Symptomatic Increased Intracranial Pressure Due to Arachnoid Cysts

Sashank Prasad, MD *, Robert A. Avery, DO *, Alejandra de Alba Campomanes, MD, MPH *, Leslie N. Sutton, MD †, and Grant T. Liu, MD *

Intracranial arachnoid cysts are typically benign lesions, but rarely, they may cause signs and symptoms relating to increased intracranial pressure. We report 4 pediatric patients with arachnoid cysts who received successful surgical treatment after failing to respond to conservative medical therapies. After undergoing a shunting procedure, each patient experienced improvement in symptoms, resolution of optic disc swelling, and reduction or elimination of medications necessary to manage the condition. These cases illustrate the potential for arachnoid cysts to outstrip compensatory mechanisms and cause signs and symptoms requiring definitive surgical intervention. © 2011 Elsevier Inc. All rights reserved.


Introduction

Intracranial arachnoid cysts are a common incidental neuroradiologic finding, but the incidence of symptomatic arachnoid cysts is believed to be low [1,2]. Rarely, these lesions may cause increased intracranial pressure or focal neuro-ophtalmic deficits. In this series, we report 4 pediatric patients with intracranial arachnoid cysts with evidence of increased intracranial pressure. Medical therapies did not demonstrate sustained success for these patients. Ultimately, the children benefited from a cerebrospinal fluid diversion procedure, with resolution of the signs and symptoms related to increased intracranial pressure. Although arachnoid cysts typically display a benign course, these cases illustrate their potential to cause morbidity requiring more aggressive therapy. Awareness of this possibility will allow more effective treatment of these complicated cases.

Case Reports

Case characteristics are summarized in Table 1.

Patient 1

A 14-year-old girl presented with 3 weeks of increasing headaches, which were worse in the supine position. She noticed blurry vision in both eyes and pulsatile tinnitus. She was known to have a left middle temporal fossa arachnoid cyst and Chiari 1 malformation, which were diagnosed by magnetic resonance imaging when she developed an increasing head circumference at age 2. She had been treated with a cystoperitoneal shunt at that time.

Examination revealed bilateral severe optic disc swelling with peripapillary hemorrhages and venous distension. Visual acuity was 20/30 with each eye; color vision, pupillary responses, and ocular ductions and alignment were normal. Automated perimetry revealed enlarged blind spots bilaterally. Brain magnetic resonance imaging confirmed a large left middle temporal fossa arachnoid cyst (Fig 1). Continuous intracranial pressure monitoring revealed pressures ranging from 14 to 54 cm H2O.

Acetazolamide 500 mg daily was prescribed. Initially she had substantial improvement in her headaches, but after 1 month, her symptoms recurred. Examination revealed persistent optic disc swelling. Furosemide 20 mg daily was added, but the headaches persisted. The cystoperitoneal shunt was then revised, and within 3 days, the headaches fully resolved. Examination 2 months later revealed resolution of optic disc swelling and hemorrhages.

After 6 months, her headache recurred, and mild optical disc elevation had reappeared. The shunt was explored and found not to be draining; it was repositioned into the lateral ventricle. Her headaches improved again, and the disc swelling remained fully resolved at examination 3 months later.

Patient 2

A 15-year-old boy with migraine headaches sought care for a new, worsening headache for 10 days. The headache was exacerbated in the supine position. He noticed intermittent binocular horizontal diplopia. He was known to have a left middle temporal fossa arachnoid cyst and a Chiari 1 malformation. He had been previously treated with a cystoperitoneal...
shunt. He received topiramate 125 mg daily, and he had tried a brief course of oral prednisolone without success.

Examination revealed normal acuity and pupillary responses. He had limited abduction of the right eye with 12 prism diopter esotropia in primary gaze. Dilated fundus examination revealed no optic disc swelling.

