Headache
© 2014 American Headache Society
ISSN 0017-8748
doi: 10.1111/head.12415
Published by Wiley Periodicals, Inc.

Expert Opinion

Headache and Arachnoid Cysts

Jacob Cherian, MD; Ashwin Viswanathan, MD; Randolph W. Evans, MD

Arachnoid cysts are commonly encountered when neuroimaging is obtained for headaches. Their clinical relevance is not always immediately clear and they may confound medical management.

Key words: arachnoid cysts, headaches, migraine

(*Headache* 2014;••:••-••)

CASE HISTORY

This is a 27-year-old male with a history of headaches consistent with migraine without aura since childhood. Headaches have increased to twice a week in frequency. Past medical history is negative. Neurological examination is normal. Magnetic resonance imaging (MRI) of the brain shows a left anterior cranial fossa arachnoid cyst measuring 1.2-cm anteroposterior by 1.6-cm transverse by 1.5-cm craniocaudad without significant parenchymal compression. He is placed on a triptan with a good response.

QUESTIONS

What are arachnoid cysts and their prevalence? Are headaches or other neurological symptoms associated? What is the natural history? When is surgery indicated?

From the Department of Neurosurgery, Baylor College of Medicine, Houston, TX, USA (J. Cherian and A. Viswanathan); Department of Neurology, Baylor College of Medicine, Houston, TX, USA (R.W. Evans).

Address all correspondence to: A. Viswanathan, Department of Neurosurgery, 6501 Fannin Street, Suite NC-100, Houston, TX 77030, USA.

Accepted for publication May 21, 2014.

EXPERT COMMENTARY

Arachnoid cysts are frequently found intracranial lesions and are the most common congenital cystic abnormality in the brain.[1] They have a reported occurrence of 1.4%[2] according to a recent natural history study surveying an adult population. They consist of a confined diverticula emanating from the natural septations of the arachnoid membrane and are filled with cerebrospinal fluid (CSF). A 1-way slit-valve mechanism has been proposed by some to underlie their growth.[3,4]

RADIOLOGIC FEATURES

The radiographic features of arachnoid cysts are well defined. The cysts are extra-axial and the contents have the same signal as CSF. There may be evidence of bone remodeling on computed tomography (CT) including thinning and scalloping of the bone (Fig. 1).[1] By MRI, they are bright on T2 and dark on T1. They do not typically diffusion restrict, but occasionally they may have slightly increased signal intensity on diffusion-weighted imaging because of stasis of fluid within the cyst (Fig. 2). Epidermoid cysts can have similar appearance by CT and T2-weighted imaging, but can be differentiated by their avid diffusion-weighted intensity.

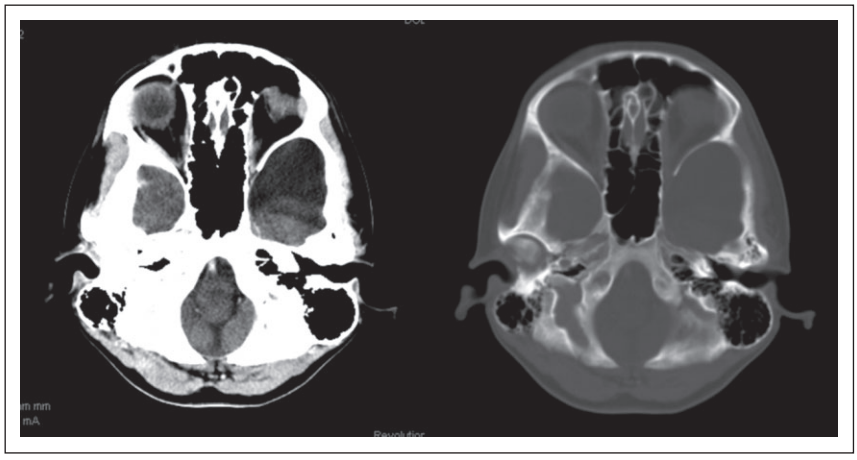

Fig 1.—Brain and bone windows of left middle fossa arachnoid cyst with remodeling of the lateral orbit and temporal bones.

