

Huge Arachnoid Cyst of the Occipital Cerebral Convexity —Case Report—

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Abstract

A 68-year-old woman presented with a rare huge right occipital cystic lesion manifesting as progressive left hemiplegia. Cranial computed tomography revealed a 4 × 7 cm right occipital lobe thin-walled cyst with midline shift and compression of the ipsilateral ventricle. The patient underwent a single burr hole operation for cystography and delineation of the cyst anatomy, then a separate right parieto-occipital craniotomy with complete cyst evacuation, corticotomy, and ventriculostomy. The presumptive diagnosis was arachnoid cyst. The symptoms had completely resolved by 4 months after surgery with nearly normal neuroimaging appearance after 7 months. Cystography followed by craniotomy and ventriculostomy may be effective for supratentorial arachnoid cysts.

Key words: arachnoid cyst, porencephaly, cystography, corticotomy, ventriculostomy

Introduction

Here we describe a case of an elderly patient presenting with focal signs and a huge cystic lesion with mass effects on cranial computed tomography (CT). The etiology, differential diagnosis, and treatment options are discussed.

Case Report

A 68-year-old woman presented to our Department of Neurosurgery on May 31, 2004 with a 1-month history of progressive left hemiplegia. The patient had a 1-year history of hypertension and liver function abnormalities. There was no history of infection or trauma.

Neurological examination indicated diminished sensation to pin prick on the left distal upper extremity, decreased grip strength of the left hand (20 kg versus 24 kg on the right), and left leg weakness (manual muscle test 5-/5). Cranial CT on admission revealed a low density area (4 × 7 cm) in the right occipital lobe with midline shift involving significant displacement of the right lateral ventri-

cle, but no evidence of ventriculomegaly (Fig. 1A). Cranial T₂-weighted and fluid-attenuated inversion recovery magnetic resonance (MR) imaging on June 4, 2004 demonstrated the arachnoid cyst and surrounding anatomy (Fig. 1B, C). There was no clear evidence of communication with the right ventricle. Cranial coronal T₁-weighted MR imaging demonstrated the presence of several internal membranes within the cyst (Fig. 1D). The presumptive diagnosis was arachnoid cyst.

As a first step we decided to perform cystography to investigate the anatomy of the cyst and the relationship with the right ventricle and surrounding structures. Cystography performed on June 8, 2004 confirmed the absence of communication between the cyst and the right lateral ventricle (Fig. 2A). This procedure also provided important preoperative anatomical information delineating the relationship between the cyst and the right lateral ventricle for the planning of the corticotomy (Fig. 2B).

Right parieto-occipital craniotomy was performed to achieve cyst ablation via cyst aspiration, corticotomy, and permanent communication (ventriculostomy) between the cyst space and the right lateral ventricle. At surgery the dura was opened and immediately a large, thin-walled cyst easily

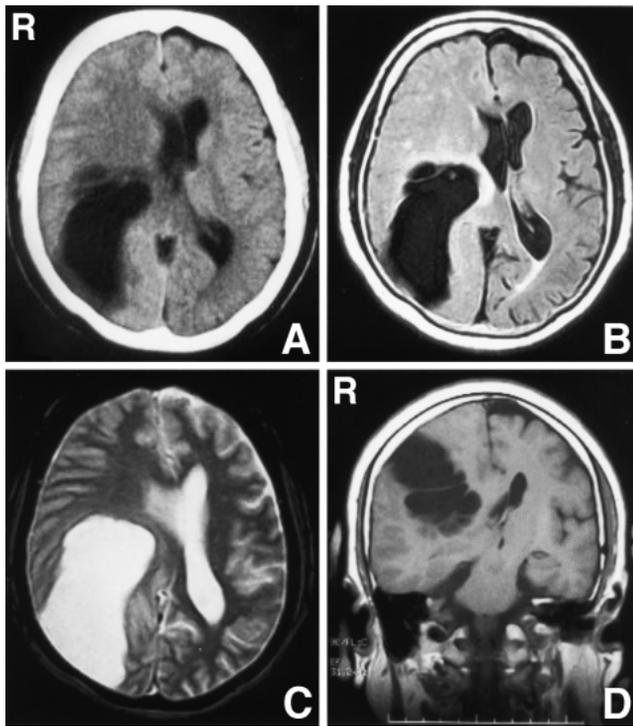


Fig. 1 Cranial axial computed tomography scan on May 31, 2004 (A), and fluid-attenuated inversion recovery (B) and T₂-weighted (C) magnetic resonance (MR) images on June 4, 2004 showing a 4 × 7 cm right parieto-occipital arachnoid cyst and midline shift with displaced right lateral ventricle, but no evidence of ventriculomegaly or communication with the right lateral ventricle. Coronal T₁-weighted MR image (D) showing internal membranes within the arachnoid cyst.

separable from the arachnoid layer, was found that had no obvious communication with the subarachnoid space. There was no obvious macroscopic evidence of chronic inflammation or hemosiderin deposit. The cyst was incised and aspirated. Normal brain cortex was observed adjacent to the medial surface of the cyst. Further decompression of the cyst was performed via corticotomy and ventriculostomy at the trigone and inferior horn of the right lateral ventricle.

