal cord, especially dorsal arachnoid cysts, is required for proper surgical planning. When ISCH is misdiagnosed as other pathologies, the herniation may not be identified during surgery; this results in an unfavorable postoperative course. According to a systematic review [4], among 174 patients, 37 (21%) were misdiagnosed and 30 of the 37 (81%) underwent the wrong kind of surgery. ISCH was identified intraoperatively in only 7 of the 30 patients (23%); the 23 other initially misdiagnosed patients (77%) required a second operation. Patency of the subarachnoid space ventral to the displaced cord and isolation of the subarachnoid space dorsal to the cord suggest a dorsal arachnoid cyst [3]. Dorsal arachnoid cysts are commonly

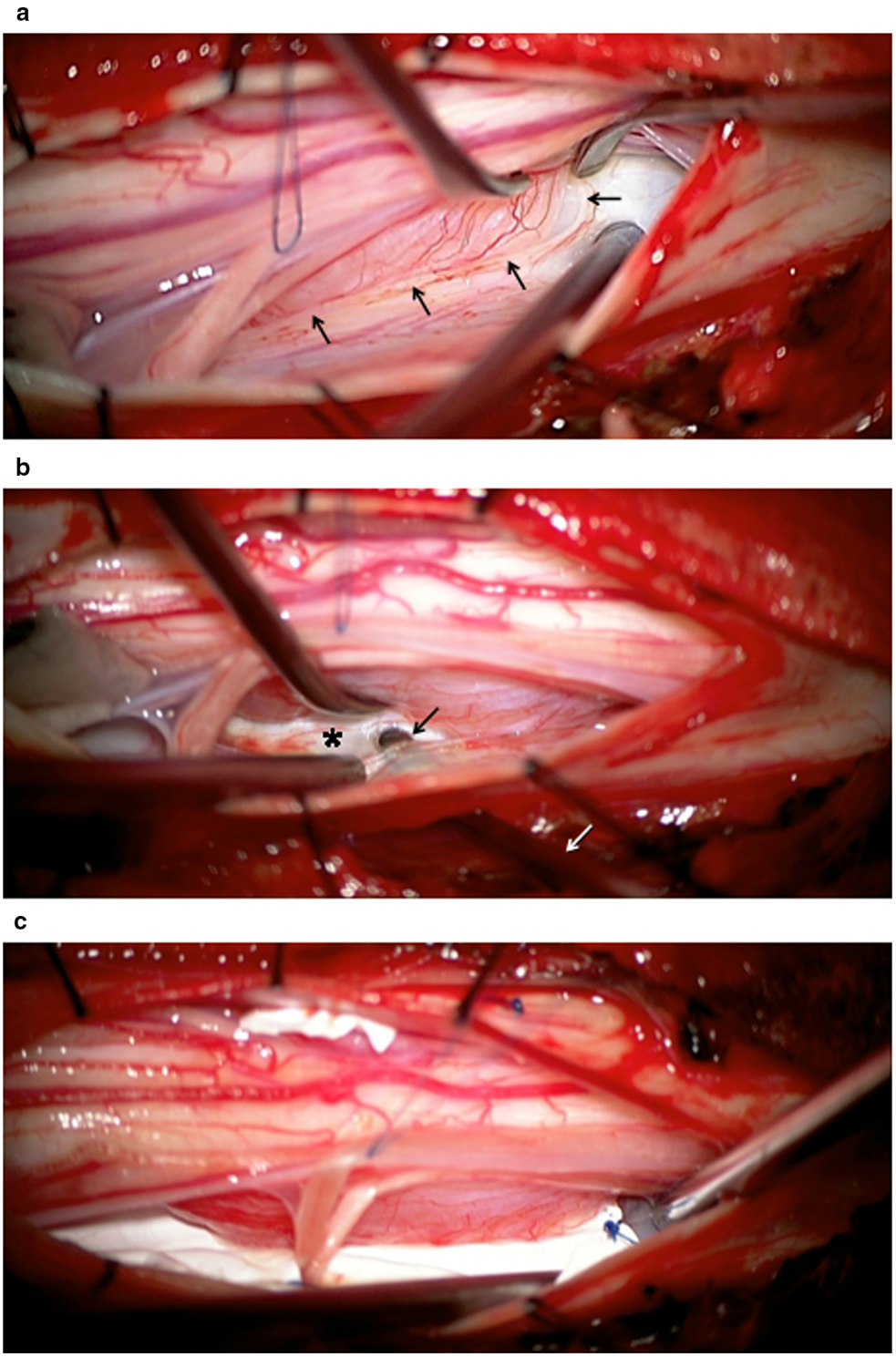

Fig. 2 Intraoperative views. The spinal cord herniating into the ventral dural opening at the T5–6 intervertebral level was exposed via the right posterolateral subdural epiarachnoid approach. The rostral dural opening was round (**a**). *Arrows* indicate the edge of the dural opening. After releasing the herniation, the posterior longitudinal ligament (*asterisk*) was visible at the bottom of the dural opening (**b**). A dissector (*arrows*) inserted from the lateral epidural space readily reached the bottom. To complete the procedure, the spinal cord was lifted with an expanded polytetrafluoroethylene sheet (hammock method) (**c**)

demonstrated as a partial block on myelograms; occasionally, they are filled with contrast medium [19]. On delayed CTM, cysts without opacification on myelograms can appear to be filled with contrast medium [19]. Cine-mode MRI depicting stasis in the pulsatile cerebrospinal fluid flow at the site of the cyst [20], constructive interference in steady state (CISS) MRI showing a site of communication between the cysts and the subarachnoid space [21], and the visualization of septa dividing the lesion into multiple cysts on CISS MRI [21] are diagnostically useful, as are phase-contrast MRI, CTM, and anomalous nerve