Case Report

Multiple extradural arachnoid cysts at the spinal cord and cauda equina levels in the young

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Study design: A case report of multiple extradural arachnoid cysts at the spinal cord and cauda equina levels in the young.

Objective: To report an exceedingly rare case of multiple extradural arachnoid cysts at the spinal cord and cauda equina levels in the young.

Setting: Department of Orthopaedic Surgery, Tokai, Japan.

Case report: An 11-year-old boy was diagnosed with multiple extradural arachnoid cysts at the spinal cord and cauda equina levels extending from the T5 to L5 vertebrae and surgery was performed. At 2 years after surgery, no recurrence was observed and muscle weakness of the lower extremities and sensory disturbance improved.

Conclusion: Excision of only the arachnoid cysts at the spinal cord level led to a favorable outcome.

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Keywords: arachnoid cyst; extradural tumor; spinal cord tumor; cauda equina tumor

Introduction

Spinal arachnoid cyst is a relatively rare disease that accounts for approximately 1% of primary spinal cord tumors; however, the widespread use of magnetic resonance imaging (MRI) has been associated with an increasing number of reported cases in recent years. Here we report a case of multiple extradural arachnoid cysts at the spinal cord and cauda equina levels in the young.

Case report

The patient was an 11-year-old boy who had a chief complaint of gait disturbance without a significant family history. He was born prematurely and weighed 1155 g at birth without gross external congenital anomaly; there have been no known growth abnormalities. At 1 month before being referred to our institution, he had a contusion of the buttocks when falling from stairs at school and a diagnosis of coccygeal fracture was made, which was treated conservatively at another clinic. About a week later, gait disturbance emerged and he was referred to our institution for suspected spinal epidural hematoma.

Examination showed hyperesthesia at the level of T8–T10 and hypesthesia at the level of T10 and below, together with weakness of the lower extremities MRC grade 3–4 on manual muscle testing. Increased deep tendon reflexes of the lower extremities and ankle clonus were observed; Babinski reflex was present.

Plain radiographs on admission revealed a deformity suggesting apophyseal separation in the L1 vertebra and spina bifida in the L5 vertebra. An anteroposterior radiograph of the thoracic and lumbar vertebrae showed increased interpedicular distance, while a lateral radiograph revealed increased anteroposterior diameter of the spinal canal. MRI disclosed multiple cystic mass densities, which were isointense to cerebrospinal fluid, extending from the upper margin of T5 to the upper margin of L5 on the dorsal aspect of the spinal cord and cauda equina (Figure 1). Myelography and subsequent computed tomography (CT) showed leakage of contrast medium into the cysts disclosed by MRI (Figure 2).

These findings led to a diagnosis of multiple extradural arachnoid cysts at the spinal cord and cauda equina levels extending from the T5 to L5 vertebrae, and surgery was performed. Laminectomy was performed from the T5 to T9 vertebrae and from the T11 to L1 vertebrae. Arachnoid cysts were identified: (1) from the upper margin of T5 to the upper margin of T8, (2) from the upper margin of T8 to the upper margin of T10,

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Figure 1 Sagittal T1-weighted MRI disclosed multiple cystic mass densities, which were isointense to cerebrospinal fluid, extending from the upper margin of T5 to the upper margin of L5 on the dorsal aspect of the spinal cord and cauda equina

Figure 2 Sagittal CT myelography showed leakage of contrast medium into the cysts. In delayed CT (right), cystic mass of L2–L5 is imaged more

(3) from the lower margin of T10 to the lower margin of T12, (4) from the lower margin of T12 to the upper margin of L2, and (5) the upper margin of L2 and below. The communications between the above-mentioned cysts and the dura mater were found as follows: (1) in the left T7 nerve root, (2) in the left T8 nerve root, (3) from T11 to T12 vertebrae on the left, and (4) from T12 to L1 vertebrae on the right (Figure 3). The communications (1) and (2) were ligated and the corresponding arachnoid cysts were excised. The arachnoid cysts (3) and (4) were excised, the dura mater sutured, and the cysts removed. The arachnoid cyst located at the level of L2 and below was left untreated. Histopathological examination of the cyst wall showed layered collagenous fibers and epithelial tissue with flat lining cells, confirming arachnoid cysts (Figure 4).

As of this writing, 2 years after surgery, no recurrence was observed and muscle weakness of the lower extremities and sensory disturbance improved. However, scoliosis with a Cobb angle of 16° was found between the T7 and L1 vertebrae. Between the T2 and L1 vertebrae, backward curvature with a preoperative Cobb angle of 28° changed to 62° postoperatively, without significant effects on his growth.