PREVALENCE AND NATURAL HISTORY

Arachnoid cysts are thought of as benign lesions. They are often found during the workup of generalized presenting symptoms such as headache. They are most commonly found in the middle fossa, but can occur throughout the neuroaxis. A 2007 population-based study by Vernooij et al[5] reviewing incidental findings on brain MRI in 2000 patients with a mean age of 63.3 years found arachnoid cysts in 22 patients (1.1%). A similar 1999 study looking at a younger population by Katzman et al[6] reviewed MRIs in 1000 asymptomatic volunteers with a mean age of 30.6 years found arachnoid cysts in 3 patients (0.3%).

A recent excellent study by Al-Holou and colleagues examined the prevalence and natural history of arachnoid cysts in an adult population.[7] The charts of all 48,417 patients, 19 years or older, who underwent brain MRI at the University of Michigan over a 12-year period were retrospectively reviewed. Six hundred sixty-one patients or 1.4% were found to have arachnoid cysts, 1.8% males and 1.1% females. The top 3 locations were middle fossa (34%), retrocerebellar (33%), and cerebral convexity (14%). Thirty-five patients were found to have neurologic symptoms thought to be attributable to their arachnoid cysts; for 15, headache was the presenting symptom.

Of these 661 patients, 203 patients with 213 arachnoid cysts were able to be followed for greater than 6 months with both imaging and clinical follow-

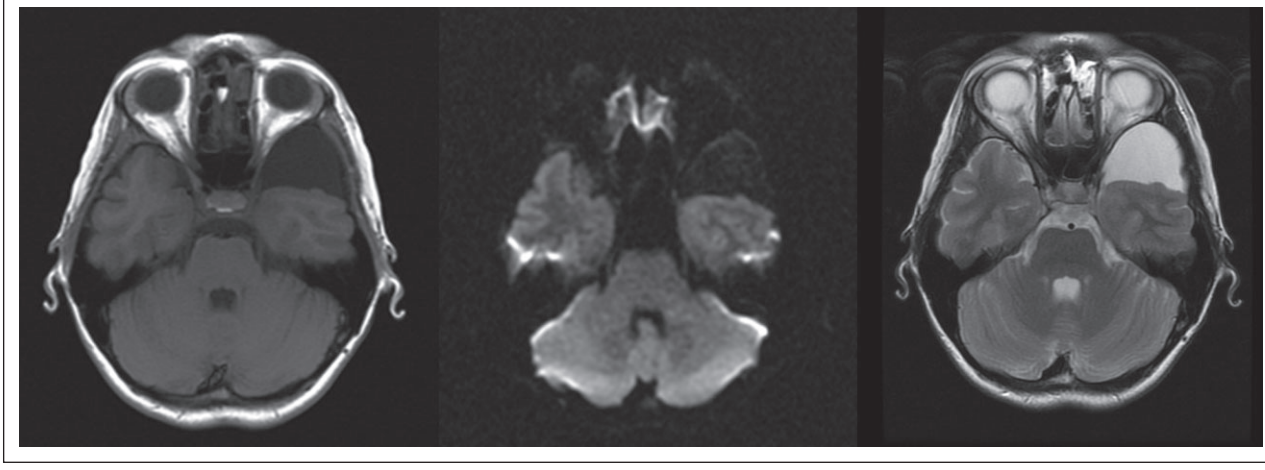

Fig 2.—T1-, diffusion-, and T2-weighted imaging characteristics of a left middle fossa arachnoid cyst.

up. Mean follow-up was 3.8 years. At the last available follow-up, only 5 cysts were larger than at presentation and 2 were smaller. Most were thus unchanged. None of the patients followed developed cyst-associated hemorrhage or symptomatic hygroma.