Postoperatively there was significant improvement in the patient's symptoms. The patient was discharged on June 27, 2004 and during the first 4 months after surgery, the patient reported normal walking ability and all of her preoperative complaints disappeared. Neuroimaging showed the midline shift had resolved and the cyst gradually disappeared. CT showed some evidence of subgaleal

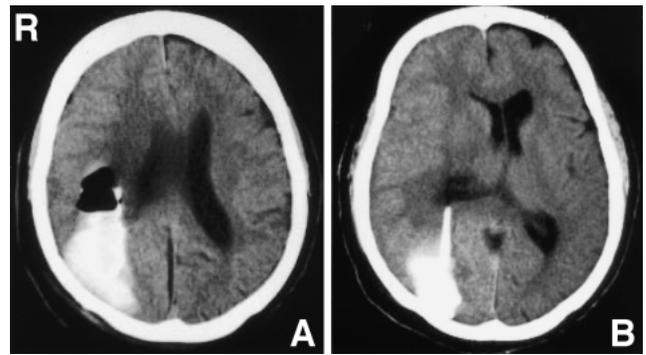


Fig. 2 Cystograms performed on June 8, 2004 showing no evidence of communication between the arachnoid cyst and the right lateral ventricle (A), and the anatomical relationship between the arachnoid cyst and the right lateral ventricle and directional approach for corticotomy and ventriculostomy (B).

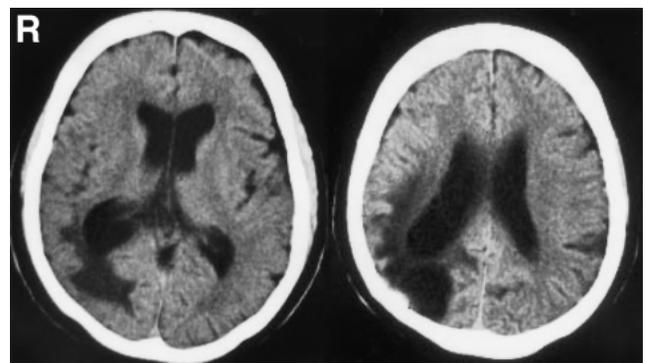


Fig. 3 Cranial computed tomography scans showing nearly normal anatomy at 7 months after the surgery.

fluid accumulation at 2 months postoperatively, which had resolved at 3 months postoperatively, and the anatomy was nearly normal at 7 months postoperatively (Fig. 3).

Discussion

Arachnoid cysts account for approximately 1% of all intracranial lesions^{1,15}) and the overall population incidence of arachnoid cysts is low, with most cases identified in the first 2 decades of life.^{10,12}) To date there have been less than 60 cases of symptomatic arachnoid cysts over 60 years of age reported in the literature.¹⁰) Less than 10% of arachnoid cysts in adults occur in the cerebral convexity, with a slightly higher percentage found in children.^{11,16,17})

Cerebral convexity arachnoid cysts are more common in females, as in our patient. Occipital convexity arachnoid cyst lesions are a rare entity and there is little direct reference in the literature. Symptoms are based on the extent of the lesion in accordance with focal compressive mass effects on adjacent anatomy. Elderly patients typically present with focal neurological signs as seen in our case, but occasionally patients present with symptoms resembling those caused by chronic subdural hematoma and global symptoms of dementia as seen in normal pressure hydrocephalus.^{18,21)}

In contrast to the typical case of a congenital arachnoid cyst, this case probably represents a cyst of secondary origin. Theories for the origin of secondary arachnoid cysts include infection (meningitis), trauma, hemorrhage, chemical irritation, and tumors.^{9,13,20)} Most arachnoid cysts are static but occasionally involute and disappear over time.²³⁾ However, some cysts progressively enlarge resulting in cranial enlargement and localized cranial bulging. The reason for such expansion remains controversial, but a potential cause is the pressure gradient from the movement of cerebrospinal fluid (CSF) into the cyst due to transient increases in CSF pressure via arterial systolic oscillation.⁴⁾ The resulting mass effects of arachnoid cysts on the surrounding neurological structures may cause the development of seizures, psychomotor retardation, and focal neurological signs. In the present case, there was no history of trauma or infection, suggesting that the etiology may be related to one of the congenital mechanisms, or gradual development of the arachnoid cyst as a secondary phenomenon. However, minor trauma unnoticed by the patient could have triggered the enlargement process.