Magnetic resonance imaging demonstrated a left middle temporal fossa arachnoid cyst (Fig 2), and magnetic resonance venography revealed normal venous sinuses. Lumbar puncture revealed an opening pressure of 54 cm H₂O.

The cystoperitoneal shunt was explored and found to be cracked in the chest. Replacement of the tubing restored its function. There was near-complete resolution of the headaches. After 1 week, there was improvement of right eye abduction, with normal alignment in primary gaze, and a 6 prism diopter residual esodeviation in right gaze.

**Patient 3**

A 12-year-old girl manifested severe headaches with episodes of graying out of vision. She had a history of hydrocephalus diagnosed at age 18 months. At that time, a posterior fossa arachnoid cyst was found and a ventriculoperitoneal shunt was placed. She required a shunt revision at age 2 years. At age 6 years, a Chiari 1 malformation and cervical syrinx was diagnosed, and she was treated with suboccipital decompression.

Examination revealed optic disc swelling and normal automated perimetry. After initiating therapy with acetazolamide 250 mg 3 times daily, the visual symptoms resolved and the headaches improved. Six months later, persistent optic disc swelling and one peripapillary hemorrhage were observed. Automated perimetry demonstrated an enlarged blind spot. Visual acuity, ocular ductions, and alignment were normal. Magnetic resonance imaging revealed a stable posterior fossa arachnoid cyst, without compression of the cerebral aqueduct or ventricular enlargement.

She underwent direct cystoventricular shunting of the arachnoid cyst. Her headaches improved dramatically, and after 1 month, the optic disc swelling fully resolved.

**Table 1. Patient clinical characteristics**

<table>
<thead>
<tr>
<th>Age, yr/Sex</th>
<th>Symptoms</th>
<th>Signs</th>
<th>Arachnoid Cyst Location</th>
<th>Other Intracranial Pathology</th>
<th>Intracranial Pressure (method)</th>
<th>Treatments</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>14/F</td>
<td>Positional headache, blurry vision, pulsatile tinnitus</td>
<td>Acuity 20/30, papilledema OU, enlarged blind spot</td>
<td>Left middle temporal fossa</td>
<td>Chiari 1 malformation</td>
<td>14-54 cm H₂O (continuous monitoring)</td>
<td>Acetazolamide, furosemide, cystoperitoneal shunt revision</td>
<td>Improvement of headache, vision and papilledema with acetazolamide, complete resolution of headaches after shunt revision</td>
</tr>
<tr>
<td>15/M</td>
<td>Positional headache, intermittent horizontal diplopia</td>
<td>Esotropia, abduction deficit OD</td>
<td>Left middle temporal fossa</td>
<td>Chiari 1 malformation</td>
<td>54 cm H₂O (LP opening pressure)</td>
<td>Oral prednisone, topiramate, cystoperitoneal shunt revision</td>
<td>Improvement in headache and resolution of ET with only mild residual abduction deficit after shunt revision</td>
</tr>
<tr>
<td>12/F</td>
<td>New onset of headache, visual obscuration</td>
<td>Papilledema, enlarged blind spot</td>
<td>Posterior fossa</td>
<td>Hydrocephalus, Chiari 1 malformation, cervical syrinx</td>
<td>Not measured</td>
<td>Ventriculoperitoneal shunt/revision, Suboccipital decompression, acetazolamide, cystoperitoneal shunt</td>
<td>Improvement of headaches and resolution of papilledema after cystoperitoneal shunt</td>
</tr>
<tr>
<td>12/M</td>
<td>Positional frontal headache, tinnitus, horizontal</td>
<td>Acuity 20/30, papilledema, macular edema, esotropia, abduction deficit OD</td>
<td>Left middle temporal fossa</td>
<td>None</td>
<td>37 cm H₂O (LP opening pressure)</td>
<td>Cystoperitoneal shunt</td>
<td>Complete resolution of headache, esotropia, and papilledema after cystoperitoneal shunt</td>
</tr>
</tbody>
</table>

Abbreviations:
- ET = Esotropia
- LP = Lumbar puncture
- OD = Right eye
- OU = Both eyes
incomitant esotropia (18 prism diopters in primary gaze, 20 prism diopters in right gaze, and 16 prism diopters in left gaze).

