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CASE REPORT

Spinal extradural meningeal cyst and Scheuermann's disease: coincidence or causative factor?

Ho-Young Park · Sun-Ho Lee · Eun-Sang Kim · Whan Eoh

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Introduction

Extradural meningeal cysts, previously called extradural arachnoid cysts, are relatively rare lesions of the spinal canal and are uncommon causes of spinal cord compression in children and adolescents [4, 15]. They are extradural outpouchings of arachnoid tissue that communicate with the intraspinal subarachnoid space through a small dural defect [5]. According to the classification system by Nabors et al. [16], spinal meningeal cysts are divided into three major categories: extradural cysts without spinal nerve root fibers (type I), extradural cyst with spinal nerve root fibers (type II), and intradural cysts (type III). Type I meningeal cysts are further divided into extradural meningeal cysts (type IA) and sacral meningoceles (type IB).

The etiology of spinal extradural meningeal cysts remains unclear. Although some can be acquired from trauma, infection, or inflammation, the majority of dural defects are thought to be congenital in nature [3]. In a few reported cases, authors suggested that a type IA cyst (extradural meningeal cyst) may be combined with Scheuermann's disease (kyphosis dorsalis juvenilis) [4, 10]. In this report, we present a rare case of spinal cord compressing type IA spinal extradural meningeal cyst with Scheuermann's disease in an adolescent patient.

H.-Y. Park · S.-H. Lee (✉) · E.-S. Kim · W. Eoh
Department of Neurosurgery, Samsung Medical Center,
Sungkyunkwan University, School of Medicine,
50 Ilwon-dong, Gangnam-gu,
Seoul 135-710, South Korea
e-mail: sobotta72@hotmail.com

Case report

A 15-year-old male presented with progressive ataxia and gait disturbance for 15 days. In his past history, the patient had a small head circumference at birth and had surgery for unilateral cryptorchidism. He also showed a developmental delay and mental retardation from the age of two. Fifteen days before admission, gait disturbance developed and he had also progressive difficulty in defecating 3 days before admission.

On physical examination, short stature (<3 %), low body weight (25–50 %), small head circumference (<30 %), and tic-like movement were noted in the right hand. On neurological examination, he showed grade 4 spastic paraparesis including upper motor neuron signs in both lower extremities. His deep tendon reflexes were hyperactive and his gait was severely ataxic. Laboratory tests including complete blood count, serum chemistry profile, electrolyte profile, erythrocyte sedimentation rate, C-reactive protein, and routine urinalysis were normal.

On radiologic evaluation, plain spinal X-ray and computerized tomography revealed irregular narrowing of the disc spaces, irregular endplates, and kyphosis, which are consistent with Scheuermann's disease. Whole spine magnetic resonance imaging (MRI) revealed a dorsally located 14.5×7.4×75-mm-sized cerebrospinal fluid (CSF) intensity cystic mass at the T12–L1 level which was noted to extend from the epidural space into both neural foramens. The cystic lesion had low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and no contrast enhancement (Fig. 1a, b). His thecal sac and spinal cord were compressed. Also, mild disc bulging on

