J Neurosurg (Pediatrics 3) 103:266–269, 2005

Noncommunicating spinal extradural arachnoid cyst causing spinal cord compression in a child

Case report

JAMES K. LIU, M.D., CHAD D. COLE, M.D., M.SC., GREGORY T. SHERR, M.D., JOHN R. W. KESTLE, M.D., AND MARION L. WALKER, M.D.

Department of Neurosurgery, Primary Children's Medical Center, University of Utah School of Medicine, Salt Lake City, Utah

✓Extradural arachnoid cysts in the spine are relatively uncommon causes of spinal cord compression in the pediatric population that are thought to arise from congenital defects in the dura mater. Most reports describe such cysts communicating with the intrathecal subarachnoid space through a small defect in the dura. The authors describe the case of a child who presented with spinal cord compression caused by a large spinal extradural arachnoid cyst that did not communicate with the intradural subarachnoid space. An 11-year-old girl presented with urinary urgency, progressive lower-extremity weakness, myelopathy, and severe gait ataxia. Magnetic resonance imaging of the spine demonstrated a large extradural arachnoid cyst extending from T-8 to T-12. The patient underwent a thoracic laminoplasty for en bloc resection of the spinal extradural arachnoid cyst. Intraoperatively, the dura was intact and there was no evidence of communication into the intradural subarachnoid space. Postoperatively, the patient's motor strength and ambulation improved immediately, and no subsequent cerebrospinal fluid leak occurred.

Noncommunicating spinal extradural arachnoid cysts are extremely rare lesions that can cause spinal cord compression in children. Because the dura remains intact, they can be removed entirely without subsequent dural repair. The authors review the literature and discuss the proposed underlying mechanisms of formation of these arachnoid cysts.

KEY WORDS • **arachnoid cyst** • **spinal cord compression** • **pediatric neurosurgery**

EXTRADURAL arachnoid cysts are relatively rare lesions of the spinal canal and are uncommon causes of spinal cord compression in children and adolescents.[1,4,10,13] These cysts are often described as diverticula or protrusions of arachnoid membrane that herniate through a small dural defect, producing a cyst containing CSF with a pedicle that is in communication with the spinal subarachnoid space.[1,4,6–8,13,14,16,18,20,21] The majority of these dural defects are thought to be of congenital origin; however, some can be acquired from trauma, infection, or inflammation.[4] These extradural arachnoid cysts are most commonly found in the middle to lower thoracic spine; less commonly they appear in the upper thoracic, lumbar, sacral, and cervical regions.[3,10–13] On rare occasions, enlargement of extradural arachnoid cysts can cause symptomatic spinal cord compression that results in progressive spastic paraparesis.[11,13,18] Excision of the cyst with obliteration of the communicating dural defect is the mainstay of treatment in symptomatic patients.

Pathologically, the cyst wall consists of fibrous connective tissue with an inner single-cell arachnoid lining; however, the inner arachnoid lining is sometimes absent on histological examination.[10,18] The term extradural arachnoid cyst has thus been used interchangeably with extradural meningeal cyst. Nabors, et al.,[14] described a classification system of spinal arachnoid cysts divided into three major categories: extradural cysts without spinal nerve root fibers (Type I); extradural cysts with spinal nerve root fibers (Type II); and intradural cysts (Type III). Type I is further divided into extradural arachnoid cysts (Type IA) and sacral meningoceles (Type IB). In some cases, an extradural cyst has significant intradural extension.[6,7]

In nearly all cases of Type IA cysts, communication of CSF between the cyst and the intrathecal subarachnoid space through a dural defect has been reported.[4,6,12,14,15] We report the case of a large Type IA extradural arachnoid cyst of the thoracic spine that produced symptomatic spinal cord compression in an 11-year-old girl. The case is unique because there was no dural defect or fistulous communication into the intradural subarachnoid space. Complete excision was performed without dural repair.

Abbreviation used in this paper: CSF = cerebrospinal fluid.

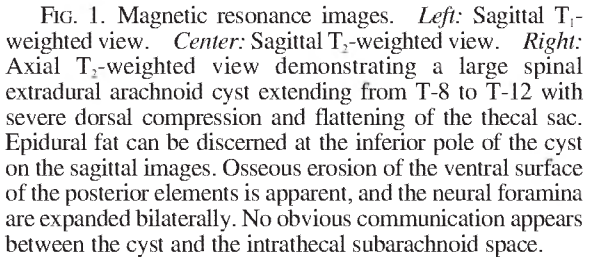

FIG. 1. Magnetic resonance images. *Left:* Sagittal T_1-weighted view. *Center:* Sagittal T_2-weighted view. *Right:* Axial T_2-weighted view demonstrating a large spinal extradural arachnoid cyst extending from T-8 to T-12 with severe dorsal compression and flattening of the thecal sac. Epidural fat can be discerned at the inferior pole of the cyst on the sagittal images. Osseous erosion of the ventral surface of the posterior elements is apparent, and the neural foramina are expanded bilaterally. No obvious communication appears between the cyst and the intrathecal subarachnoid space.

