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Case Report:

Congenital spinal intradural arachnoid cyst associated with intrathoracic meningocele in a child

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Abstract: Congenital spinal intradural arachnoid cyst associated with intrathoracic meningocele is very rare. We report a case in a 9-year-old Chinese boy who presented with a two-week history of progressive paraparesis and gait ataxia. Magnetic resonance imaging revealed that a dorsal intradural extramedullary cystic lesion extended from T1 to T5 and compressed the spinal cord. A left lateral intrathoracic meningocele pouch was found incidentally at the level of T1. The arachnoid cyst as well as meningocele was removed and the spinal cord compression was relieved. Arachnoid cyst was confirmed by histological examination. The patient recovered well postoperatively. This is the second report of such a case in the world according to the available literature. The take-home message for our case is that the surgical approach should be individualized, depending on the size and location.

Key words: Spinal intradural arachnoid cyst, Intrathoracic meningocele, Spinal cord compression, Surgical treatment
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1 Introduction

Spinal intradural arachnoid cyst (AC) is a relatively uncommon lesion that causes symptomatic spinal cord compression, mostly dorsal to the thoracic spinal cord. The cysts may occur at any age, and often cause symptoms in the age group of 30–50 year-olds (Bassiouni *et al*., 2004). The pediatric spinal intradural ACs are, however, even more uncommon (Lee and Cho, 2001), and the exact pathogenesis remains unknown. Spinal ACs have been described in children associated with other neural tube anomalies, including myelomeningocele and diastematomyelia (Rabb *et al*., 1992) and kyphoscoliosis (Alvisi *et al*., 1987). But spinal AC associated with intrathoracic meningocele is very rare (Baysefer *et al*., 2001).

We report on the case of a Chinese boy with spinal intradural AC (T1–T5) associated with a left lateral intrathoracic meningocele and further discuss the diagnosis and treatment.

2 Case report

A 9-year-old Chinese boy presented with a two-week history of progressive paraparesis and gait ataxia. There was no history of trauma, previous spinal surgery, infection or spinal anesthesia. Paraparesis was confirmed on the neurological examination. Muscle strength of both legs was grade 4/5 proximally and grade 3/5 distally. Deep tendon reflex was bilaterally increased with positive Babinski's signs. No focal neurological sign or clinical evidence of neurofibromatosis was found. The emergent spinal cord magnetic resonance imaging (MRI) was performed and indicated a dorsal intradural extramedullary cystic lesion ranged from T1 to T5 segments, compressing the spinal cord. A left lateral intrathoracic meningocele pouch was also found incidentally at the T1 level. The meningocele extended into the left hemithorax through an enlarged intervertebral foramen and the spinal cyst protruded into the meningocele pouch (Fig. 1). The signal intensity of the cyst and meningocele was similar to that of the cerebral spinal fluid (CSF). Spinal intradural cyst (T1–T5)

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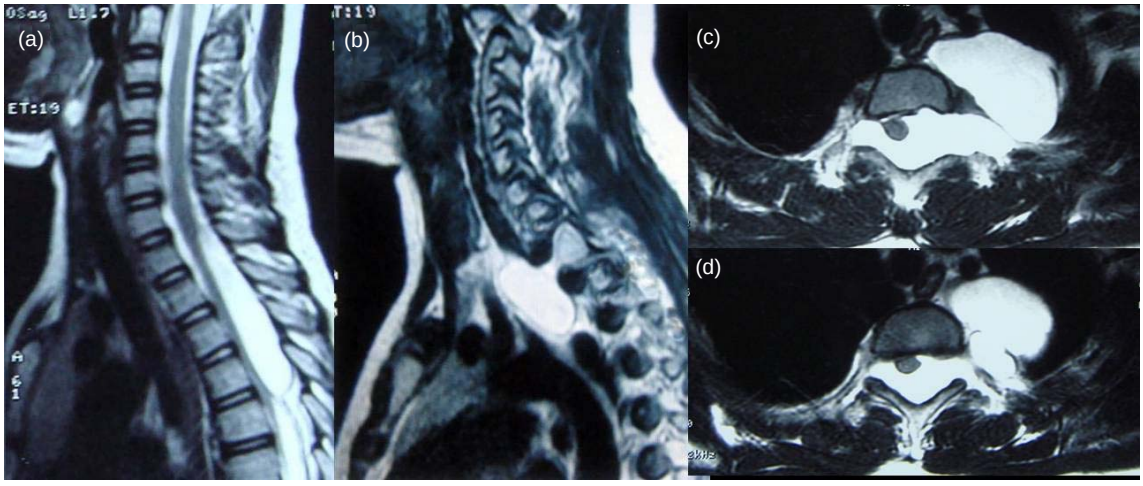

