

A rare but serious cause of spinal cord compression in an adolescent female

A young woman presents to the ED for evaluation after falling down the stairs a few times and developing extremity paresthesias and midthoracic back pain.

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CASE

A 19-year-old female presented to the emergency department, accompanied by her mother, with complaints of back pain and paresthesias. The patient stated that she had fallen down the night before and developed paresthesias in both lower extremities. She was also having difficulty with ambulation. She noted that she had fallen down a short flight of stairs on two different occasions within the previous month. After one of the falls, she developed midthoracic level pain and sought evaluation and treatment with a chiropractor. Plain radiographs taken of the thoracic spine failed to show any abnormalities. The patient's pain had continued, and she began to notice weakness in the lower extremities. She also experienced increasing difficulty with ambulation. No bowel or bladder dysfunction was present. The patient's history was significant only for anxiety and sinus infection, and she denied any trauma or surgery to the spine.

Physical examination demonstrated a female of normal weight, development, and tone. Cardiopulmonary and GI examinations were both unremarkable. Neurologic examination revealed muscle strength of 5+/5 in the upper extremities; however, proximal muscle strength in the lower extremities, particularly the hip flexors and quadriceps, was 4/5 bilaterally. The patient was alert and oriented to time, person, and place. Speech and language were intact. Plantar flexion and dorsiflexion of the ankles were normal. Cranial nerves II to XII were grossly intact. Sensory testing in both the upper and lower extremities was normal to light touch and pinprick. Plantar responses were downgoing. Deep tendon reflexes were brisk, especially in the lower extremities. Pain was elicited while palpating the midthoracic spine.

MRI of the cervical, thoracic, and lumbar spine without gadolinium was performed. A mass lesion approximately 1.9×1.6 cm was found dorsal to the spinal cord at the T6-T7 level, causing about 50% stenosis of the canal (Figure 1). It appeared to be an extradural lesion. The patient was admitted to the hospital for further evaluation.

Thoracic MRI with contrast revealed the same nonenhancing lesion at T6-T7 (Figure 2). The imaging results were discussed with the patient and her family. Given the size and location of the lesion as well as her deteriorating clinical symptoms, surgical removal was recommended. Based on the MRI images, the differential diagnosis included schwannoma, meningioma, arachnoid cyst, and Tarlov's perineurial cyst.

A complete laminectomy at T6 and T7 revealed a cystic, cerebrospinal fluid (CSF)-filled lesion dorsal to the spinal cord. The lesion was gradually dissected off the dura of the spinal canal and was found to have a small pedicle attaching it to the dural sac of the nerve root exiting the spinal cord between T6 and T7. There appeared to be communicating flow of CSF through the pedicle opening. This connection was ligated with a surgical clip and disconnected. The lesion was then removed en bloc. The patient tolerated the surgical procedure well. The pathology report confirmed an epidural arachnoid cyst.

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FIGURE 1. MRI showed a lesion dorsal to the spinal cord.



FIGURE 2. Thoracic MRI with contrast confirmed the lesion.

The patient had an uncomplicated postoperative course. The lower extremity paresthesias resolved within the first day after surgery. She experienced mild incisional and thoracic back pain postoperatively and received inpatient physical therapy for 3 days prior to being discharged from the hospital. No detectable lower extremity weakness or pain was present at her 4-week follow-up visit.

DISCUSSION

Arachnoid cysts are lesions within the spinal canal that are a rare but serious cause of spinal cord compression. These lesions can be intradural or extradural. Extradural arachnoid cysts are meningeal outpouchings that arise either from congenital defects in the dura mater or as a result of previous trauma, infection, or inflammation. They are CSF-filled lesions that are most commonly found on the dorsal aspect of the spinal canal. Congenital extradural cysts often appear at the dural sleeve of the nerve root and can protrude through the neural foramen. Extradural lesions have been associated with congenital pigmented nevus, diastematomyelia, multiple sclerosis, Marfan syndrome,

neural tube defects, spinal dysrhapism, and syringomyelia.¹⁻³ Noncongenital extradural cysts have been reported in patients with previous arachnoiditis, surgery, or trauma.^{1,4}

Extradural arachnoid cysts are seen most frequently in the thoracic spine (65-70%), followed by the lumbosacral, thoracolumbar, and cervical spine.³ Twice as many extradural cysts occur in males than in females, and they manifest most often in the second decade of life.⁵ Patients typically present with painless progressive spastic or flaccid paraparesis and difficulty walking. Motor weakness is more pronounced than sensory loss, and sphincter tone is typically spared. Back pain is generally uncommon in thoracic lesions but more common in the lumbar cysts. Symptoms may be intermittent and can sometimes be exacerbated by Valsalva maneuvers. Slowly expanding cysts may result in osseous erosion and remodeling of the spinal canal, and kyphoscoliosis is seen on plain radiographs.¹⁻³

