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Case Report

Parasellar arachnoid cyst presenting with a nonpupil sparing third nerve palsy mimicking a posterior communicating artery aneurysm in an adult

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Abstract

Background: Arachnoid cysts are congenital lesions that contain fluid identical to cerebrospinal fluid (CSF). They usually do not communicate with CSF spaces. The vast majority of arachnoid cysts are congenital asymptomatic lesions that are discovered incidentally. Those lesions that do become symptomatic typically present in childhood with signs and symptoms of intracranial hypertension, seizures, and focal neurologic deficits specific to cyst location.

Case Description: A rare case of a parasellar arachnoid cyst presenting with oculomotor palsy is presented. The patient is a 45-year-old male who presented with acute onset diplopia and frontal headache. Neurologic examination revealed right ptosis, pupillary dilation, and opthalmoparesis consistent with an oculomotor palsy. Computed tomography (CT) scan and lumbar puncture failed to reveal evidence of a subarachnoid hemorrhage. Magnetic resonance imaging (MRI) of the brain demonstrated a 1 cm right parasellar nonenhancing mass that was hyperintense on T2 flair and with a fluid-fluid level concerning for a thrombosed posterior communicating artery (PCommA) aneurysm. There was an additional finding of a left occipital pole intraparenchymal hemorrhage in the setting of multiple hereditary cavernomas. Formal cerebral angiography revealed normal intracranial and extracranial vasculature. The patient was taken to the operating room for a right frontotemporal craniotomy, which revealed compression of the right oculomotor nerve by an arachnoid cyst. The cyst was fenestrated and resected with decompression of the oculomotor nerve. Postoperatively, the third nerve palsy had completely resolved.

Conclusions: The above case demonstrates that arachnoid cysts should be considered in the differential for patients presenting with nonpupil sparing third nerve palsy and require timely surgical intervention. As is the case for an expanding PCommA aneurysm, prompt decompression results in the best chance for recovery of oculomotor nerve function.

Key Words: Arachnoid cyst, cavernoma, oculomotor palsy

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INTRODUCTION

Arachnoid cysts are well-described congenital fluid-filled structures lined with meningothelial cells first reported in the 16th century by Dutch Anatomist Pieter Paux and first described by Bright in 1831.[6] These cysts generally do not communicate with cerebrospinal fluid (CSF) spaces and contain fluid that is identical to CSF. Presented here is a case of an arachnoid cyst in an adult presenting as an acute onset unilateral nonpupil sparing third nerve palsy mimicking a thrombosed posterior communicating artery (PCommA) aneurysm. While two cases of arachnoid cysts at the level of the cerebral peduncles presenting with isolated unilateral third nerve palsies have been documented in the literature, it is exceedingly rare and the demographics of the patients are variable. Both cases in the literature occurred in very young children, and this case is therefore the first report of such a diagnosis in an adult.

Ashker *et al.* described a case similar to the one presented here, in which a 3-month-old boy presented with anisocoria, a sluggishly reactive right pupil and isolated unilateral internal opthalmoplegia.[2] Imaging several months later revealed a right-sided arachnoid cyst in the interpeduncular cistern compressing the third nerve exiting the brain stem. The magnetic resonance imaging (MRI) findings in this child were classic. Hustler *et al.* presented a similar case in a 16-month-old boy who presented initially with right-sided mydriasis and subsequently developed a progressive oculomotor palsy over the ensuing 14 months. He was found to have an arachnoid cyst located anterior to the right cerebral peduncle, which pushed the 3rd nerve inferiorly as it exited the midbrain. Subsequent fenestration of the cyst improved the patient's mydriasis; however, he developed oculomotor paralysis postoperatively, which did not resolve .[4]

Additionally, there are reports of arachnoid cysts of the cavernous sinus presenting with third nerve palsies;[3] however, these lesions are far distal to the lesion discussed in this case report. A complete discussion of cavernous sinus arachnoid cysts is beyond the scope of this report.