Fig. 1 Preoperative T2-weighted magnetic resonance images: (a) Sagittal image shows that the spinal arachnoid cyst extends from T1 to T5 vertebrae and compresses the spinal cord; (b) Sagittal image shows the intrathoracic meningocele at the level of T1; (c) & (d) Axial images show that the meningocele extends into the left hemithorax through an enlarged intervertebral foramen and the spinal arachnoid cyst protrudes into the meningocele pouch

and intrathoracic meningocele with spinal cord compression were the primary diagnoses.

A T1–T4 laminectomy was performed. Under the microscope, the dura was opened in the midline. Upon opening of the arachnoid layer, CSF escaped under high pressure and the cyst, filled with clear CSF, became visible. The cyst covered and compressed the dorsal aspect of the spinal cord, and spanned from the T1 to T5 segment. The spinal cord became thin with decreased pulsatile movement. The cyst protruded into the meningocele pouch at 3-cm depth and no nervous structures passed through the communication stalk. The cyst membrane was exposed and resected carefully, preventing further injury. Because a part of the anterior membrane was adherent firmly to the dorsal portion of the spinal cord, we had to leave these portions in situ. After excision of the cyst, the spinal cord gradually regained its normal diameter with good pulsation. Then, the meningocele pouch was pulled with biopsy forceps and resected, and was found to extend 6 cm into the left thoracic cavity through an enlarged left T1 intervertebral foramen. The stalk was carefully ligated and the enlarged intervertebral foramen was repaired with a piece of muscle and reinforced with fibrin glue.

The postoperative course was uneventful. The patient recovered well and could walk on postoperative Day 7. Pathologic examination of the resected cyst wall confirmed the diagnosis of an arachnoid cyst. One year after the surgery, the boy could walk and run without any difficulty and follow-up MRI scans showed no recurrence of the arachnoid cyst or intrathoracic meningocele. Meanwhile, minimal kyphoscoliosis of the spinal column was found (Fig. 2).

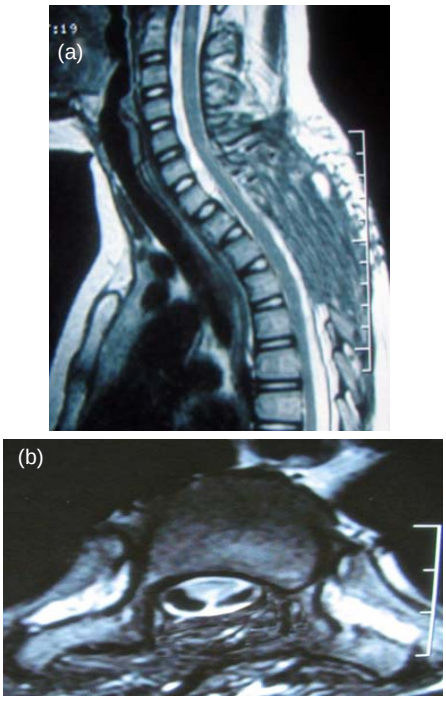

Fig. 2 Postoperative T2-weighted magnetic resonance images show that the intrathoracic meningocele and intraspinal arachnoid cyst have disappeared and there is minimal kyphoscoliosis of the spinal column