root findings on axial images [3]. The roots traverse the dorsal subarachnoid space in ISCH; in the presence of a dorsal arachnoid cyst, they course along the periphery of the lesion. A possible association with dorsal arachnoid cyst, reported in 20–25% of ISCH patients [4], should be considered in a differential diagnosis.

Rationale for treatment and evidence-based literature

Most patients with ISCH who were conservatively treated experienced no change or worsening of their neurological symptoms [3, 4]. Spontaneous resolution of the herniation [22] can occur but is usually unexpected. Surgery yielded favorable results; 74% of patients reported symptom improvement [4]; motor function improved in 75% of patients with Brown-Séquard syndrome and in 55% of patients with spastic paraparesis [2].

The aim of surgery is release of the herniated spinal cord and treatment of the dural opening to prevent recurrent herniation [2, 3]. There are two strategies for addressing the dural opening. One is closing the opening by direct suture of the dura [1], inserting filling materials into the opening [3], or covering the opening with a patch [2, 9, 11]. The other, applicable to an opening in the inner layer of the duplicated dura mater, is widening the dural opening to avoid the recurrence of cord wedging [7, 10, 11, 16–18]. Widening of the dural opening yielded better motor function improvement than patch closure (82 vs 64%) [2]. Postoperative worsening of the symptoms is thought to be attributable to excessive retraction of the spinal cord during closure of the ventral dural opening. To reduce the risk of iatrogenic damage to the spinal cord during exposure of the ventral dural opening, the posterolateral approach has been recommended [3]. Laminectomy or hemilaminectomy, costotransversectomy, and lateral durotomy can be combined [2]. The transthoracic approach is not widely used and advances in diagnostic preoperative imaging render biopsy of the bulge of the herniated spinal cord to rule out tumors [1] unnecessary.