Figure 3 Postoperative sketch illustrating the location

Figure 4 Histopathological examination of the cyst wall showed layered collagenous fibers and epithelial tissue with flat lining cells

Discussion

Spinal arachnoid cyst was first reported by Spiller in 1903.¹ Although spinal arachnoid cyst is a benign tumor accounting for approximately 1% of overall spinal cord tumors,² incidence of the disease has been frequently reported in recent years because of the development of MRI. Most of the reported cases are of solitary cysts occurring commonly at the thoracic level in individuals in a wide range of ages from 10 to 69 years. Extradural arachnoid cysts may be caused by congenital or acquired factors, including inflammation, trauma, and iatrogenic ones, although most cases are believed to be congenital in origin. In all cases, the disease results from sliding of the arachnoid from defective or fragile dura mater.² The arachnoid may be enlarged by the ball-valve mechanism of the nerve root or change in spinal pressure associated with pulse, breathing, and coughing.^{3,4} Clinical course should be closely monitored in asymptomatic individuals, while surgical treatment is indicated in patients with significant neurologic findings; complete closure of the communication is essential for complete cure. Dorsal spinal arachnoid cysts may be associated with fragility of spinal posterior supporting tissues, requiring consideration of spinal reconstruction.

There have been only five reported cases of multiple arachnoid cysts, including this case, regardless of intradural or extradural ones (Table 1).^{2,5-7} Multiple arachnoid cysts occurred in a narrow age group from 9 to 12 years of age, and their affected sites varied widely from the thoracic to sacral vertebrae. In spinal development, union of the vertebral arch occurs by approximately 7 years of age, and the cricoid cartilage develops

Table 1 Summary of cases of multiple arachnoid cysts regardless of intradural or extradural ones

Case No.	Authors	Age (years)	Gender	Level of cysts	Number of cysts	Treatment	Recurrence	Outcome
1	Andrew W Duncan (1978)	9	M	T6–T10	6 (Intradural)	With no publication		
2	Shuichirou Otake (1995)	11	F	T1–L2	3 (Intradural)/ 3 (Extradural)	Laminoplasty As much as possible excision (intradural) Tumorectomy (extradural)	(–)	Kyphosis Scoliosis
3	Lynn M Myles (1999)	9	F	T2–S	6 (Extradural)	Laminoplasty Tumorectomy Arachnoid cyst at the cauda equinal level was left untreated	(–)	Kyphosis
4	Kazuhiro Obara (2000)	12	M	T2–T12	11 (Extradural)	Laminoplasty Tumorectomy + closed communication Tumorectomy + unclosed communication Cyst-abdominal shunt	(–)	Kyphosis
5	Current case (2004)	11	M	T5–L5	5 (Extradural)	Laminectomy Tumorectomy + closed communication Arachnoid cyst at the cauda equinal level was left untreated	(–)	Kyphosis Scoliosis

all along the vertebrae at approximately 10 years of age, followed by longitudinal growth of the vertebrae.⁸⁻¹⁰ From this fact, we speculate that, multiple arachnoid cysts predominantly occur in this age group because the longitudinal growth of the vertebrae is involved in the onset of the disease, together with extensively and ventrally compressed spinal cord.

Regarding treatment, three of the four patients with multiple arachnoid cysts previously described in Cases 2-5 were surgically treated with laminoplasty. In Case 3, as with our case, only excision of arachnoid cysts at the spinal cord level resulted in a favorable outcome. In all cases, kyphosis occurred postoperatively. In Case 4, recurrence was observed in the arachnoid cyst with unclosed communication and the cyst with a shunt procedure, requiring additional surgery. In the present case, which involved extensive lesions in the young, the arachnoid cysts at the spinal cord level were excised with their communications closed, and the arachnoid cyst at the cauda equina level was left untreated. Marked thinning of the vertebral arch and externally extending arachnoid cysts made laminoplasty difficult; therefore, laminectomy was performed for excision of the arachnoid cysts. Although the patient had an uneventful postoperative growth with improvement in neurological symptoms, residual scoliosis and kyphosis will require close follow-up.

Conclusions

We report a case of multiple extradural arachnoid cysts at the spinal cord and cauda equina levels in the young,

which was treated with excision of only the arachnoid cysts at the spinal cord level, leading to a favorable outcome.

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