Case Report

History and Examination. This 11-year-old girl presented with several years of urinary urgency that progressed to lower-extremity weakness, myelopathy, and severe gait ataxia over a period of 2 months. She had progressive difficulty in walking and frequently bumped into walls. On neurological examination, she had 5/5 strength in all muscle groups except in both hip flexor muscles, which were 4/5. Proprioception and vibratory sensation were diminished in both legs. Her reflexes were hyperactive and her gait was severely ataxic. She had no cutaneous stigmata of neurological disease.

Neuroimaging. Magnetic resonance imaging with Gd enhancement of the spine demonstrated a large, nonenhancing extradural mass extending from T-8 to T-12 with severe dorsal compression and flattening of the thecal sac (Fig. 1). The mass was hypointense on T_1-weighted imaging, was hyperintense on T_2-weighted imaging, and suppressed signal intensity on fluid-attenuated inversion recovery imaging. Diffusion-weighted imaging demonstrated no restriction in the epidural spinal mass. The signaling patterns were consistent with CSF suggestive of a spinal extradural arachnoid cyst. The anterior surface of the posterior elements was eroded and the neural foramina at T9–10, T10–11, and T11–12 were expanded bilaterally. No obvious communications between the cyst and the intrathecal subarachnoid space were detected on neuroimaging.

Operation. The patient underwent a thoracic laminoplasty to expose the extradural spinal mass (Fig. 2). At surgery, a large extradural mass resembling an arachnoid cyst was identified, with a thick but translucent wall. The cyst was easily dissected from the exterior surface of the thecal sac and removed en bloc, keeping the wall of the cyst intact (Fig. 2). The dura mater was inspected and there was no evidence of a dural defect, arachnoid pedicle, or fistulous communication into the intradural subarachnoid space. The thecal sac was decompressed and reexpanded. The thoracic laminae were replaced with titanium miniplates and screws.

Pathological Examination. Grossly, the specimen measured 6 × 3 × 0.7 cm (Fig. 2). It was lined by a thin grayish white slightly cloudy membrane and was filled with clear colorless fluid. Histological examination of the cyst wall demonstrated fibrous connective tissue and some areas were lined by a single layer of flattened cells, consistent with an arachnoid cyst. Meningothelial cells, some of which were multinucleated, were found within the fibrous connective tissue.

Postoperative Course. The patient's motor function improved to full strength and she was ambulating well. A CSF leak did not develop, and she was discharged on the 3rd postoperative day.

Discussion

Clinical Features

Type IA spinal extradural arachnoid cysts are uncommon causes of progressive spastic paraparesis in adolescence. According to Cloward,[4] spinal arachnoid cysts are most common in the second decade of life and are twice as frequent in male as in female patients. Approximately 65% of these lesions affect the thoracic spine, particularly the middle to lower thoracic regions, followed by the thoracolumbar (12%), lumbar and lumbosacral (13%), sacral (7%), and cervical (3%) spine.[4] Type IA lesions most commonly arise dorsally and can partially protrude into the adjacent neural foramen. These lesions can extend over several spinal segments as a single cyst and can even occur as multiple cysts each associated with a separate dural defect and communicating pedicle (Fig. 3).[13] Thoracic cysts usually occur in young adolescents, whereas thoracolumbar and lumbar cysts usually occur in adults in the fourth decade of life.[4,5]

FIG. 2. *Upper:* Intraoperative photograph demonstrating a large extradural spinal arachnoid cyst arising from the dorsal aspect of the spinal canal. The cyst was easily dissected off the thecal sac and removed en bloc, keeping the cyst wall intact. No evidence was seen of a dural defect or communicating pedicle into the intradural subarachnoid space. *Lower:* Photograph of the gross specimen, which measured 6 × 3 × 0.7 cm.

When these lesions compress the intraspinal neural elements, symptoms usually manifest as painless progressive spastic paraparesis and difficulty in walking, with initial relative sparing of the sphincter tone. Motor weakness is usually more predominant than sensory loss.[10,17,21] Radiculopathy may be present at the level of the lesion. Bowel and bladder disturbances are present in approximately 20% of cases.[4] Slow expansion of these cysts over a long period can result in osseous erosion, remodeling of the spinal canal, and kyphoscoliosis.

