Extradural Giant Multiloculated Arachnoid Cyst Causing Spinal Cord Compression in a Child

Serdar Kahraman, MD; Ihsan Anik; Selcuk Gocmen, MD; Sait Sirin, MD

Department of Neurosurgery, GATA, Ankara, Turkey

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Abstract

Background: Spinal extradural arachnoid cysts are rare expanding lesions in the spinal canal. Enlargement may cause progressive signs and symptoms caused by spinal cord compression. They are associated with trauma, surgery, arachnoiditis, and neural tube defects. Most nontraumatic spinal extradural arachnoid cysts are thought to be congenital.

Design: Case report and literature review.

Findings: A 9-year-old boy with mild paraparesis was found to have an extradural multiloculated arachnoid cyst with fibrous septa at T4-L3 levels and anterior compression and displacement of the spinal cord.

Conclusions: Definitive treatment of arachnoid cyst entails radical cyst removal and dura cleft repair. Formation of a postoperative cerebrospinal fluid fistula may require external lumbar drainage.

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Key Words: Spinal cyst, extradural; Arachnoid cyst; Cystoperitoneal shunt; Dura, defect, graft; Paraparesis; Hypesthesia; Spinal cord compression

INTRODUCTION

Causes of spinal extradural arachnoid cysts remain unclear. They appear to be extradural outpouchings of arachnoid that communicate with the intraspinal sub-arachnoid space through a small defect in the dura. These cysts are most commonly located in the thoracic spine, followed by the lumbosacral junction. The most common presenting symptoms are pain, paresthesias, intermittent claudication, and variable degrees of spastic weakness (1).

Case Report

A 9-year-old boy underwent surgery with the preoperative diagnosis of thoracic arachnoid cyst and cystoperitoneal shunt. Postoperative pathology revealed an arachnoid cyst. One year after surgery, he was admitted to our hospital with back pain and mild paraparesis. Neurologic examination revealed motor strength of 3/5 for both lower extremities. Hypesthesia was apparent below the level of T4 bilaterally. Bilateral plantar extension responses were present, and both Achilles tendons were hyperreflexic. Thoracolumbar magnetic

resonance imaging (MRI) demonstrated a T4-L3 extradural multiloculated arachnoid cyst (Figure 1A through C).

After insufficient cystoperitoneal shunt, we planned resection of the cyst. At surgery, the cyst was dissected carefully from the underlying dura after cyst contents were removed. Two communicating defects between the dura and cyst were found during surgery. Defects were on the caudal side (inferior border) of each compartment without any connection to each other.

Duraplasty was performed by closing the defects with grafts from the patient's lumbar fascia, and artificial dura was laid out to each defect. Two days after the operation, a cerebrospinal fluid (CSF) fistula occurred. A second operation was carried out. Duraplasty was revised using fibrin glue, and external (intradural) lumbar drainage was performed. On postoperative day 5, the drainage tube was removed. There was no subsequent CSF leakage. Symptoms improved in 2 weeks.

DISCUSSION Pathogenesis

Arachnoid cysts are assumed to be the result of pathologic proliferation and distribution of the arachnoid trabeculae during the embryonic period. The arachnoid trabeculae, consisting of collagen and elastic fibers, are embryological remnants of the common origin of the arachnoid and pia mater, which pass from through the

Please address correspondence to Ihsan Anik, Department of Neurosurgery, GATA, Etlik, Ankara, Turkey; phone: 0090.533.247.6735; fax: 0090.312.304.7008 (e-mail: drianik@gmail).

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Figure 1. (A) Sagittal T2-weighted image of the thoracic spine reveals loculated arachnoid cysts separated by fibrous septa; the small part, nearly 1 cm in diameter, is located at T4-T5 intervertebral disk level, and the big part is located at T5-T12 levels that caused mild scalloping. The spinal cord is compressed and displaced to the anterior of the spinal canal. (B) On sagittal T2-weighted image of the lumbar spine, another loculation of the arachnoid cyst is seen, located at L1-L3 levels and separated from the superior one with fibrous septa. Note that all loculations are homogeneous and demonstrate high signal intensity. (C) Axial T2-weighted image from distal thoracic level shows extradurally located arachnoid cyst compressing the spinal cord.

