Long-term follow-up of a multiloculated arachnoid cyst of the middle cranial fossa

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Abstract

Arachnoid cysts are benign intracranial lesions that are typically diagnosed incidentally. We describe the case of a 56-year-old man who presented with a multiloculated arachnoid cyst of the middle cranial fossa that extended into the sphenoid sinus. The lesion was identified on computed tomography of the head, which had been obtained for an unrelated investigation. However, establishing a definitive diagnosis proved to be difficult. Because the cyst had caused extensive skull base erosion, the patient was managed conservatively with close observation. We report the radiographic progression of this lesion during more than a decade of follow-up, and we review the literature pertaining to the presentation, pathophysiology, and treatment of arachnoid cysts.

Introduction

Arachnoid cysts are uncommon benign masses.\(^1\,\^2\) They are more common in men than in women. This entity was first described by Bright in 1831 as a “serous cyst forming in connection with the arachnoid.”\(^3\) Since then, a substantial number of reports have described the presentation and management of arachnoid cysts. However, the patterns of their occurrence and their associated pathophysiology have yet to be entirely elucidated.

Approximately 50% of intracranial arachnoid cysts are located in the middle cranial fossa and 30% in the posterior fossa; most (64%) arise on the left side.\(^2\,\^4\) Our review of the English-language literature found only 2 cases in which an arachnoid cyst extended into the sphenoid sinus.\(^5\,\^6\) Moreover, we found only one citation that pertained to a multiloculated arachnoid cyst; that report concerned 3 patients with cervical spinal cord postsurgical collections that appeared to be arachnoid cysts.\(^7\)

In this article, we report a case of multiloculated arachnoid cyst that originated in the middle cranial fossa and extended into the sphenoid and ethmoid paranasal sinuses, causing substantial skull base erosion. The diagnosis in this case initially remained uncertain, even after we performed computed tomography (CT), magnetic resonance imaging (MRI), and surgical exploration for a presumed erosive sphenoid sinus mucocele.

Case report

A 56-year-old man presented to an outside hospital in 1994 after he had been involved in a motor vehicle accident. At that time, he had a history of chronic rhinosinusitis that had been treated with endoscopic sinus surgery. CT of his head identified a sphenoid sinus mass that had eroded the left skull base (figure 1, A). MRI confirmed the presence of an extra-axial cystic lesion in the sphenoid sinus that extended to the floor of the middle cranial fossa (figure 1, B and C). The presumptive diagnosis was a large mucocele, and the patient underwent an endoscopic exploration. Unfortunately, the posterior ethmoid roof was violated during the exploration, and a CSF leak occurred. The procedure was terminated, and Gelfoam was placed over the site of the skull base lesion. Definitive drainage of the presumed mucocele was not performed.

The patient was transferred to our institution with a significant fever and symptoms consistent with meningitis and a CSF leak. The diagnosis of meningitis was confirmed by lumbar puncture, and intravenous antibiotic therapy was initiated. A neuroradiologist reviewed the preoperative CTs and MRIs and those obtained during the immediate postoperative period but could not make a definitive diagnosis. We then entertained a differential diagnosis of an expansile mucocele extending intracranially or a temporal lobe arachnoid cyst extending into the sphenoid sinus.

After 2 days, the patient improved clinically and underwent a second endoscopic exploration. A significant...
skull base defect and active CSF leak were identified in the area of the right posterior ethmoid sinus roof (planum ethmoidale). After the Gelfoam was removed, a nasal septal mucoperiosteal graft was harvested from the opposite side of the nose, and the skull base defect was repaired. The sphenoid sinus ostium was identified and enlarged. The fluid was evacuated, and no active CSF leak was identified within the sphenoid sinus. It was not immediately evident whether the lesion did indeed represent a mucocele with fluid content or a meningocele arising from an arachnoid cyst. The lining of the cyst was firmly adherent to thinned dura, and extensive bone erosion was observed.

An intraoperative biopsy of the cyst lining submitted for frozen-section analysis was nondiagnostic; it was reported as just necrotic tissue. Repeat biopsies were performed during the procedure in an attempt to establish a definitive diagnosis. During removal of another section of the cyst lining for pathologic evaluation, CSF began to leak through the extremely thin dura. However, because a mucocele could not be ruled out on the basis of frozen-section analysis, a graft was not placed over a mucosal membrane lining the thinned dura.

The patient was managed with a spinal drain for 72 hours postoperatively. His recovery was uneventful, and he experienced no CSF rhinorrhea. The final pathology report identified necrotic/fibrotic tissue that was possibly mucosal and possibly arachnoid. The patient has been followed long-term with regular MRI scans (figure 2), and the lesion has remained relatively stable. Based on the lesion’s lack of progression and our review of the imaging and pathology, we believe that this lesion most likely represented an arachnoid cyst extending into the sphenoid sinus as a poorly communicating meningocele. In early 2007, the patient remained free of neurologic and otolaryngologic complaints.

Discussion
For a lesion to be considered an arachnoid cyst, it must meet three criteria: (1) it must be enveloped by an arachnoid membrane, (2) it must contain arachnoidea mater cells, and (3) it must contain CSF. These requirements, by definition, ensure duplication of the arachnoid membrane, leading to a distinct cyst.

Arachnoid cysts are divided into two types: congenital and acquired. Congenital cysts are considered to occur secondary to aberrant embryogenesis of the central nervous system. Patients who present with arachnoid cysts early in childhood have associated central nervous system malformation, which often leads to obstructive hydrocephalus at the level of the foramen of Monro. Acquired arachnoid cysts usually arise after trauma, infection, or hemorrhage.