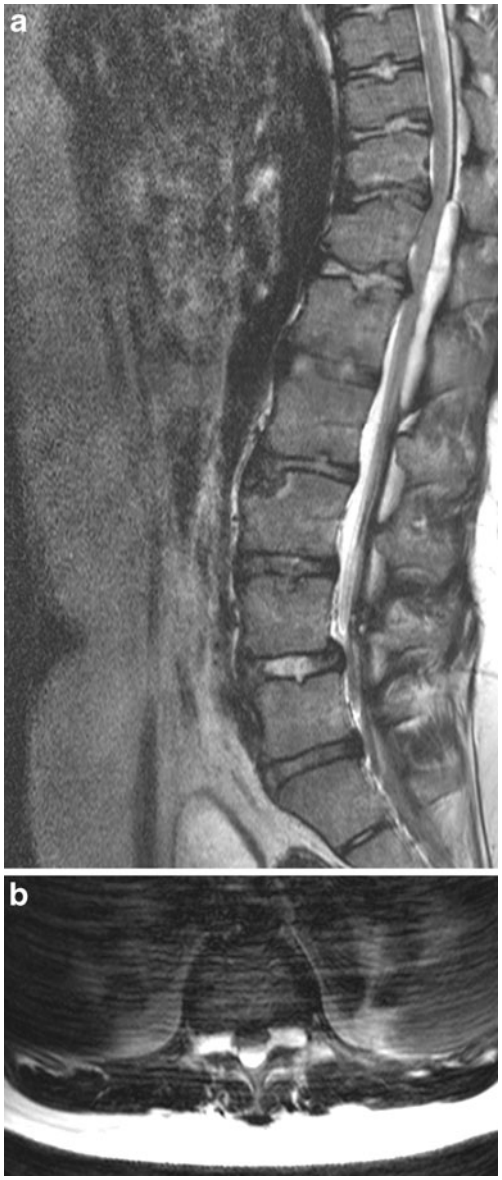

Fig. 1 Preoperative **a** sagittal T2 and **b** axial T2-weighted magnetic resonance imaging of the patient

T12–L1 was seen, and a signal change on spinal cord was not obvious.

We performed a laminectomy to expose the spinal mass on T11–12. After removal of a ligamentum flavum, a thin-walled 7.5-cm-sized extradural cystic mass occupying the spinal canal was found. CSF-like cystic fluid was aspirated, and then the cyst wall was totally removed. Two small defects, thought to be a connection between the cyst and subarachnoid spaces, were seen on both sides of the dura below both the T12 nerve roots. Furthermore, a pulsatile, breath-synchronous outflow of CSF was seen on these dural defects. They were repaired with a primary suture (Fig. 2). In addition to wide laminectomy with excessive facetectotmy, T12–L1 posterior screw fixation was performed. On pathological examination, the cyst wall showed fibrous tissue focally admixed with a few glandular structures of epithelial cells, consistent with an arachnoid membrane (Fig. 3).

The postoperative course was uneventful and the patient's signs and symptoms steadily improved without perioperative complications. Although the patient experienced a slight gait disturbance during the follow-up period, the patient was able to walk without difficulties and motor grade was normalized at follow-up examination 12 months after surgery.

Discussion

Type 1A spinal extradural meningeal cysts are relatively rare lesions of the spinal canal and uncommon causes of spinal cord compression in children and adolescents [4, 15]. This type of meningeal cyst accounts for about 1 % of all spinal tumors [2]. It has a male predilection and their peak incidence is in the second decade of life. Approximately 65 % of these lesions affect the thoracic spine, particularly the middle to lower thoracic regions, followed by the thoracolumbar (12 %), lumbar and lumbosacral (3 %), sacral (7 %), and cervical (3 %) spine [3]. Most of the lesions are located posteriorly in the spinal canal and can partially protrude into the adjacent neural foramen [2].

The causes of extradural meningeal cysts are unclear. They are known to be associated with trauma, surgery, arachnoiditis, and neural tube defects [15, 17]. In many cases, however, there is no underlying cause and most of the non-traumatic spinal extradural meningeal cysts are thought to be a result of congenital dural defects [14, 15]. We postulated a congenital contributing factor in our case because our patient revealed several findings including short stature, small head circumference, developmental delay, and mental retardation. In addition, the patient harbored Scheuermann's disease. Although the exact cause of this pathologic condition has not been fully proved, some studies have suggested a dominant mode of inheritance with incomplete penetrance [7]. In addition, previous report found an association between genetic disease and Scheuermann's disease [13]. It seems that dural defect develops as a result of insufficiency in mesodermal tissue of dura due to congenital malformation in patients with Scheuermann's disease, although no evidence of such occurrence is shown between Scheuermann's disease and meningeal cyst.

Meanwhile, it has been proposed that spinal extradural meningeal cysts can cause Scheuermann's disease by cystic compression and occlusion of the venous channels draining the vertebral bodies [4, 6]. In summarizing the literature, Wise and Foster [18] found dorsal kyphosis to be reported in 19 of 33 patients with spinal extradural cysts. After these