Computed tomography of the head revealed a left middle temporal fossa arachnoid cyst. Lumbar puncture revealed an opening pressure of 37 cm H₂O and normal spinal fluid constituents. He received a cystoperitoneal shunt, and within days, the headache and diplopia resolved. Three weeks later, examination revealed decreased optic disc swelling and resolution of hemorrhages, normal ductions, and alignment, and 20/20 acuity in each eye. Four months later, he remained without any symptoms or evidence of optic disc swelling. After 2 months, a computed tomographic scan revealed reduction of the size of the cyst, and after 5 months, he continued to have no symptoms or optic disc elevation.

**Discussion**

Arachnoid cysts are common lesions. Their true prevalence is not known, but they are thought to account for 1% of intracranial mass lesions [2]. Although these lesions are often considered to be benign, this case series highlights their potential to cause signs and symptoms of increased intracranial pressure. The relationship between an arachnoid cyst and increased intracranial pressure is supported by the resolution of headache and neuroophthalmic signs and symptoms after a cerebrospinal fluid diversion procedure. Confirmation of the presumably normalized intracranial pressure at follow-up was not clinically indicated because the deficits of all patients improved.

The reasons why a long-standing arachnoid cyst would become symptomatic are not fully understood. The cysts are thought to occur from leptomeningeal maldevelopment during embryogenesis, and they typically remain stable in size or demonstrate very slow growth [1-4]. With a slowly expanding intracranial mass, compensatory mechanisms (including increased cerebrospinal fluid absorption and collapse of cerebral veins and sinuses) typically permit homeostatic maintenance of the intracranial pressure [5-7]. Nevertheless, there are limits to these compensatory mechanisms, and increase of intracranial pressure typically depends more on the rate of expansion of a mass than on its absolute size. Therefore, an arachnoid cyst that has become symptomatic during childhood may be growing at a rate that surpasses a critical threshold for maintaining the intracranial pressure.

Symptomatic arachnoid cysts appear to be more common in children than adults [8-10]. In fact, 3 of our 4 adolescent patients had received initial surgical intervention for the cyst before age 2. In these 3 patients, the cyst may have demonstrated very early rapid growth, exceeding compensatory mechanisms and causing symptoms at an early age. Although it is not clear why the incidence of symptomatic arachnoid cysts is higher in children, it may be that relative expansion of the cyst during a period of normal growth and development leads to a disproportionately higher risk of increased mass effect and increased intracranial pressure. Later in life, this risk of significant relative growth of the cyst may be greatly reduced, leading to a fairly benign natural history of these lesions in adulthood.

It is noteworthy that 3 of the 4 patients presented here also had a coexisting Chiari 1 malformation. This anatomical variant may have altered cerebrospinal fluid pressure.
homeostatic mechanisms and contributed to the onset of symptomatic increased intracranial pressure. Further studies are necessary to determine whether patients with both an arachnoid cyst and Chiari 1 malformation are in fact at higher risk of developing pathologic elevations in intracranial pressure.

In patients with headaches who are found to have an arachnoid cyst, it is critical to ascertain the presence of clinical signs or symptoms of increased intracranial pressure. In these patients, medical management aiming to lower the intracranial pressure should be initially attempted. Occasionally, however, attempts to taper these medications are unsuccessful, and it becomes clear that effective management is not possible through conservative measures alone. Ultimately, surgical treatment, either by fenestration or cerebrospinal fluid shunting, may lead to durable resolution of the neuro-ophthalmic deficits [11]. The cases presented here support the conclusion that, on rare occasions, an arachnoid cyst may cause increased intracranial pressure with related signs and symptoms that respond to surgical therapy [11].

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References