Two of the followed patients developed symptoms thought to be attributable to their arachnoid cyst that ultimately required surgical management. In one, a convexity arachnoid cyst measuring $4 \times 6 \times 7$ cm initially presented without symptoms. Nearly 4 years later, the patient presented with seizure and headache. The cyst was found to have enlarged and was associated with increasing mass effect and midline shift. Surgical fenestration was subsequently performed with reported improvement in symptoms. In the second case, the patient initially presented with an asymptomatic $1.6 \times 2.2 \times 1.9$-cm cerebellopontine angle arachnoid cyst. Over the course of 3.6 years, the patient developed ipsilateral hearing loss and tinnitus with no clear increase in cyst size. Subsequent cyst fenestration helped stabilize symptoms, but was not curative.

MANAGEMENT

Classically, management of arachnoid cysts has been observant and non-surgical. Surgical management has historically been reserved for cases in which lesions appeared locally compressive in patients with correlative neurologic findings or in those with obstructive hydrocephalus. Interventions have consisted of localized craniotomies with surgical marsupilization and drainage of the cyst. Endoscopic and shunting procedures have also been described.[8,9]

One recent series by Helland and Wester[10] offered surgical decompression of all arachnoid cysts in patients initially presenting with headache and dizziness. Utilizing postoperative questionnaires, they report total symptom relief in 82% of cases. Twelve percent of patients reported no relief and 6% reported worsening of symptoms. Significant complications causing permanent slight disability were reported in 2 out of the 156 cases. In their conclusion, the authors argue in favor of aggressive management of these lesions. They suggest that some of the generalized symptoms such as dyscognition that patients with these cysts present with may be secondary to local dysfunction of compressed cerebral tissue. There was, however, no clear association in fluid volume reduction and clinical improvement. The authors attempt to explain this disassociation by arguing that adults with congenital arachnoid cysts have a remodeled cranium with surplus intracranial space. Thus despite successful cyst decompression and relief of compressed brain tissue, relieved brain may not be large enough to fill the expanded intracranial space. This contrasts with a previous study by the same group[11] in children where there was a direct correlation between fluid volume reduction and clinical improvement. The authors believe that in children the mismatch between skull and brain volumes may not be as pronounced because they are operated on while the brain and skull are still growing.

In contrast, a recent series by Maher et al[12] managed 7 patients presenting with ruptured arachnoid cysts and associated subdural hygroma with conservative management only. All 7 patients had a good clinical outcome and complete symptomatic resolution within weeks to a few months after presentation. This was despite the fact that half of the patients in the series had objective findings of elevated intracranial pressure such as papilledema, cranial nerve VI palsy, and progressive macrocephaly. The authors, nonetheless, write that they would prefer surgical treatment as an initial therapy for patients with severe symptoms or an especially concerning neurological examination. They would also consider surgical treatment in those patients whose condition was refractory to initial conservative management.

Arachnoid cysts of the middle fossa may also be associated with chronic subdural hematomas, which have variably been reported as occurring in 2.4%[13] to 6.6%[14] to 17.5% of patients,[15] although another large study suggests this is a rare event.[7] Rarely, arachnoid cysts may rupture into the subarachnoid space causing a symptomatic CSF hygroma.[7] Though the exact pathogenesis for the development of subdural hematomas is not clear, 2 theories have been proposed. The first theory suggests that the

movement of fluid within the arachnoid cyst causes a higher risk of tearing the small bridging vessels between the outer arachnoid membrane and the dura mater. The second theory postulates that chronic subdural hematomas are caused by bleeding of veins within the cyst wall itself. In patients with chronic subdural hematomas, both removal of the hematoma alone,[15] and removal of the hematoma along with fenestration of the arachnoid cyst,[14] have been effective treatment strategies.

CONCLUSION

Given the overall benign natural history, we prefer a conservative, nonsurgical approach in patients who present without neurological symptoms. We advocate a nonsurgical approach also for asymptomatic patients in whom imaging studies suggest local mass effect. In patients who present with headaches and without progressive neurological deficits, a trial of medical management and close observation is warranted to help determine the underlying cause of the headaches.

