Association between an arachnoid cyst and intracranial aneurysms misdiagnosed as a cystic tumor with a mural nodule

Case report and review of the literature

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✓The authors report on a 63-year-old man initially admitted to an outside hospital for altered mental status and respiratory distress. A head computed tomography scan disclosed a right frontal cystic mass, suspected to be a neoplasm. An open biopsy was performed at an outside institution, and on visualization of the cyst, an aneurysm was found incidentally. Postoperatively, an angiogram and magnetic resonance image confirmed the presence of a distal right M₁ segment aneurysm. The patient was transferred to our institution where, in addition to the middle cerebral artery lesion, a right anterior choroidal artery aneurysm was found intraoperatively; the necks of both aneurysms were clipped successfully. A review of the literature revealed 14 additional cases of intracranial aneurysms associated with arachnoid cysts. Data in the present report highlight the importance of considering an intracystic aneurysm in the differential diagnosis when reviewing cases that involve a cystic mass with a mural nodule. The authors provide a comprehensive summary of documented cases of aneurysms associated with arachnoid cysts. In addition, they include a discussion of prevailing thoughts on the origin and evolution of arachnoid cysts.

KEY WORDS • aneurysm • arachnoid cyst • craniotomy • mural nodule

IN GENERAL, when CT scans reveal a cystic mass with a mural nodule, a malignant glioma or other tumor is highly suspected.¹ There have been sporadic reports of arachnoid cysts associated with aneurysms resembling a cystic mass with a mural nodule, but no review in which these findings are summarized. Arachnoid cysts have been previously associated with chronic subdural hematomas^{2,9,11,18,30} and less frequently with aneurysms. More arachnoid cysts are being diagnosed given the increased frequency of neuroimaging procedures. On neuroimages, these lesions appear as smooth-bordered, noncalcified, extraparenchymal cystic masses with a density similar to CSF and no enhancement with the intravenous administration of contrast material. They can cause expansion and remodeling of nearby bone, which typically depicts its chronic nature.^{9,10,28,30} Most occur in the anterior half of the middle cranial fossa.³

An arachnoid cyst is a leptomeningeal cyst, perhaps congenital, forming from the splitting of arachnoid membranes. It can be categorized as primary or secondary. Primary arachnoid cysts are developmental abnormalities, whereas secondary arachnoid cysts can result from trauma, infection, surgery, or previous hemorrhage.^{7,9,15} A number of hypotheses have evolved regarding the origins of

arachnoid cysts and their potential relationship to intracranial aneurysms. Here, we describe a case encountered at our institution and review the literature documenting additional institutional experiences with arachnoid cysts associated with aneurysms.

Case Report

History. This 63-year-old man, who was initially admitted to an outside hospital for pneumonia, presented with an altered mental status and respiratory distress. A head CT was obtained and disclosed a right frontal cystic mass (Fig. 1). An open biopsy was performed given the suspiciousness of the lesion. On visualization of the cyst, an aneurysm was found incidentally. Postoperatively, an angiogram confirmed the presence of a distal right M₁ segment aneurysm (Fig. 2).

Examination. The patient was transferred to our institution, where he arrived neurologically intact. An MR image confirmed the presence of an enhancing right frontal cystic lesion measuring approximately 4 × 4.8 × 4.5 cm (transaxial × anteroposterior × craniocaudal) as well as the distal right MCA aneurysm incorporated along the inferior margin of the lesion (Fig. 3). The cyst itself appeared to contain material consistent with CSF on MR imaging.

Operation. The patient was taken to the operating room and underwent a standard right-sided craniotomy. The

Abbreviations used in this paper: ACA = anterior cerebral artery; AChA = anterior choroidal artery; CSF = cerebrospinal fluid; CT = computed tomography; MCA = middle cerebral artery; MR = magnetic resonance; SAH = subarachnoid hemorrhage.

FIG. 1. *Left and Right:* Axial non-contrast-enhanced CT scans revealing a right frontal cystic mass with an intracystic nodule.

arachnoid cyst was marsupialized and drained. Intraoperative findings revealed clear CSF from within the cyst on decompression. Inferiorly, hemosiderin staining was noted to overlie the dome of the right MCA aneurysm. On further dissection, a right AChA aneurysm was discovered. The necks of both the MCA and AChA aneurysms were clipped using an appropriate microvascular technique. The pathological specimens disclosed reactive gliosis surrounding the cyst. Focal vascular proliferation and chronic inflammation were also noted, suggesting a cyst wall lining.

Discussion

In a review of the available literature, we found that six of 15 cases reported involved female patients (Table 1).^{3,4,11–14,20,26,31,35} Symptoms typically included headache, nausea, and vomiting. Most arachnoid cysts were found in the anterior half of the middle cranial fossa. Six patients had right-sided aneurysms, whereas nine had left-sided ones. Patients in four cases were noted to have multilocular cysts, three of which involved the posterior circulation. Five cases involved MCA aneurysms, and all of the patients in these cases presented with unilocular cysts. Four

FIG. 2. Transorbital cerebral angiogram revealing a right MCA aneurysm (arrow).

