REVIEW



GWYNETH HUGHES, MD Department of Neurological Surgery, Cleveland Clinic

KENE UGOKWE, MD Department of Neurological Surgery, Cleveland Clinic EDWARD C. BENZEL, MD Chairman, Cleveland Clinic Spine Institute, Department of Neurological Surgery, Cleveland Clinic

A review of spinal arachnoid cysts

ABSTRACT

The symptoms of spinal arachnoid cysts are variable and nonspecific, so they are commonly misdiagnosed. Often the cysts are discovered incidentally on magnetic resonance imaging (MRI). If they cause no symptoms, no treatment is warranted regardless of the size of the cyst. Cysts that cause symptoms from mechanical compression of the spinal cord are best evaluated with MRI and surgically excised if possible.

KEY POINTS

Spinal arachnoid cysts can occur at any age and at any spinal level.

Symptoms vary widely but typically include waxing and waning pain and spastic or flaccid paraparesis.

Most spinal arachnoid cysts are asymptomatic when diagnosed and are discovered incidentally on MRI or myelography.

MRI and computed tomography help characterize spinal arachnoid cysts and differentiate them from abscesses and tumors.

Symptomatic cysts should be surgically resected. If complete resection is impossible, fenestration of the cyst wall, drainage, or shunting may relieve symptoms.

An asymptomatic spinal arachnoid cyst should be followed annually with serial imaging.

ANY PATIENTS with spinal arachnoid cysts complain of symptoms suggesting spinal cord compression, and are often initially evaluated by their primary physicians. However, these cysts are often discovered incidentally.

This article discusses how to manage spinal arachnoid cysts, whether found incidentally or during an evaluation for symptoms of spinal cord compression.

PRESENTATIONS CAN VARY WIDELY

A patient with a clinically relevant spinal arachnoid cyst is most likely to be a boy in his teens, but these cysts can occur in either sex and have been reported in patients as young as a few months and as old as nearly 80 years.^{1–6}

In their typical presentation, spinal arachnoid cysts cause progressive signs and symptoms suggesting spinal cord compression. But because a cyst can occur at any spinal level and in a patient of any age, no one clinical presentation is pathognomonic, and the clinical sequelae can differ drastically from patient to patient. Nevertheless, we can make certain generalizations: a spinal arachnoid cyst that compresses the spinal cord typically causes waxing and waning pain and progressive spastic or flaccid paraparesis, which often are exacerbated by Valsalva maneuvers.^{1,6} Spinal arachnoid cysts can also present with symptoms suggestive of an isolated radiculopathy.

Less typical presentations include noncardiac chest pain, isolated gait difficulty, and isolated urinary urgency.^{2–4}

Missed diagnosis is common

Because the symptoms are so variable and nonspecific, the diagnosis of spinal arachnoid cysts is often missed. For example, a sacral extradural arachnoid cyst can cause pain in the low back and perineal region, which is

CLEVELAND CLINIC JOURNAL OF MEDICINE VOLUME 75 • NUMBER 4 APRIL 2008 311



FIGURE 1. A magnetic resonance image of the spine in a 52-year-old woman. The cyst (arrow) was an incidental finding.

Most spinal arachnoid cysts are discovered incidentally on MRI

often relieved by lying flat and aggravated by Valsalva maneuvers.⁷

Complicating the picture, spinal arachnoid cysts can also coexist with other disorders of the central nervous system. Cases have been reported of sacral extradural arachnoid cysts coexisting with lumbar disk prolapse⁷ and of spinal arachnoid cysts located near a syrinx (a tube-shaped cavity in the spinal cord).^{3,8} A patient can have more than one spinal arachnoid cyst, or both a spinal arachnoid cyst and a concurrent intracranial arachnoid cyst or a tumor.⁹

EXTRADURAL VS INTRADURAL CYSTS

Like other types of spinal meningeal cysts, spinal arachnoid cysts can be broadly characterized as either extradural or intradural.¹⁰

Extradural cysts are extradural outpouchings of arachnoid that are contiguous with the spinal subarachnoid space via a small dural defect. They typically occur in the thoracic spine dorsal to the spinal cord, although they may be found elsewhere. **Intradural cysts** are outpouchings of arachnoid that, regardless of size, lie entirely within the dural space. Intradural arachnoid cysts are more common than extradural cysts.