The above described cases, while rare, still follow the general rule that arachnoid cysts, when symptomatic, most often present early in life. The case presented here is similar with respect to the constellation of symptoms; however, it differs in that this patient presented with acute onset of symptoms and presented as an adult, and the location of the cyst is different than any of the other reported cases.

CASE REPORT

The patient is a 45-year-old male with a past medical history significant for multiple hereditary cavernous hemangiomas and Hodgkin's lymphoma in remission following treatment with chemotherapy and radiation. He presented to his primary care physician with acute onset double vision, frontal headaches, and lid lag of the right eye. On computed tomography (CT) imaging [Figure 1], he was found to have a left occipital hemorrhage and was transferred to this facility for further work-up and management. On examination in the emergency room, he was found to have right eye ptosis, papillary dilation as well as restriction of leftward and upgaze consistent with a right third nerve palsy. There was no evidence of subarachnoid blood on the CT scan. A lumbar puncture was performed, which was unremarkable. MRI of the brain [Figure 2] demonstrated a 2 cm left occipital pole hemorrhage and a 1 cm nonenhancing right parasellar mass with a fluid-hematocrit level that was hyperintense on T2 flair imaging without diffusion restriction that appeared to compress the right third cranial nerve [Figure 3]. These findings were concerning for a

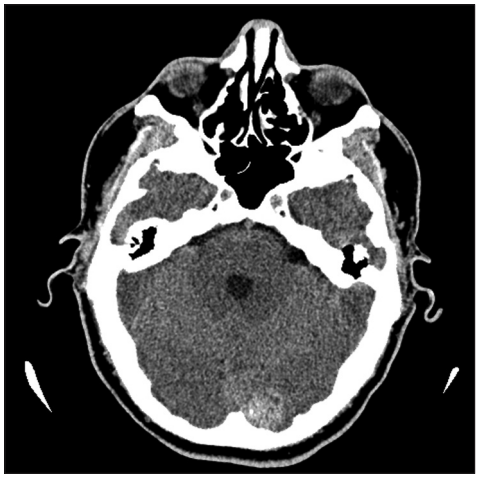

Figure 1: Head CT without contrast demonstrating hemorrhage in the left occipital pole

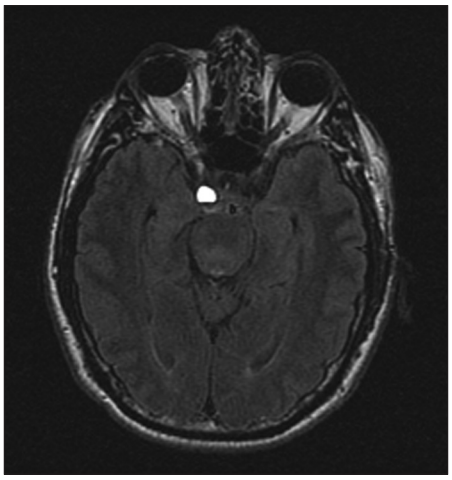

Figure 2: T2 FLAIR MRI revealing 1 cm hyperintense parasellar mass with a possible fluid-hematocrit level (a) and T1 weighted image with gadolinium that demonstrates a nonenhancing parasellar mass (b)

thrombosed PCommA aneurysm. Subsequent CT angiography of the head/neck and conventional cerebral angiography revealed normal intracranial and extracranial vessels without filling of the right parasellar mass.

The patient was taken to the operating room and underwent a right frontotemporal craniotomy. Intraoperatively, a right parasellar cystic lesion resembling an arachnoid cyst was identified, medial to the third nerve, which was pushed laterally [Figure 4]. The cyst was fenestrated and clear fluid was expressed, which appeared grossly similar to CSF. The fluid was easily aspirated, and there was no evidence of hemosiderin deposition in the cyst wall or of blood products within the cyst. This is in contrast to the suggestion of MRI, which raised the possibility of the content being hemorrhagic. The wall of the cyst was then carefully dissected and sent for permanent section analysis, which confirmed the diagnosis of an arachnoid cyst [Figure 5].