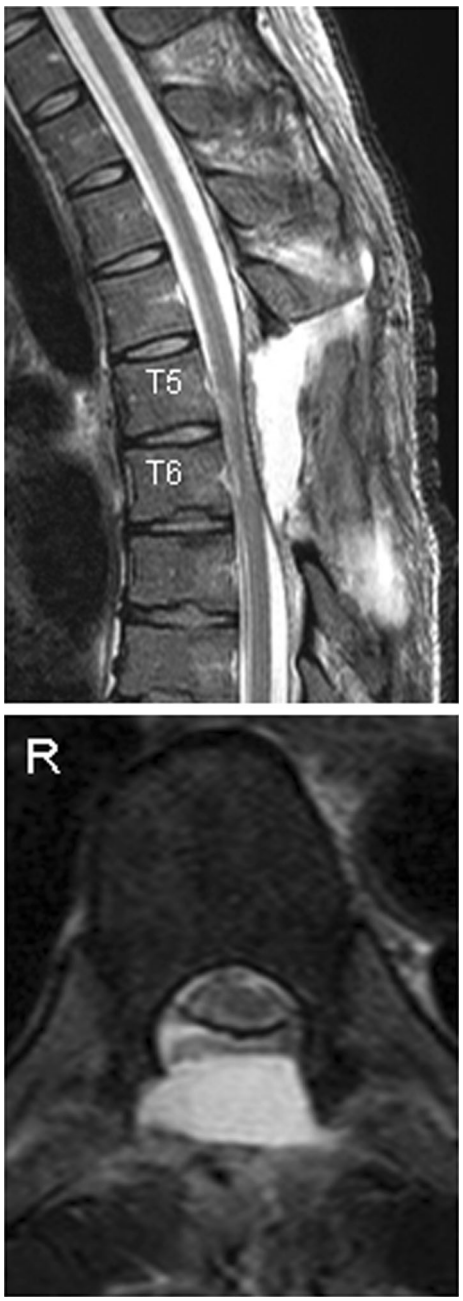

Fig. 3 Postoperative *T2*-weighted MRI. Sagittal- (*upper row*) and axial images at the *T5–6* intervertebral level (*lower row*) confirm restoration of the course of the cord. The subarachnoid space ventral to the cord is observed

In our patient, we did not consider conservative treatment, because it is ineffective [3, 4] and because his symptoms were progressive at the time of presentation. We chose closure of the opening by the less invasive hammock method [23]. The risk for iatrogenic injury to the epidural venous plexus ventral to the dural opening off the midline ruled out widening of the dural opening in this case.