Spiller and colleagues reported the first arachnoid cyst in 1903. Later in 1934, Elsberg and associates were the first to characterize these lesions based on the painless progressive paraplegia they produced.⁶ They also noted the fluctuating severity of symptoms in patients with an arachnoid cyst and the high incidence of these cysts in adolescents. Elsberg also believed that the pathogenesis of arachnoid cysts was either a herniation of the arachnoid through a dural defect or a diverticula of the dura mater itself. Soon thereafter, Cloward and Bucy reported the association of extradural cysts with Scheuermann disease or kyphosis dorsalis juvenilis.^{1,7} It was believed that the rhythmic fluctuations in CSF pressure caused bony erosions surrounding these lesions. Some researchers proposed that extradural arachnoid cysts enlarge due to a ball valve mechanism in which CSF enters into the cyst but cannot exit.⁸

Diagnosis Prior to the availability of MRI to produce high quality images, myelography was the imaging study of choice to diagnose extradural arachnoid cysts. During myelography, the cysts could easily be identified as they filled with contrast medium injected into the subarachnoid space. Currently, most cysts are diagnosed using MRI of the spine either with or without contrast medium. Similar to the way CSF presents, the lesion appears with low intensity on a T1-weighted image and high intensity on a T2-weighted image.⁹ Although bony erosion—demonstrated by widening of the canal, pedicle erosion, foraminal enlargement, and

TEACHING POINTS

- Arachnoid cysts are lesions within the spinal canal that are a rare but serious cause of spinal cord compression. These lesions can be intradural or extradural.
- A patient who presents with a history of progressive spastic or flaccid paralysis, backache, paresthesias, and difficulty with ambulation may have a compressing lesion.
- Diagnosis is mainly by MRI, but plain radiographs may also be used.
- While extradural arachnoid cysts are often symptomatic and require surgical removal, intradural arachnoid cysts, which are more commonly seen, do not have a tendency to enlarge and most often do not require surgery.

scalloping of the vertebral bodies—may be found on plain radiographs, it should not be the sole diagnostic criterion.^{2,9}

Treatment Symptomatic extradural arachnoid cysts should be treated as any other spinal tumor. Laminectomy with complete resection of the lesion without causing neurologic deficit is standard. Identification of the communication between the cyst and dural sac as well as ligation and possible repair of any dural defects are the goals of treatment. Frequently, the lesion can be removed en bloc without complication. Patients typically recover fully with minimal discomfort. On occasion, postoperative physical therapy may be indicated in patients with significant preoperative gait disturbance. While extradural arachnoid cysts are often symptomatic and require surgical removal, intradural arachnoid cysts, which are more commonly seen, do not have a tendency to enlarge and most often do not require surgery.

Conclusion Extradural arachnoid cysts are a rare but serious cause of spinal cord compression. A patient who presents with a history of progressive spastic or flaccid paralysis, backache, paresthesias, and difficulty with ambulation may have a compressing lesion. MRI is the radiographic study of choice, but plain radiographs may also reveal bony changes in the area of the lesion. Once an MRI study has been performed, the clinician can identify the lesion as being intradural or

extradural. An arachnoid cyst will have the same density as CSF on the MRI. Surgical excision without neurologic sequelae is the optimal treatment. Most patients recover to become completely asymptomatic. **JAAPA**

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REFERENCES

1. Cloward RB. Congenital spinal extradural cysts: case report with review of literature. *Ann Surg.* 1968;168(5):851-864.
2. Cilluffo JM, Gomez MR, Reese DF, et al. Idiopathic ("congenital") spinal arachnoid diverticula. Clinical diagnosis and surgical results. *Mayo Clin Proc.* 1981;56(2):93-101.
3. Liu JK, Cole CD, Sherr GT, et al. Noncommunicating spinal extradural cyst causing spinal cord compression in a child. *J Neurosurg.* 2005;103(3 suppl):266-269.
4. Swanson HS, Fincher EF. Extradural arachnoidal cysts of traumatic origin. *J Neurosurg.* 1947;4(6):530-538.
5. Chynn KY. Congenital spinal extradural cyst in two siblings. *Ann J Roentgenol Radium Ther Nucl Med.* 1967;101(1):204-215.
6. Elsberg CA, Dyke CG, Brewer ED. The symptoms and diagnosis of extradural cysts. *Bull Neurol Inst NY.* 1934;3:395-417.
7. Cloward RB, Bucy PC. Spinal extradural cyst and kyphosis dorsalis juvenilis. *Am J Roentgenol Radium Ther Nucl Med.* 1937;38:681-706.
8. Rohrer DC, Burchiel KJ, Gruber DP. Intraspinal extradural meningeal cyst demonstrating ball-valve mechanism of formation. Case report. *J Neurosurg.* 1993;78(1):122-125.
9. Nabors MW, Pait TG, Byrd EB, et al. Updated assessment and current classification of spinal meningeal cysts. *J Neurosurg.* 1988;68(3):366-377.