Either type of cyst may or may not communicate with the subarachnoid space.^{1–3}

Other cystic lesions of the spine exist. One of the most common is the Tarlov cyst, which may look similar to a spinal arachnoid cyst, as both types of cysts are collections of cerebrospinal fluid. But, unlike typical spinal arachnoid cysts, Tarlov cysts occur only in the sacral spine and appear solely within the sacral root on radiographic imaging.

HOW DO CYSTS FORM?

How spinal arachnoid cysts start to form is open to conjecture, and several theories exist.^{1,2,7} They are often attributed to congenital defects. Another possibility is that arachnoid adhesions develop secondary to inflammation, which may arise from infection (meningitis), hemorrhage, or an iatrogenic cause such as injected contrast media or anesthetics or from the intraoperative contaminants of fibrin glue.¹¹ Some cysts are due to trauma from lumbar puncture, anesthetic procedures, or intradural surgery. Other cysts are idiopathic.

WHY DO CYSTS ENLARGE?

Several mechanisms have been proposed to explain why spinal arachnoid cysts enlarge.² The cells in the cyst wall probably do not secrete fluid: many spinal arachnoid cyst walls are composed primarily of simple connective tissue, and many completely lack an inner arachnoid lining—the cells that normally secrete spinal fluid—or have only a sparse lining.⁶ A unidirectional "valve" might let fluid in but not out. Another mechanism is pathologic distribution of arachnoid trabeculae, leading to fluid shifts within the cyst, thereby causing an increase in size.

DIAGNOSIS IS OFTEN INCIDENTAL

Spinal arachnoid cysts are rare, so an algorithm to diagnose them solely on the basis of common presenting symptoms would be impractical.

312 CLEVELAND CLINIC JOURNAL OF MEDICINE VOLUME 75 • NUMBER 4 APRIL 2008

Most spinal arachnoid cysts are asymptomatic and are discovered incidentally on magnetic resonance imaging (MRI) or myelography performed because of neck or back pain, myelopathy, or radiculopathy (FIGURE 1).⁸ Cysts in the thoracic spine may be discovered during MRI evaluation for intra-abdominal diseases, and lumbar cysts may be found during MRI evaluation for isolated hip pain.

Whenever an arachnoid cyst is discovered, one must determine whether the cyst—or another problem—is actually causing the symptoms. If treatment is to succeed, the clinical presentation must correspond to the radiographic findings. For example, removing a cervical arachnoid cyst is unlikely to relieve low back pain.

Imaging studies help evaluate pain from suspected nerve compression

Although most arachnoid cysts are found by MRI, it is inappropriate to initially order MRI to evaluate a cyst's common presenting symptoms (eg, back pain, radiculopathy).

Plain radiography should be done first. Although arachnoid cysts are composed of fluid and soft tissue, which are not easily detectable on plain films, subtle and indirect signs of a chronic, large cyst may be visible.⁵

MRI is the next step if plain radiographs do not reveal bony abnormalities that could explain a patient's symptoms.

MRI is the most sensitive and specific study for detecting a spinal arachnoid cyst^{6,12} and for assessing the extent of the cyst wall. Intravenous gadolinium contrast can help distinguish between cystic tumors, synovial cysts, and arachnoid cysts. On T1- and T2-weighted images, the signal within a cyst has the same intensity as cerebrospinal fluid (FIGURE 2).

Further studies help characterize the lesion

Diffusion-weighted MRI can help differentiate an epidermoid cyst from an arachnoid cyst. It may also help differentiate a cyst from an abscess or tumor: abscesses have areas of restricted diffusion, and tumors tend to lack cerebrospinal fluid signal in their central core. Diffusion-weighted MRI can also help evaluate spinal cord atrophy and inflammatory changes.^{1,6,12} If an arachnoid cyst accompanies a nerve root as it enters the neural foramen, this would also appear on MRI.