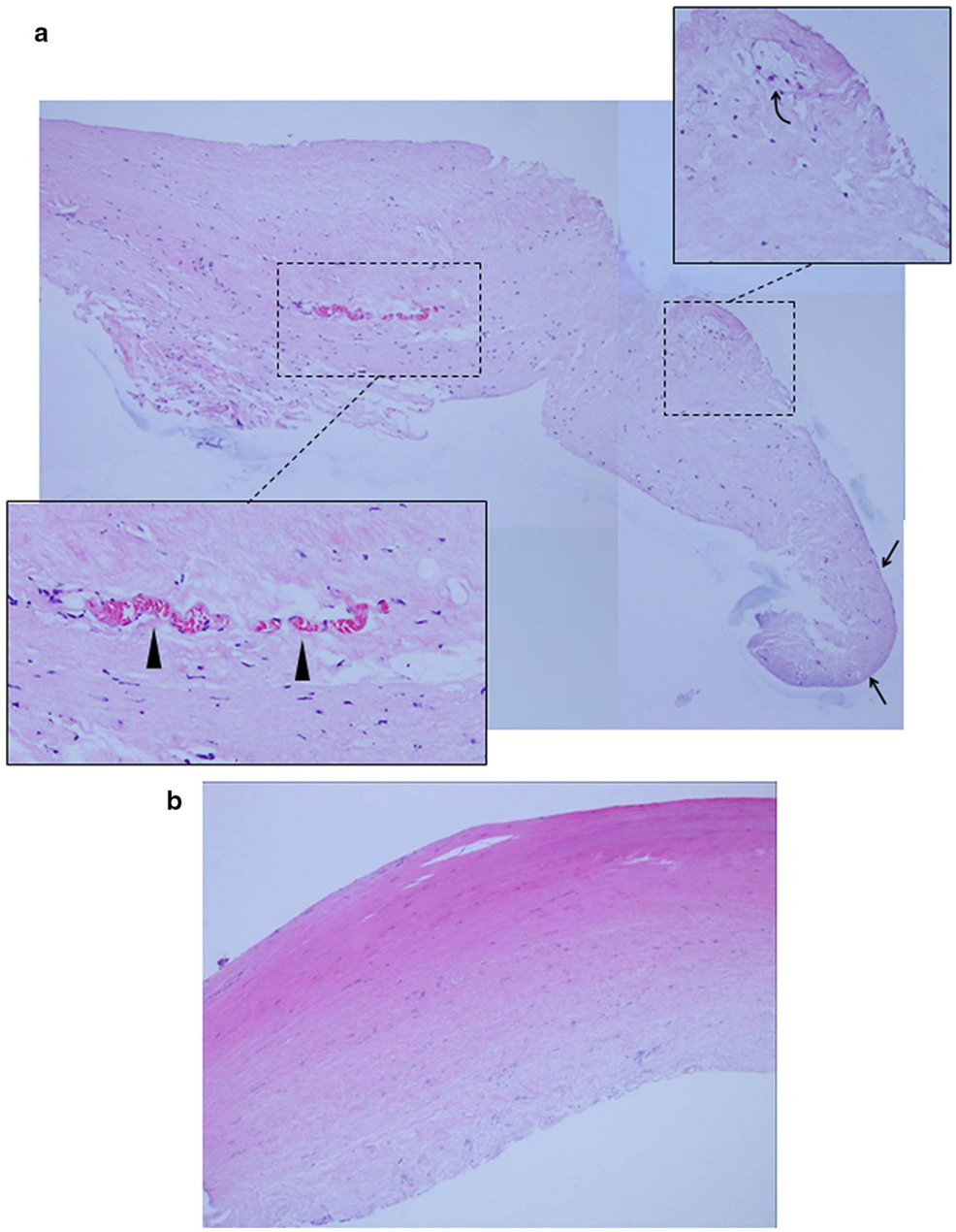

Fig. 4 Photomicrographs of the dura mater. At the edge of the ventral opening (**a**), a loose arrangement of collagen fibers and edematous change, indicative of degeneration, is observed (hematoxylin-eosin stain; magnification ×40). The epidural side is at the *bottom*. *Arrows* indicate the edge facing the herniated spinal cord. The magnified photographs (×100) reveal macrophages (*curved arrow*) and angiogenesis (*arrowheads*). In the dorsal dura mater (**b**), the collagen fiber arrangement is normal; it is tighter near the epidural side than the intradural side. There is no evidence of inflammation. The epidural side is at the *top* (hematoxylin–eosin stain; magnification ×40)

Procedure

We performed laminectomy from T5 to T6, predominantly on the right side for a posterolateral approach. The normal-appearing dorsal dura was opened on the right to avoid tearing the arachnoid membrane and postoperative cerebrospinal fluid leakage. The site of the spinal cord herniation in the ventral aspect of the dural theca was exposed via a subdural epiarachnoid approach with lifting of the right dentate ligament (Fig. 2a). The herniated cord bulge, covered by the arachnoid membrane, was released gently; the tissue appeared to be congestive. The dural opening was a longitudinal oval approximately 20 × 5 mm in diameter. The posterior longitudinal ligament was visible at the bottom of the dural opening. A dissector inserted from the lateral epidural space readily reached the bottom

(Fig. 2b) and whole-layer penetration of the dura was confirmed. A minute biopsy of the dura was performed at the edge of the ventral opening and in the dorsal durotomy. Using the hammock method [23], a 0.1-mm-thick expanded polytetrafluoroethylene sheet (GORE, PRECLUDE®; W.L. Gore & Associates, Inc., Flagstaff, AZ, USA) was placed to lift the spinal cord over the dural opening, both lateral ends of the sheet were sutured on the lateral dural walls (Fig. 2c), and the dorsal durotomy was closed. Intraoperative motor- and somatosensory evoked potentials of the lower extremities showed no deterioration throughout the surgical procedures.

Outcome, follow-up

In the course of the first 3 postoperative weeks, his spasticity of the right lower leg improved gradually and he could walk independently; his sensory disturbance persisted. Postoperative MRI confirmed restoration of the course of the cord and the ventral subarachnoid space was visualized (Fig. 3). Although his sensory disturbance persisted, 1 year later, he was able to ascend and descend stairs using handrails.

Histopathological examination of the dura mater at the edge of the ventral opening revealed degeneration reflected by a loose arrangement of collagen fibers and edematous changes. Minor inflammatory cell (macrophage) infiltration and angiogenesis were observed (Fig. 4a). The dorsal durotomy specimen revealed a normal arrangement of collagen fibers; they were tighter near the epidural side than the intradural side. There was no evidence of inflammation (Fig. 4b). The small volume of both specimens disallowed additional immunohistochemical and electron micrographic studies to demonstrate the paucity of collagen fibers and inflammatory cell infiltration.

Conclusion

Ours is the first histopathological comparison of the ventral and dorsal dura in a patient with ISCH. Degeneration limited to the ventral opening, while not peculiar to the pathogenesis of ISCH, was at least suggestive of his ISCH being a local event in the presence of a normal dural theca. Further histopathological studies of the dura mater may help to elucidate the pathogenesis of ISCH.

Compliance with ethical standards

Conflict of interest The authors have no personal financial or institutional interest in any of the drugs, materials, or devices cited in this paper.

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