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


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 cerebrospinal fluid (CSF). Spinal intradural cyst (T1–T5)
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. 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

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