The clinical presentation can also vary depending on the location of the spine and on the severity of spinal cord and root compression.[14] Patients with cervical cysts usually present with spastic quadriparesis and sensory levels, whereas those with lesions lower in the cervical spine may present with Horner syndrome.[4] Patients with thoracic cysts tend to present with progressive spastic paraparesis. Back pain is generally uncommon in thoracic lesions. Patients with lumbar and lumbosacral cysts, on the other hand, classically present with low-back pain, radiculopathy, and bowel and bladder dysfunction. Symptoms may be intermittent and are sometimes exacerbated by Valsalva maneuvers or by gravitational positional forces.[20] Remissions and fluctuation of symptoms have been reported in approximately 30% of cases.[4]

Mechanisms of Pathogenesis

The exact origin and pathogenesis of Type IA spinal extradural arachnoid cysts remain unknown. Several theories have been proposed by various authors. In 1934, Elsberg, et al.,[9] attributed the origin of these cysts to either congenital diverticula of the dura or herniation of arachnoid membrane through a congenital dural defect. Congenital dural defects appear most commonly at the dural sleeve of the nerve root or at the junction of the sleeve and the thecal sac; they derive less commonly from the dorsal midline of the thecal sac.[1–4] A congenital origin is further supported by reports of a familial syndrome comprising multiple spinal arachnoid cysts, lower-extremity lymphedema (Milroy disease), and distichiasis (double row of eyelashes).[1,2,13,19] Spinal extradural arachnoid cysts have also been associated with congenital pigmented nevus, diastematomyelia, multiple sclerosis, Marfan syndrome, neural tube defects, spinal dysraphism, and syringomyelia, all of which suggest a genetic component in origin.[3,15] The loss of tissue elasticity and decrease in tensile tissue strength found in Marfan syndrome may be associated with dural ectasia in the development of these cysts.[22]

Although less common, spinal arachnoid cysts have been associated with arachnoiditis, surgery, and trauma, leading others to suggest that these cysts arise from acquired dural defects.[17,23] Spiegelmann, et al.,[20] reported a case in which hemosiderin-containing macrophages found in the cyst wall in a patient with a spinal extradural arachnoid cyst caused spastic paraparesis 10 years after craniospinal injury.

The mechanism of cyst enlargement leading to spinal cord compression has been a subject of debate. Some theories include active fluid secretion from the cyst wall, passive osmosis of water, and hydrostatic pressure of CSF.[10] Because the cyst walls are composed of simple connective tissue with frequent absence of the inner arachnoid lining, the theory of active fluid secretion is unlikely. The theory that osmosis causes cystic enlargement has often been dismissed because most authors have indicated that the fluid content of the cyst is the same as that of CSF.[4] On the other hand, Gortvai[10] found xanthochromic fluid in the cyst, which may have given rise to a higher osmotic pressure, thus allowing osmosis of water as an adjunct to cyst enlargement.

Some have postulated that cyst expansion is triggered by a ball–valve mechanism in the communicating pedicle associated with pulsatile CSF dynamics.[10,12,17] According to the theory of McCrum and Williams,[12] intermittent surges of pressure in the subarachnoid space are communicated to the cyst and flow occurs into the pouch. When pressure falls again, egress of fluid is impeded by compression of the cyst pedicle. According to the Laplace law, the body of the cyst exerts a force on the neck sufficient to close the communication, because its radius and wall tension are greater. This mechanism then allows further enlargement, with persistent CSF pulsations. This ball–valve mechanism has been observed intraoperatively by Rohrer, et al.[17] Bone erosion of the spinal canal may imply the presence of a valve mechanism that is responsible for producing forces of CSF pressure within the cyst that are greater than normal hydrostatic forces.[14]

Our case is unusual in that there was no detectable dural defect or communication with the intradural subarachnoid space. We postulate that the noncommunicating cyst evolved from a preexisting communicating cyst that initially formed as a result of a small dural defect. Over time, the cyst enlarged and eventually obliterated the communicating channel to the subarachnoid space because of the

FIG. 3. *Left:* Drawing of a spinal cross-section illustrating the dural defect and communication of the cyst with the subarachnoid space. *Right:* Drawing of a Type IA spinal extradural arachnoid cyst with a communicating pedicle at the site of the dural defect. Reprinted from Bergland RM: Congenital intraspinal extradural cyst. Report of three cases in one family. **J Neurosurg 28:**495–499, 1968.

Laplace law.[12] The closure of the dural defect may be due to the proliferation of arachnoid cells. In the thoracic region, where spinal fluid pressure in the upright position approaches zero, early closure by proliferation of arachnoid cells may be facilitated, thereby producing a symptomatic cyst that is easier to identify than are the cysts located in other areas of the spinal cord. Magnetic resonance imaging is valuable in determining the presence of the cyst. Computerized tomography myelography is the diagnostic study of choice to demonstrate the communication of the cyst with the subarachnoid space;[8,11,13] however, this study was not performed in our case.