In patients presenting with headache and symptoms suggestive of increased intracranial pressure, a surgery-first approach may be warranted. However, the potential benefits of surgery must be balanced with the risks of surgical intervention which include: infection, CSF leak, seizures, shunt dependence and overdrainage, and hydrocephalus. Recent literature suggests that conservative management can even be extended to, and effective for, patients with ruptured arachnoid cysts associated with subdural hygromas, papilledema, and cranial nerve palsy.[11] Surgical intervention may also be appropriate in patients with a focal neurologic finding attributable to local mass effect and in cases with hydrocephalus secondary to blocked ventricular outflow. Most often, however, arachnoid cysts are benign incidental findings that can be managed observantly. In the case illustration described above, the good response to migraine management highlights once again the importance of accurate history and good clinical acumen.

REFERENCES

1. Yousem DM. *Neuroradiology: The Requisites*, 3rd edn. Philadelphia, PA: Mosby/Elsevier; 2010.
2. Al-Holou WN, Terman S, Kilburg C, Garton HJL, Muraszko KM, Maher CO. Prevalence and natural history of arachnoid cysts in adults. *J Neurosurg*. 2013;118:222-231.
3. Halani SH, Safain MG, Heilman CB. Arachnoid cyst slit valves: The mechanism for arachnoid cyst enlargement. *J Neurosurg Pediatr.* 2013;12:62-66.
4. Schroeder HW, Gaab MR. Endoscopic observation of a slit-valve mechanism in a suprasellar prepontine arachnoid cyst: Case report. *Neurosurgery*. 1997; 40:198-200.
5. Vernooij MW, Ikram MA, Tanghe HL, et al. Incidental findings on brain MRI in the general population. *N Engl J Med*. 2007;357:1821-1828.
6. Katzman GL, Dagher AP, Patronas NJ. Incidental findings on brain magnetic resonance imaging from 1000 asymptomatic volunteers. *JAMA J Am Med Assoc*. 1999;282:36-39.
7. Al-Holou WN, Terman S, Kilburg C, Garton HJ, Muraszko KM, Maher CO. Prevalence and natural history of arachnoid cysts in adults. *J Neurosurg*. 2013;118:222-231.
8. Arai H, Sato K, Wachi A, Okuda O, Takeda N. Arachnoid cysts of the middle cranial fossa: Experience with 77 patients who were treated with cystoperitoneal shunting. *Neurosurgery*. 1996; 39:1108-1112, discussion 1112-1113.
9. Choi JU, Kim DS, Huh R. Endoscopic approach to arachnoid cyst. *Childs Nerv Syst ChNS Off J Int Soc Pediatr Neurosurg*. 1999;15:285-291.
10. Helland CA, Wester K. A population based study of intracranial arachnoid cysts: Clinical and neuroimaging outcomes following surgical cyst decompression in adults. *J Neurol Neurosurg Amp Psychiatry*. 2007;78:1129-1135.
11. Helland CA, Wester K. A population-based study of intracranial arachnoid cysts: Clinical and neuroimaging outcomes following surgical cyst decompression in children. *J Neurosurg*. 2006; 105(Suppl. 5):385-390.
12. Maher CO, Garton HJL, Al-Holou WN, Trobe JD, Muraszko KM, Jackson EM. Management of subdural hygromas associated with arachnoid cysts. *J Neurosurg Pediatr.* 2013;12:434-443.
13. Parsch CS, Krauss J, Hofmann E, Meixensberger J, Roosen K. Arachnoid cysts associated with subdural hematomas and hygromas: Analysis of 16 cases, long-term follow-up, and review of the literature. *Neurosurgery*. 1997;40:483-490.

14. Kwak YS, Hwang SK, Park SH, Park JI. Chronic subdural hematoma associated with the middle fossa arachnoid cyst: Pathogenesis and a review of its management. *Childs Nerv Syst*. 2013; 29:77-82.

15. Domenicucci M, Russo N, Giugni E, Pierallini A. Relationship between supratentorial arachnoid cyst and chronic subdural hematoma: Neuroradiological evidence and surgical treatment. *J Neurosurg*. 2009;110:1250-1255.