Immediately following surgery, the third nerve deficit improved but was still present. At his routine 2-week postoperative visit, he had subjective complaints of intermittent diplopia without evidence of a third nerve palsy on neurologic examination. When he presented for his three-month postoperative visit, he was symptom free and neurologically intact. MRI at that time revealed no evidence of a residual or recurrent arachnoid cyst [Figure 6].

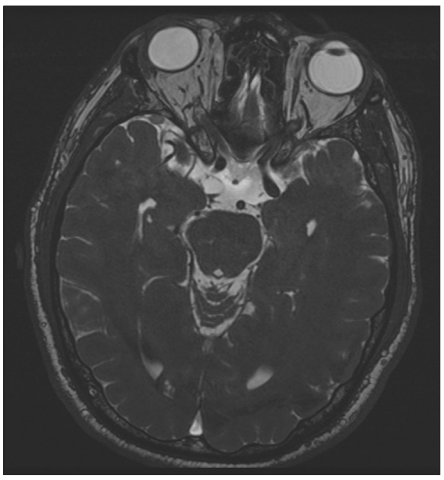

Figure 3: FIESTA MRI revealing the course of the right third nerve as it meets the arachnoid cyst

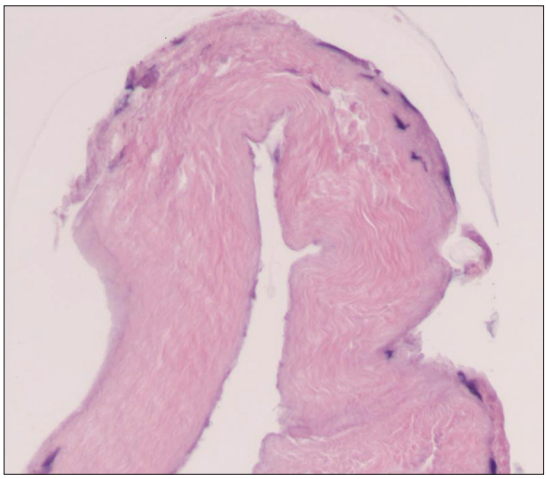

Figure 5: Formalin-fixed paraffin-embedded slides stained with hematoxylin and eosin. Sections showed a fragment of dense fibrous connective tissue attached to a long thin fibrous piece of tissue. There was no inflammation or other pathological changes. The thin fibrous material is likely leptomeningeal tissue, while the more fibrous fragment could be a portion of dura or fibrotic leptomeninges. (H and E, ×200)

DISCUSSION

Arachnoid cysts are fluid-filled CSF-containing structures typically found incidentally on imaging. They are widely thought to be congenital abnormalities, and as such, they may be associated with hypoplasia of surrounding brain tissue and focal bony expansion of the skull overlying the cyst. There are reports of arachnoid cyst development

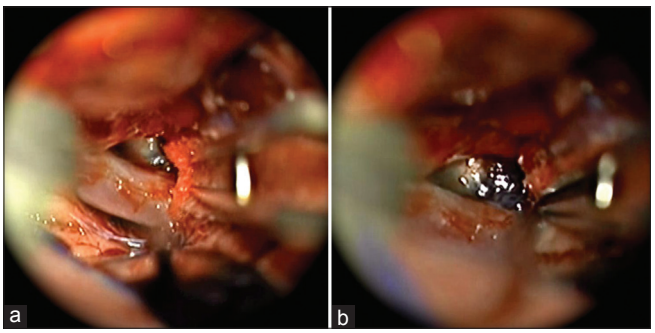

Figure 4: Intraoperative images demonstrating a frontotemporal approach to the middle fossa. Internal carotid artery and optic nerve (a). Arachnoid cyst bounded inferiorly by the internal carotid artery (b)