FIG. 3. Contrast-enhanced MR image revealing a right frontal cystic mass with an intracystic enhancing nodule, confirmed to be an MCA aneurysm on angiography.

cases were described as having aneurysmal calcification and/or thromboses, and rupture was not apparent on presentation. In two cases, including the one featured in the present report, a second aneurysm was noted on the same side.¹¹ In all documented cases, the dome of the aneurysm always projected toward the cystic space.

Authors of five reports documented the communication between an aneurysm and arachnoid cyst within the context of a recent aneurysm rupture and intracystic hematoma. The arachnoid cyst was located in the middle cranial fossa in 10 cases; four involved an intracystic hematoma and three had xanthochromic fluid.^{3,4,11,13,14} Subarachnoid hemorrhage was present in only three of the 15 cases. Only two cases entailed a subdural hematoma, suggesting a local tear in the arachnoid membrane during aneurysm rupture.^{4,13}

Authors of three cases noted xanthochromic fluid on surgically opening the cyst.^{12,20,35} In the case described by Marcoux et al.,²⁰ a cyst had formed 17 months after an SAH and embolization of an ACA aneurysm. These authors ascribed this acquired arachnoid cyst to a postinflammatory process. Two other cases, including the one featured in the present report, demonstrated hemosiderin layering on preoperative MR imaging, suggesting prior hemorrhage.²⁶

Starkman et al.³⁰ defined arachnoid cysts as closed compartments situated within the arachnoid and segregated from subarachnoid spaces. We propose that the patient in the present case could have experienced a remote episode of SAH with subsequent arachnoiditis and adhesion formation initiating the development of a secondary arachnoid cyst. Given the contrast enhancement and hemosiderin layering at the base of the cyst on MR images and on intraoperative examination, respectively, we admitted that an age-indeterminate SAH or parenchymal hemorrhage might have occurred, disrupting the preexisting subarachnoid membranes. Based on our review, once an arachnoid cyst has formed, an intracystic aneurysm rupture is likely to appear initially as an intracystic hematoma, with fewer instances of acute SAH.

TABLE 1
Literature summary of patients harboring an arachnoid cyst associated with an intracranial aneurysm*

Authors & Year	Age (yrs), Sex	Clinical Symptoms	Aneurysm Rupture?	SAH?	Aneurysm Location	Thrombosis	Ca ⁺⁺	Additional Aneurysm	Cyst	Intracystic Hematoma	Aneurysm Dome Location w/ Respect to Cyst	Outcome
Jinkins et al., 1987	40, M	headache, nausea, vomiting	yes	yes	rt MCA bifurcation	none	none	none	unilocular	positive	uncertain relationship	no treatment
Hirose et al., 1995	45, M	headache	yes	no	lt ICA bifurcation (saccular)	none	none	lt MCA	unilocular	positive	posteriorly & perforating cyst wall	surgical clipping
Barker et al., 1998	29, F	nausea, vomiting, photophobia, neck stiffness	yes	yes	lt PCoA	none	none	none	unilocular	positive	uncertain relationship	surgical clipping
Hirota et al., 1999	71, M	speech & writing difficulties	no	no	lt MCA	none	none	none	unilocular	xantho	anteromedially into cyst wall	surgical clipping
Huang et al., 1999	61, F	mild headache	yes	no	lt MCA	none	none	none	unilocular	positive	medial wall of cyst	surgical clipping
Sato et al., 2000	46, M	headache	no	no	rt ACA	none	none	none	unilocular	negative	medial wall of cyst	surgical clipping
	57, M	unknown	no	no	rt or lt giant ACA	prominent	prominent	none	unilocular	negative	abutting left margin of cyst	died of aneurysm rupture
	51, M	seizures 2 yrs previously	no	no	rt PCA	prominent	prominent	none	multilocular	negative	anterosuperiorly into walls of cyst	biopsy: reactive gliosis, embo offered
	62, M	none	no	no	lt ICA bifurcation	none	prominent	none	multilocular	negative	superiorly into cyst wall	endovascular treatment offered
	35, F	headache	no	no	rt PCA	none	none	none	multilocular	not evaluated	anteromedially into cyst wall	hydrocephalus, surgical clipping
Zanini et al., 2000	35, M	headache	no	no	lt ICA bifurcation	none	none	none	unilocular	xantho	medial wall of cyst	surgical clipping
Takai et al., 2001	64, F	alexia & anomic aphasia	no	no	lt PCA	partial	partial	none	multilocular	negative	medial wall of cyst	embo, endoscopic septostomy, & cyst fenestration
Marcoux et al., 2002	62, F	headache, urinary frequency, confusion	yes†	yes	lt A ₁ -A ₂ junction	none	none	none	unilocular	xantho	superiorly into cyst wall	cyst fenestration after initial fenestration
Berthouma et al., 2006 present case	37, F	headache, rt hemiparesis	yes	no	lt MCA bifurcation (saccular)	none	none	none	unilocular	positive	adherent to posterior wall of cyst	surgical clipping
	63, M	headache, respiratory distress	no	no	rt MCA bifurcation	none	none	rt AChA	unilocular	negative	anterior wall of cyst	surgical clipping; biopsy: reactive gliosis