The formation of arachnoid cysts after surgical trauma—especially in the spine—has been noted previously. It has been suggested that post-traumatic changes can lead to pressure increases that foster an environment suitable for arachnoid layer separation that leads to cyst formation. In trauma patients with skull damage, the arachnoid membranes can invaginate into the intraosseous defects and cause a post-traumatic intraosseous arachnoid cyst. Furthermore, trauma can lead directly to adhesions of the arachnoid membrane. There is speculation that loculated accumulations involving arachnoid fluid may develop after trauma to the spine.

Autopsy studies have indicated that approximately 0.5% of the population has an arachnoid cyst.
percentage is markedly higher than the percentage of patients who present with symptoms of this lesion, regardless of etiology. It is unclear why some arachnoid cysts expand and cause symptoms while others remain stable. A number of cases of spontaneous regression of arachnoid cysts have been reported.

Four pathophysiologic mechanisms have been proposed to explain arachnoid cyst formation:

- The first theory involves a ball-valve mechanism, in which the cyst is in communication with the CSF compartment. According to this theory, clefts in the cyst wall allow fluid to enter the cyst from the subarachnoid space, but since the fluid cannot regress, cystic expansion occurs. The plausibility of this hypothesis has been proven by cine MRI. This hypothesis correlates with the supposition that post-traumatic arachnoid cysts form or enlarge during periods of rapid cerebral growth or an increase in CSF pressure.

- The second hypothesis applies to noncommunicating arachnoid cysts. This theory maintains that there is a flow gradient through endothelial cells into the cyst that is generated by differences in fluid composition between the CSF and the cystic fluid. However, this hypothesis neglects the fact that the subarachnoid space and cystic fluid have been demonstrated to be osmotically similar. Therefore, one would not expect to see a passive transcellular flow of fluid in two spaces with identical composition.

- According to the third hypothesis, cystic fluid expansion occurs secondary to an increase in the production of CSF by the arachnoid cells that line the collection. This hypothesis is supported by the finding that the arachnoid lining of the cysts has characteristics of secretory cells, including the presence of apical cell surface Na⁺/K⁺ ATPase. This hypothesis is tenable whether or not the cyst communicates with the CSF compartment. No speculation has been offered to explain what inciting events would lead to the growth of a seemingly stable cyst.

- The fourth hypothesis holds that the accumulation of fluid is the result of CSF pulsation and extravasation of venous or possibly arterial fluid.

The presentation of an arachnoid cyst varies markedly, depending on its etiology and location. As noted previously, many patients with arachnoid cysts are asymptomatic, and many of these cysts are found incidentally as a byproduct of the recent increase in the use of radiologic imaging to evaluate the central nervous system. When symptoms are present, they usually manifest secondary to the direct compression of surrounding structures. In children, direct compression can lead to macrocephaly secondary to obstructive hydrocephalus. Gosalakal recently speculated that children with asymptomatic congenital arachnoid cysts are at increased risk of developing attention-deficit hyperactivity disorder and experiencing a delay in speech development.

In symptomatic adults, common presentations include headache, seizures, and cranial nerve abnormalities. In the elderly, arachnoid cysts have been associated with both dementia and normal-pressure hydrocephalus. They have also been implicated in psychosis. Regardless of the etiology, the vasculature on the cyst may rupture spontaneously or after a minor trauma, leading to hemorrhage and a subsequent subdural hematoma. Hemorrhage into the cyst can occur even in the absence of trauma. In rare cases, symptoms related to arachnoid cysts occur secondary to the rupture of these entities and...
the formation of a subdural hygroma and intracranial hypertension. Arachnoid cysts of the middle cranial fossa have also been documented to cause local bulging and thinning of the temporal bone and erosion of the sphenoid bone in the context of elevated intracranial pressure. Finally, speculation has recently been expressed that regression of an arachnoid cyst that abuts the paranasal sinuses can lead to pneumosinus dilatans—that is, an abnormal enlargement of the paranasal sinuses. Occasionally, extremely large cysts remain asymptomatic. This was perhaps the case in our patient, who had a large lesion that likely originated in the middle fossa and expanded into the sphenoid sinus. Alternately, it is possible that symptoms that might have been caused by the cyst’s erosion through the sphenoid bone were obscured by the patient’s chronic sinusitis.

The treatment of arachnoid cyst is controversial. Certainly, almost all authors are in favor of treatment if the patient is symptomatic, especially in the setting of elevated intracranial pressure or progressive hydrocephalus. However, some authors have questioned the efficacy of treatment for certain symptoms, such as seizures and headaches, that are often attributed to the presence of an arachnoid cyst. Neurosurgical options for the management of symptomatic arachnoid cysts include cystoperitoneal shunting, complete excision via a cranietomy, and fenestration. Fenestration of an arachnoid cyst is associated with a higher recurrence rate (30%) than is shunting (0%), but it avoids the permanent ramifications of shunting dependence. A neuroendoscopic approach to intracranial arachnoid cysts has the benefits of being minimally invasive—to a similar degree as shunting—and being effective without shunt dependence. Additionally, there may be a role for sinonasal endoscopic treatment for selected patients with a symptomatic arachnoid cyst that abuts or invaginates into the paranasal sinuses.

In conclusion, our case demonstrates that the diagnosis of arachnoid cyst may be difficult when a cyst extends extracranially and that it can be confused with a mucocele in the setting of chronic rhinosinusitis. Accordingly, the differential diagnosis of arachnoid cyst should be considered for lesions that involve the sphenoid sinus and extend intracranially. Because the dura adjacent to the lesion may be thinned, biopsy may be difficult, fraught with the potential for causing CSF rhinorrhea, and nondiagnostic. In our patient, long-term follow-up did not detect any disease progression over time.

References