References

1. Bergland RM: Congenital intraspinal extradural cyst. Report of three cases in one family. **J Neurosurg 28:**495–499, 1968
2. Chynn KY: Congenital spinal extradural cyst in two siblings. **Am J Roentgenol Radium Ther Nucl Med 101:**204–215, 1967
3. Cilluffo JM, Gomez MR, Reese DF, Onofrio BM, Miller RH: Idiopathic ("congenital") spinal arachnoid diverticula. Clinical diagnosis and surgical results. **Mayo Clin Proc 56:**93–101, 1981
4. Cloward RB: Congenital spinal extradural cysts: case report with review of literature. **Ann Surg 168:**851–864, 1968
5. Cloward RB, Bucy PC: Spinal extradural cyst and kyphosis dorsalis juvenilis. **AJR Am J Roentgenol 38:**681–706, 1937
6. Congia S, Coraddu M, Tronci S, Nurchi G, Fiaschi A: Myelographic and MRI appearances of a thoracic spinal extradural arachnoid cyst of the spine with extra- and intraspinal extension. **Neuroradiology 34:**444–446, 1992
7. Dickson RA, Arabi K, Goodfellow J: Congenital spinal extradural cyst (lateral meningocele) simulating acute transverse myelitis. Report of a case. **J Bone Joint Surg Br 60:**412–415, 1978
8. DiSclafani A II, Canale DJ: Communicating spinal arachnoid cysts: diagnosis by delayed metrizamide computed tomography. **Surg Neurol 23:**428–430, 1985
9. Elsberg CA, Dyke CG, Brewer ED: The symptoms and diagnosis of extradural cysts. **Bull Neurol Inst NY 3:**395–417, 1934
10. Gortvai P: Extradural cysts of the spinal canal. **J Neurol Neurosurg Psychiatry 26:**223–230, 1963
11. Khosla A, Wippold FJ II: CT myelography and MR imaging of extramedullary cysts of the spinal canal in adult and pediatric patients. **AJR Am J Roentgenol 178:**201–207, 2002
12. McCrum C, Williams B: Spinal extradural arachnoid pouches. Report of two cases. **J Neurosurg 57:**849–852, 1982
13. Myles LM, Gupta N, Armstrong D, Rutka JT: Multiple extradural arachnoid cysts as a cause of spinal cord compression in a child. Case report. **J Neurosurg (Spine 1) 91:**116–120, 1999
14. Nabors MW, Pait TG, Byrd EB, Karim NO, Davis DO, Kobrine AI, et al: Updated assessment and current classification of spinal meningeal cysts. **J Neurosurg 68:**366–377, 1988
15. Rabb CH, McComb JG, Raffel C, Kennedy JG: Spinal arachnoid cysts in the pediatric age group: an association with neural tube defects. **J Neurosurg 77:**369–372, 1992
16. Rengachary SS, O'Boynick P, Karlin CA, Batnitzky S, Price H: Intrasacral extradural communicating arachnoid cyst: cases report. **Neurosurgery 8:**236–240, 1981
17. Rohrer DC, Burchiel KJ, Gruber DP: Intraspinal extradural meningeal cyst demonstrating ball-valve mechanism of formation. Case report. **J Neurosurg 78:**122–125, 1993
18. Sato K, Nagata K, Sugita Y: Spinal extradural meningeal cyst: correct radiological and histopathological diagnosis. **Neurosurg Focus 13(4):**E1, 2002
19. Schwartz JF, O'Brien MS, Hoffman JC Jr: Hereditary spinal arachnoid cysts, distichiasis, and lymphedema. **Ann Neurol 7:** 340–343, 1980
20. Spiegelmann R, Rappaport ZH, Sahar A: Spinal arachnoid cyst with unusual presentation. Case report. **J Neurosurg 60:** 613–616, 1984
21. Stechison MT, Hendrick EB, Cohen E: Spinal extradural arachnoid cyst. **Pediatr Neurosci 15:**36–38, 1989
22. Stern WE: Dural ectasia and the Marfan syndrome. **J Neurosurg 69:**221–227, 1988
23. Swanson HS, Fincher ER: Extradural arachnoidal cysts of traumatic origin. **J Neurosurg 4:**530–538, 1947

Manuscript received January 10, 2005.
Accepted in final form April 27, 2005.

Address reprint requests to: Marion L. Walker, M.D., Department of Neurosurgery, University of Utah, 30 North 1900 East, Suite 3B409, Salt Lake City, Utah 84132. email: marion.walker@hsc.utah.edu.