* Ca⁺⁺ = calcification; embo = embolization; ICA = internal carotid artery; PCA = posterior cerebral artery; PCoA = posterior communicating artery; xantho = xanthochromic.

† Cyst formed 17 mos postembolization.

With the presence of hemolyzed blood in the subarachnoid spaces, a cascade of events occurs, creating a fibroproliferative reaction that results in arachnoiditis. Data from molecular studies have shown that the increase in procollagen propeptides found in the CSF after SAH can be correlated with the increased rate of meningeal collagen synthesis.²²⁻²⁴ A number of authors have documented the formation of spinal arachnoiditis and an arachnoid cyst after SAH, particularly those involving posterior fossa circulation aneurysms.^{16,19,32}

Sato et al.²⁶ have also described a case that supports the concept of cyst formation following parenchymal hemorrhage. Three additional cases have entailed xanthochromic fluid on opening the arachnoid cyst, again suggesting a previous SAH.^{12,20,35} Interestingly, all documented cases have involved an aneurysm dome projecting toward the cystic space. In particular, five cases involved aneurysm rupture and intracystic hematoma. These examples also suggest a previous SAH event that initiated a postinflammatory cascade, creating a cystic cavity.

Fox and Al-Mefty⁸ described a pediatric case with a history of head trauma. The initial cyst origin was an outpouching of an imperforate membrane of Lilliequist by a previous hemorrhage, infection, or maldevelopment. These authors hypothesized that the cysts begin as diverticulae of the membrane of Lilliequist, allowing the entrance of CSF. The fibroproliferative reaction involving the multiple layers of the membrane of Lilliequist may likely explain the predominant multilocular nature of cysts involving posterior circulation aneurysms noted in our review. Sato and colleagues²⁶ have suggested that arachnoid cysts evolve from exudation or edema induced by vascular endothelial growth factor, a premise similar to theories regarding cyst formation in tumors.²⁹

Krawchenko and Collins¹⁷ have described a ball-valve action between the cyst and subarachnoid space with the possible association of an osmotic gradient as one potential mechanism for arachnoid cyst enlargement.²⁵ Some authors have suggested that a valve action exists only in traumatic cysts; however, endoscopic observations reveal that congenital cysts can also expand by this mechanism.^{6,17} Williams and Guthkelch³⁴ have posited that arachnoid cyst enlargement involves a pulsatile pump mechanism driven by transient pressure gradients established by the responsiveness of the subarachnoid space to changes in venous pressure.²⁵ Santamarta and colleagues²⁵ have used cine phase-contrast MR imaging to support the theory of a pressure gradient between the subarachnoid space and the cyst that would operate like a one-way valve. They have suggested that the gradient is sustained by expansion of the brain during arterial inflow,^{7,21} as has been shown in previous measurements obtained using cine phase-contrast MR imaging.^{5,6,25,27}

Its nodular appearance on CT or MR images means that an aneurysm can be mistaken for a mural nodule when surrounded by a cystic mass. Key to making a correct diagnosis is an understanding of the vascular anatomy in the immediate vicinity of the nodule. The case in the present report clearly highlights its association with the distal M₁ segment. In addition to our case, there have been other documented cases suspected to be a cystic neoplasm.^{1,26,33} Our pathological results coincide with the documented findings of Sato et al.,²⁶ in that they are consistent with

mild reactive gliosis without evidence of hemorrhage. Oversight of an aneurysm when performing resection or biopsy of a cystic mass with a mural nodule can lead to unanticipated aneurysm rupture. The gravity of such an error could explain the paucity of such cases in the literature.

Conclusions

The featured case marks the importance of considering the presence of an aneurysm in the differential diagnosis of a cystic mass with a mural nodule. Given that only 15 cases of aneurysms with associated arachnoid cysts have been described in the literature, much more investigation is necessary. Additional studies in which authors use cine MR imaging and endoscopy will reveal the potential origins of such interesting pathophysiology. Having a precipitant SAH may play a greater role in the development of arachnoid cysts than previously noted. Because aneurysmal SAHs are being diagnosed and treated more frequently, we will continue to gather more information regarding this interesting phenomenon.

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Manuscript submitted November 16, 2006.

Accepted January 8, 2007.

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