FIGURE 2. A magnetic resonance image in a 32-year-old woman with upper extremity weakness and spasticity. Note that the cyst fluid (arrow) has the same signal intensity as cerebrospinal fluid, appearing bright white anterior to the spinal cord.

Myelography or computed tomographic (CT) myelography were used to further characterize the form and structure of spinal arachnoid cysts discovered on MRI in most reported cases, and most authors advocate these studies.^{1,3,8,12} Specifically, CT myelography has been used to look for a communication between the intraspinal subarachnoid space and the spinal arachnoid cyst, and it is sensitive in determining whether a communication exists, although it does not pinpoint the location of the communication very well.¹² CT myelography is also invaluable for imaging the spine of patients who have contraindications to MRI.

Kinematic MRI (cine-MRI) is now widely available and can help evaluate for the presence of communications between the cyst and the subarachnoid space. Dural defects may be located by carefully scrutinizing cine-MRI images for pulsating turbulent flow voids, facilitating a more focused and minimally invasive treatment strategy.¹³ Asymptomatic cysts—even large ones need no treatment

SPINAL ARACHNOID CYSTS HUGHES AND COLLEAGUES

Neo et al¹² used cine-MRI to evaluate and plan the surgical resection of a giant spinal extradural arachnoid cyst. MRI helped determine the initial diagnosis, and a pulsating turbulent flow void was observed by cine-MRI in the area later confirmed surgically to contain the communication between the cyst and the spinal subarachnoid space.

Cine-MRI is not necessary as part of the initial diagnostic evaluation for spinal arachnoid cysts. It is of particular value only to the surgeon, who can request it if needed.

HISTOPATHOLOGY

With hematoxylin and eosin staining, the walls of spinal arachnoid cysts are typically seen as fibrous and lined by meningothelial cells.

TREATMENT

Surgery is most

likely to

succeed if

symptoms

match findings

Observe asymptomatic cysts

For incidentally discovered spinal arachnoid cysts that cause no symptoms—ie, most of them—surgery is not recommended. No correlation exists between the size of a cyst and the need for treatment. Yearly imaging should be done to detect any new abnormality and determine whether the cyst is truly benign.

If symptoms arise, reevaluation of the cyst with MRI should be immediately undertaken.

Remove symptomatic cyst if possible

For a patient with symptoms, treatment offers an excellent chance of neurologic recovery.

Aspiration of the cyst is not routinely advised. Although aspiration may intuitively seem like the best initial approach to management, it only temporarily improves symptoms. However, percutaneous aspiration under fluoroscopic guidance may be appropriate for determining whether a cyst is causing a patient's symptoms and thereby predicting whether surgery can help. Surgery should be undertaken only after careful consideration, as postoperative complications, though uncommon, may be very troublesome for both the patient and the surgeon.

Complete resection is ideal treatment. The standard treatment of an isolated spinal arachnoid cyst is complete surgical removal of the cyst.¹ Surgery typically results in excellent outcomes in terms of resolution of symptoms, and is effective across a large range of cyst sizes.

Drain cysts that cannot be resected. Unfortunately, not all isolated spinal arachnoid cysts can be fully resected, owing to their location or to intraoperative findings such as extensive adhesion of a cyst to the spinal cord. In such cases, fenestration of the cyst wall, percutaneous drainage, or shunting the cyst into the peritoneal cavity may relieve symptoms.^{1-3,6}

Minimally invasive surgical techniques have also met with some success. Neo et al¹² reported that they successfully treated a giant spinal extradural arachnoid cyst by selectively closing the dural defect with clips. Cine-MRI was used to pinpoint the communication, allowing for a focused, limited surgical approach requiring only fenestration. The dural surface of the cyst was examined with an operating microscope.