The diagnosis of arachnoid cysts may be confused with several intracranial cysts of various etiologies, including porencephaly, epidermoid cysts, dermoid cysts, chronic subdural hygromas, cerebral abscess, cystic tumors, parasitic cysts, basal midline cysts (midline cysts associated with agenesis of corpus callosum and other causes of hydrocephalus), posterior fossa cysts (dorsal cysts associated with holoprosencephaly, Dandy-Walker cyst, and vein of Galen aneurysms), and cystic craniopharyngiomas.²²⁾ The initial diagnosis of arachnoid cyst can be made using cranial CT which typically shows arachnoid cysts as continuous smooth-bordered, fluid-filled lesions which do not communicate with normal CSF pathways.¹¹⁾ Therefore, CT evidence of absence of ventricular communication is necessary to distinguish arachnoid cysts from false cysts such as Dandy-Walker, hydranencephalic, and porencephalic cysts. However, there is the possibility that

porencephalic cysts can demonstrate communication with the ventricles, although this is less likely. In addition, CT contrast medium showing the absence of a capsule with vascularity can be used to exclude other space-occupying lesions such as primary and secondary malignancies or abscesses.

T₁-weighted MR imaging is the method of choice for the diagnosis of arachnoid cyst. Fluid-attenuated inversion recovery MR imaging can be used to distinguish arachnoid cysts from epidermoid cysts, where a suppressed (low) signal is only found in CSF-containing arachnoid cysts. Dermoid cysts tend to have calcifications and low density on CT and demonstration of fat content on MR imaging.²²⁾ Furthermore, recently arachnoid cysts have been accurately differentiated from other subarachnoid space involving lesions via phase contrast cine MR imaging.²⁴⁾ In this case we did not employ this modality.

Deep invagination of an arachnoid cyst into the cerebral hemisphere may simulate porencephaly to such an extent that it has been termed pseudoporencephaly. However, the inferior aspect of the arachnoid cyst shows a displaced but otherwise normal cerebral cortex, while in porencephaly, the surrounding cortex and white matter are abnormal. In our case intraoperative findings showed that surrounding white matter was observed to be normal supporting the possibility of an arachnoid cyst diagnosis. Chronic subdural hygromas usually are located subdurally rather than in the subarachnoid space and often are bilateral and flat or lentiform in profile. Hygromas compress but do not invaginate into sulci or fissures.²²⁾ And of course pathological sampling of the cyst wall, showing typical delicate single layer of arachnoid cells, is useful in the diagnosis. However, in our study we did not make any histological pathology samples of the cyst.

The optimal treatment is controversial, but surgical treatment is recommended in all symptomatic patients regardless of age at the time of diagnosis.^{4,11)} Treatment for occipital cysts is similar to other convexity lesions and arachnoid cyst in general. Conservative management with serial neuroimaging is recommended for patients with asymptomatic arachnoid cysts. There is no evidence that suggests that surgical treatment prevents associated complications such as hemorrhage.¹⁷⁾ Surgical techniques include craniotomy and cyst wall excision or fenestration of the cyst into the basal cistern or ventricle, stereotactic cyst aspiration, cyst-peritoneal shunting, cyst-ventriculoperitoneal shunting if hydrocephalus is present, cyst-subdural shunting, and endoscopic opening of the cyst into the basal cisterns.^{4,11)}

Craniotomy with fenestration of the cyst in the basal cisterns and cyst-peritoneal shunting are the most frequently used surgical procedures for the treatment of symptomatic intracranial arachnoid cysts. However, many complications have occurred due to rapid decompression (slit ventricle, hemorrhage, hindbrain herniation) or shunt failure.^{2,6,8)} Endoscopic fenestration into the ventricular system of paraventricular or intraventricular arachnoid cysts is reported to be safe.⁷⁾ Finally, stereotactic cyst-ventricular shunting is the optimally least invasive technique, but is limited by the economic access to such technology in most institutions.^{4,5)}

In our patient, all observed neurological symptoms and associated signs disappeared completely after right corticotomy and ventriculostomy. Despite the relatively invasive nature of our treatment, ventricular wall fenestration resulted in a permanent physiological path for cystic CSF production and drainage into the inherent system (similar to other endoscopic or stereotactic shunting procedures). The complete resolution of the symptoms might be explained by the reduction in direct compression of the posterior internal capsule fibers or the functioning cortex. Our case was remarkable in that there were no postoperative complications of hemorrhage from rapid decompression or inadequate cyst retention.

Cystography was very useful in this case for proving that the cyst was indeed a true cyst and not a porencephalic cyst or other false cyst. In addition, there was no evidence of a vascular capsule as found in malignancies or abscesses. Cystography was also valuable for accurate placement of the corticotomy and subsequent successful ventriculostomy. We attempted to resect the entire cyst wall, which is important in preventing cyst recurrence.^{3,19)} However, deciding on the most effective surgical procedure was difficult because of the lack of similar cases in the literature.

Primary symptomatic arachnoid cysts of the cerebral convexity are very rare and can present with focal neurological signs. Cystography followed by craniotomy and ventriculostomy may be a treatment option.

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