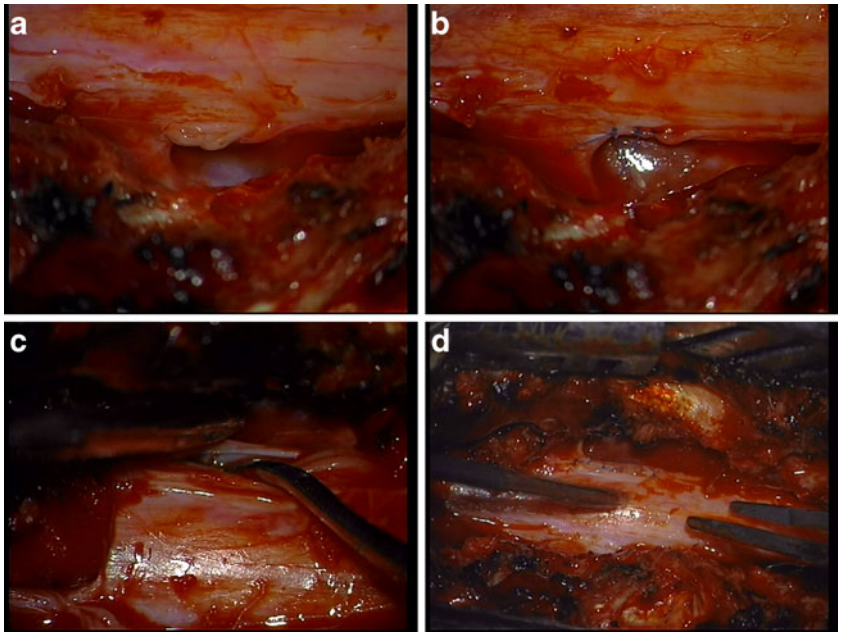

Fig. 2 Intraoperative photography obtained after thoracic laminectomy of a spinal extradural arachnoid cyst with a communicating pedicle at the site of the dural defect below both T12 nerve roots. **a** Left dural defect and **b** after repair. **c** Right dural defect and **d** after repair

reports, more authors presented similar cases [8, 10]. Because our patient had a spinal extradural meningeal cyst and local signs of Scheuermann's disease, including kyphosis with the apex at the T12–L1 level as well as multiple Schmorl's nodules, a similar mechanism might be suspected. Progressive kyphosis in extradural meningeal cysts combined with Scheuermann's disease may occur when the cysts are untreated throughout the adolescent growth period. When pediatric patients show symptoms of cord compression or when plain X-ray shows clue of Scheuermann's disease, surgeons should not forget to include spinal extradural meningeal cyst in their differential diagnosis.

In asymptomatic patients, conservative treatment or observation rather than surgical treatment is recommended [17]. Cases in which neurologic deterioration or persistent pain is attributable to spinal extradural meningeal cyst are surgically indicated. The standard surgical method for spinal extradural meningeal cyst is complete resection of the cyst wall and closure of the dural defects after laminectomy of the affected vertebra. Although myelography or intraoperative examination demonstrated communication between the extradural cysts and the subarachnoid space in approximately half of the published cases [11, 12], some reports have suggested that a communication exists in nearly all cases; Myles et al. [15] emphasized the importance of dural repair on treatment of spinal extradural meningeal cysts. In our case, two defects of the dura were found. Even though the cyst was single and a nongiant type, neurosurgeons should be reminded of the possibility of multiple dural defects which requires meticulous exploration.

The outcome of surgically treated patients with spinal extradural meningeal cysts depends on the patient's age as well as the duration and degree of neurological damage [1, 9]. Immediate pain relief after surgery is reported in most cases, but recurring back pain with and without radiation is common in long-term follow-up. To help these patients, early recognition is necessary along with surgical treatment, which can be improved. The widespread availability of MRI has also greatly improved the diagnostic possibilities.

Conclusion

Spinal extradural meningeal cysts are uncommon which may be combined with Scheuermann's disease, resulting in cord compression and progressive kyphosis, most commonly in

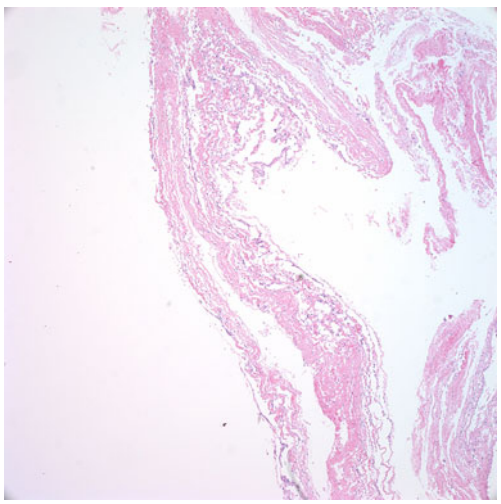

Fig. 3 Microscopy of the cyst wall demonstrates fibrocollagenous layers with a single-cell lining. H & E, original magnification ×50

young patients. The standard treatment is complete surgical removal of cyst, and a dural defect should be carefully sought and repaired. Additionally, surgeons should be reminded of the possibility of multiple dural defects which requires meticulous exploration.

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