Endoscopic approaches have also been used to treat sacral extradural arachnoid cysts.⁷

SOME CASES ARE MORE COMPLEX

Managing spinal arachnoid cysts becomes more complex as cysts become more intricate in morphology and if multiple cysts exist across different vertebral levels. Surgical planning and intraoperative monitoring are also complicated if a spinal arachnoid cyst coexists with another central nervous system problem.

Cases have been reported of patients with coexisting spinal arachnoid cysts and lumbar disk herniation; in many, the latter problem was considered to be the cause of symptoms.⁷

Holly and Batzdorf³ described patients with both intradural arachnoid cysts and syringomyelia. Cysts were resected with the aid of an operating microscope, and intraoperative ultrasonography confirmed that normal pulsation of the subarachnoid cerebrospinal fluid had returned after resection. The syrinx cavities were not surgically manipulated, yet MRI taken 3 months after surgery revealed that they had significantly diminished in each case.

The best predictor of recovery in patients

who undergo surgery for spinal arachnoid cysts is if the clinical presentation correlates with the defect.^{1,7} Usually the postsurgical prognosis is good, with significant to full neurologic recovery in patients with all cyst types and clinical presentations.

REFERENCES

- Choi JY, Kim SH, Lee WS, Sung KH. Spinal extradural arachnoid cyst. Acta Neurochir (Wien) 2006; 148:579–585.
- Kumar K, Malik S, Schulte PA. Symptomatic spinal arachnoid cysts: report of two cases with review of the literature. Spine 2003; 28:E25–E29.
- Holly LT, Batzdorf U. Syringomyelia associated with intradural arachnoid cysts. J Neurosurg Spine 2006; 5:111–116.
- Liu JK, Cole CD, Sherr GT, Kestle JR, Walker ML. Noncommunicating spinal extradural arachnoid cyst causing spinal cord compression in a child. J Neurosurg 2005; 103(3 suppl):266–269.
- Prevo RL, Hageman G, Bruyn RP, Broere G, van de Stadt J. Extended extradural spinal arachnoid cyst: an unusual cause of progressive spastic paraparesis. Clin Neurol Neurosurg 1999; 101:260–263.
- Wang MY, Levi AD, Green BA. Intradural spinal arachnoid cysts in adults. Surg Neurol 2003; 60:49–56.
- Muthukumar N. Sacral extradural arachnoid cyst: a rare cause of low back and perineal pain. Eur Spine J 2002; 11:162–166.
- Takeuchi A, Miyamoto K, Sugiyama S, Saitou M, Hosoe H, Shimizu K. Spinal arachnoid cysts associated with syringomyelia: report of two cases and a review of the literature. J Spinal Disord Tech 2003; 16:207–211.
- Kurokawa R, Kawase T. Spinal arachnoid cyst causing paraplegia following skull base surgery. Neurol Med Chir (Tokyo) 2006; 46:309–312.
- Nabors MW, Pait TG, Byrd EB, et al. Updated assessment and current classification of spinal meningeal cysts. J Neurosurg 1988; 68:366–377.
- Taguchi Y, Suzuki R, Okada M, Sekino H. Spinal arachnoid cyst developing after surgical treatment of a ruptured vertebral artery aneurysm: a possible complication of topical use of fibrin glue. Case report. J Neurosurg 1996; 84:526–529.
- Neo M, Koyama T, Sakamoto T, Fujibayashi S, Nakamura T. Detection of a dural defect by cinematic magnetic resonance imaging and its selective closure as a treatment for a spinal extradural arachnoid cyst. Spine 2004; 29:E426–E430.
- Doita M, Nishida K, Miura J, Takada T, Kurosaka M, Fujii M. Kinematic magnetic resonance imaging of a thoracic spinal extradural arachnoid cyst: an alternative suggestion for exacerbation of symptoms during straining. Spine 2003; 28:E229–E233.

ADDRESS: Edward C. Benzel, MD, Cleveland Clinic Spine Institute, S80, 9500 Euclid Avenue, Cleveland, OH 44195; email benzele@ccf.org.