subarachnoid space to combine with pia mater. The function of arachnoid trabeculae is to join and stabilize the two layers. The trabecular cells degenerate for such reasons as inflammation, hemorrhage, exposure to contrast media, penetrating trauma, and surgery. These lesions result in scar formation and adhesions that separate the subarachnoid space into a diverticulum or cyst; however, this explains only the occurrence of intradural arachnoid cysts. Pathogenesis of extradural arachnoid cysts is completely different from that of intradural arachnoid cysts. The related dural defect seems to be essential to the genesis of an extradural arachnoid cyst. In most of the cases, nontraumatic spinal extradural arachnoid cysts are considered to arise from a congenital dural defect, which allows the arachnoid membrane to herniate through the adjacent dura mater (2–4).

Enlargement of cyst dimensions is proposed to be responsible for spinal cord compression. Authors have described the continuous extension of arachnoid cysts by 3 mechanisms: (a) active CSF secretion from residual arachnoid matter, that is, noncommunicating arachnoid cyst, (b) valve effect, also termed communicating arachnoid cyst with passive fluid transport mechanism, and (c) free water entrance caused by hyperosmolar cyst content (2,3,5–13).

Differential Diagnosis

Epidermoid cysts are usually isointense or mildly hyperintense to CSF; the borders may be somewhat irregular, and they widen through cisterns and surround neurovascular structures. Unilocular type 1 hydatid cysts usually have thinner walls in comparison with their counterparts,

except for arachnoid cysts. Neuroepithelial cysts are isointense to CSF and usually multiseptated. Synovial cysts are usually small and always originate from facet articulation that can be shown on MRI. Tarlov cysts (perineural cysts) are usually located at the sacrum, are eccentrically centered over neural foramen, and always contain neurofibers. Meningoceles typically have a large extraspinal component that extends through the neural foramina. Lateral thoracic meningoceles are usually associated with neurofibromatosis type 1 and widespread dural ectasia.

Cystic tumors have a solid part that shows contrast enhancement (14,15). In this case, thoracolumbar MRI revealed an extradural multiloculated arachnoid cyst with fibrous septa at T4-L3 levels. Two loculations were in the thoracic area, and 1 loculation was in the lumbar area. The largest loculation, which was at T5-T12 levels, caused mild scalloping. The spinal cord was considerably compressed and displaced to anterior of the spinal canal. All loculations were homogeneous and demonstrated low signal intensity on T1-weighted images and high signal intensity on T2-weighted images.

Symptoms and Signs

The most common presenting symptoms and signs are pain and progressive spastic or flaccid paraparesis, which are often asymmetric and generally related to compression of the spinal cord or nerve roots (7). Symptoms typically fluctuate, with remissions and exacerbations. Valsalva maneuvers, which occur during daily activities, might increase the volume of the cyst (2). In this case, the most striking symptom was progressive spastic symmetric paraparesis and back pain, the severity of which changed with position.

Surgical Procedures

Surgical approaches of spinal arachnoid cysts include fenestration and/or resection of the cyst wall, shunt placement-cyst-to-pleural space and cyst-to-subarachnoid space (10). The aim of surgical treatment should be neural decompression and prevention of refilling of the cyst, which is best treated by complete resection of the cyst and closure of the communication between cyst and subarachnoid space (16). In this case, cystoperitoneal shunt had been performed, but it was not effective. At surgery, a large extradural mass similar to an arachnoid cyst with a thick wall was identified. The cyst was dissected from the caudal part of the dura, keeping the wall of the cyst intact. The cyst was removed en bloc after its content was drained.

There were 2 defects between dura and the cysts. Both defects were on the caudal side (inferior border) of the compartments, and there was no relation between the defects. Both defects were surgically repaired. After resection of the cyst, closure of the dura defects, and repair of the CSF fistula and external intradural lumbar

drainage for 5 days, the patient's neurologic condition improved in 2 weeks.

CONCLUSIONS

Surgical treatment of spinal arachnoid cysts should not only provide neural decompression but also prevent cyst refilling. Simple aspiration or shunting is inadequate and not recommended. Choice of treatment should be radical cyst removal and dura cleft repair. Formation of a postoperative CSF fistula may require external lumbar drainage. Neurologic improvement may follow appropriate surgical intervention.

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