3 Discussion

Intrathoracic meningocele is defined as a CSF-filled sac with spinal meningeal wall, protruding into the thoracic cavity through an enlarged intervertebral foramen or bone defects. Acquired meningocele is relatively common as a laminectomy complication, while congenital meningocele is commonly associated with neurofibromatosis (de Andrade *et al*., 2003; Ebara *et al*., 2003). Isolated intrathoracic meningoceles without neurofibromatosis are more uncommon (de Andrade *et al*., 2003). The clinical manifestation of a thoracic meningocele is closely related to its size and relationship with surrounding structures. It may include back pain, shortness of breath, coughing, and palpitation when accompanied with the lung and mediastinal structure compression. The small lesion may be asymptomatic and was found incidentally, as in our case. In this case, the radiological and operative findings clearly confirmed the diagnosis of a left lateral intrathoracic meningocele. There was no clinical evidence of neurofibromatosis and no history of trauma or operation, suggesting it was a congenital lesion.

Spinal intradural AC is uncommon and rarely leads to symptomatic spinal cord compression. It affects the thoracic segment mostly, but also cervical and lumbar segments, and often locates dorsally to the spinal cord (Osenbach *et al*., 1992). The cysts may present at any age, but frequently cause symptoms between 30 and 50 years of age (Bassiouni *et al*., 2004). Pediatric spinal intradural ACs are relatively uncommon condition and most of them are idiopathic and congenital in origin. Although the exact pathogenesis remains unknown, the cysts are regarded as resulting from an alteration of the arachnoid trabeculae and enlargement because of a valve-like mechanism (Lee and Cho, 2001). Some cysts may result from trauma, arachnoiditis (Wang *et al*., 2003), and iatrogenesis (Jea *et al*., 2005). Because our patient had no history of trauma or obvious factors such as postsurgical arachnoiditis, meningeal infection, subarachnoidal bleeding, or other insults that cause inflammation and subarachnoid adhesions, his AC was considered to be of congenital or idiopathic origin.

To our knowledge, only Baysefer *et al*. (2001) reported a similar disease. They presented a case of a 2-year-old boy with an intradural AC of the spine from T3 to L1 segments and two large left lateral intrathoracic meningoceles at the level of T8 and T9 segments with multiple meningeal diverticula. Surgery was performed with removal of the meningocele pouches and shunting of the cyst to the subarachnoid region. In our case, the diagnosis of spinal intradural AC associated with intrathoracic meningocele was confirmed by MRI, operative findings, and pathologic examination.

Expectant management is a treatment option for asymptomatic patients with spinal AC or meningocele. Further surgical evaluation is needed, however, when patients show neurological signs. For symptomatic intraspinal ACs, complete neurosurgical resection is the treatment of choice (Osenbach *et al*., 1992), but is limited in cases with extensive cysts, ventral cyst location or many adhesions to the spinal cord, as present in our patient. Partial resection and fenestration of the cyst is the second choice, allowing maximal communication with the subarachnoid space (Alvisi *et al*., 1987; Osenbach *et al*., 1992). Shunting of the cyst to the peritoneum, pleural cavity, or subarachnoid space is the third operation type (Rabb *et al*., 1992; Bassiouni *et al*., 2004; Wang *et al*., 2003). Simple aspiration of the cysts will not work, as this maneuver usually results in recurrence.

The lesion size is an important factor in the surgical approach to the symptomatic intrathoracic meningocele. For small- and medium-sized lesions, the most common surgical management is resection of the meningocele and repair of the dural defect through laminectomy. While for large lesions, operative resection or ligation of the lesion through thoracotomy is non-available (Chee, 1989; Zamponi *et al*., 1996). In our report, we performed a partial wide removal of the cyst, resected the meningocele completely, and repaired the enlarged intervertebral foramen through T1–T4 laminectomy. The postoperative follow-up demonstrated that the operation was effective. At the same time, minimal kyphoscoliosis of the spinal column was found. A lengthy follow-up is still warranted, since our patient may need an orthopedic operative correction if his kyphoscoliosis becomes worse as he ages.

In conclusion, congenital spinal intradural AC associated with intrathoracic meningocele is quite rare. The lesion's size and location should be taken into consideration when choosing an operative approach.

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