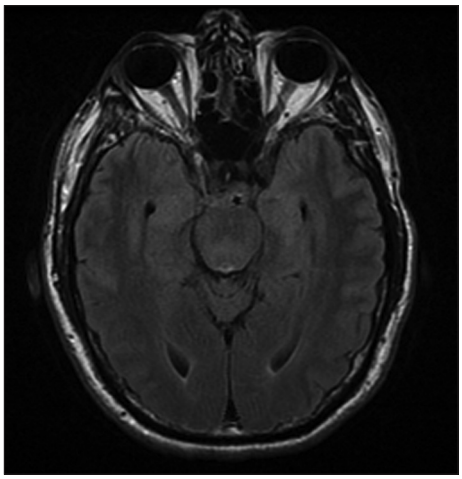

Figure 6: 3-month postoperative MRI demonstrating resolution of T2 FLAIR signal abnormality consistent with complete surgical resection of the right parasellar arachnoid cyst

as a consequence of trauma, infection, or hemorrhage; however, the pathologic components of these cysts are distinctly different from true congenital arachnoid cysts. Most arachnoid cysts are asymptomatic. The presentation of symptomatic cysts depends on the location and size of the cyst and usually occurs within the first two decades of life. Patients commonly present with signs/symptoms of elevated intracranial pressure (nausea/vomiting, headaches, visual disturbances), seizures, sudden deterioration due to hemorrhage or cyst rupture and focal neurologic deficits corresponding to cyst location. Rengachary and Watanabe[8] found that approximately 50% of arachnoid cysts occur in the sylvian fissure and middle fossa. Other common locations include the cerebellopontine angle, cerebellar vermis as well as the suprasellar, interhemispheric, and supracollicular spaces.[7]

On CT imaging, arachnoid cysts appear as extraparenchymal hypodense cystic masses approximating CSF with smooth, noncalcified walls. The cyst walls do not enhance with intravenous contrast. MRI typically reveals a cystic mass that is T2 hyperintense and typically without contrast enhancement. Arachnoid cysts generally do not exhibit restricted diffusion. Depending on the protein content of the cyst, the signal intensity on T1 imaging is variable.

The differential diagnosis of acute onset unilateral nonpupil sparing third nerve is fairly narrow and should immediately raise concern for a compressive lesion. The most common culprit is an expanding PCommA aneurysm; however, third verve tumors, uncal herniation, and mass lesions of the cavernous sinus can also produce a nonpupil sparing third nerve palsy. Approximately 90% of cases of aneurysmal third nerve palsy are due to a PCommA aneurysm; the remaining 10% of cases are due to aneurysms of the cavernous portion of the internal carotid artery or basilar tip, particularly superior cerebellar artery aneurysms.[4,5]

To date, there have been only a small number of reported cases of an arachnoid cyst presenting with unilateral nonpupil sparing third nerve palsy. The location of the arachnoid cysts in these cases is slightly more proximal to the case presented earlier and these cases occurred in children who were aged less than 18 months at the time of presentation. Further, these children presented gradually with variable components of oculomotor palsy and both cases resulted in residual oculomotor palsy following surgical intervention. The earlier case illustrates an unusual cause of a nonpupil sparing third nerve palsy caused by a parasellar arachnoid cyst in an adult whose symptoms resolved following surgical resection. The difference in outcome between the two previously reported cases and the case presented here is likely related to the timing of onset and subsequent operative intervention.

In this case, a coincident left occipital pole hemorrhage secondary to multiple hereditary cavernomas was noted. There was no indication for surgical intervention for an incidentally discovered asymptomatic cavernoma. While there are reports in the literature of coincident cavernomas and arachnoid cysts, there is no evidence to support a common underlying pathogenesis for the two distinct lesions.[1,9,10] However, venous pulsations and stasis are potential mechanisms for the enlargement of arachnoid cysts.[9] It is theoretically possible that a previously asymptomatic congenital arachnoid cyst may have enlarged later in life due to venous and CSF outflow obstruction from an acute hemorrhagic event. This provides a potential mechanism for the unusual acute presentation of arachnoid cyst in the fourth decade of life. It should be noted that the patient did not present with any focal neurological symptoms localizable to the cavernoma in question.

The above case demonstrates that arachnoid cysts should be considered in the differential for patients presenting with nonpupil sparing third nerve palsy and require timely surgical intervention. As is the case for an expanding PCommA aneurysm, prompt decompression results in the best chance for recovery of oculomotor nerve function.

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