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Adult-Onset Shunt Dependency Syndrome: A Case Report

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Background: Arachnoid cysts are benign intracranial collections formed by the accumulation of cerebrospinal fluid between the arachnoid membranes. Cerebral shunt implantation is usually reserved for selected symptomatic cases. Shunt dependency syndrome is a relatively rare and underdiagnosed complication of cerebral shunt implantation that has been described almost exclusively in children.

Case Report: A 50-year-old man with a history of cystoperitoneal shunting for a temporal lobe arachnoid cyst at age 34 presented with recurrent episodes of severe headache, nausea, vomiting, and unilateral amaurosis over the preceding 10 years. Brain imaging showed normal-sized ventricles, with no changes in the size of the arachnoid cyst or the position of the cystoperitoneal shunt. Shunt dependency syndrome was suspected, and replacement of the cystoperitoneal shunt system eventually led to resolution of symptoms.

Conclusions: This case highlights the importance of recognizing shunt dependency syndrome, even in adult patients, to prevent long-term neurological complications.

Key words: Headache; slit ventricle syndrome; ventriculoperitoneal shunt; intracranial hypertension; arachnoid cysts

BACKGROUND

Arachnoid cysts (ACs) are benign intracranial collections formed by the accumulation of cerebrospinal fluid (CSF) between the external and internal arachnoid membranes (1,2). Most ACs are congenital; that is, they result from altered development of the meninges during intrauterine life. CSF is believed to cause dissection of the arachnoid layers during the fourth month of fetal life, resulting in intrameningeal accumulation and obliteration of the normal subarachnoid space (1).

Sixty to ninety percent of ACs are detected in patients younger than 16 years. They are usually asymptomatic, although, depending on their size, location, or growth, they can cause intracranial hypertension and hydrocephalus or, much less frequently, a hypertensive subdural hygroma or a subdural or intracystic hematoma as a complication of traumatic or spontaneous rupture (2). Treatment is reserved for symptomatic ACs, which are usually of considerable size. Surgical options include endoscopic fenestration or implantation of a cerebral shunt (3). The latter is a relatively quick and simple procedure; however, it is not free of complications. Shunt dependency is a relatively rare and underdiagnosed complication that has been described almost exclusively in children (4).

Here, we present a case of adult-onset shunt dependency syndrome and discuss its implications.

CASE PRESENTATION

A 50-year-old man with a history of cystoperitoneal shunting for a temporal lobe arachnoid cyst at age 34 presented to the emergency department with severe headache, nausea, vomiting, and unilateral amaurosis. He had experienced recurrent episodes over the preceding 10 years and had been treated with topiramate and amitriptyline for a presumptive diagnosis of retinal migraine, without improvement.

Neurological examination revealed complete visual

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loss in the right eye, with a dilated pupil and diminished light reflex. Fundoscopy showed right optic nerve atrophy. Refilling of the valve pumping mechanism was adequate. Computed tomography (CT) (Fig. 1) and brain magnetic resonance imaging (MRI) showed normal-sized ventricles, with no changes in the size of the right temporal arachnoid cyst or the position of the cystoperitoneal shunt. The opening pressure on lumbar puncture was 170 mm H_2O, and CSF findings were normal. The headache resolved after the lumbar puncture, and vision in the right eye improved significantly. Shunt dependency syndrome was suspected, and a neurosurgical consultation was requested. The neurosurgical team decided to continue outpatient evaluation, and the patient was discharged.

Over the following months, the patient was readmitted because of recurrent episodes of headache and visual loss of increasing severity. A repeat lumbar puncture revealed an elevated opening pressure (260 mm H_2O). Surgical revision was performed, and an obstruction at the distal end of the shunt was identified, requiring replacement of the cystoperitoneal shunt system. The patient improved significantly and remained asymptomatic 2 years later.

DISCUSSION

The term "shunt dependency syndrome" describes a group of headache disorders characterized by signs and symptoms of increased or decreased intracranial pressure in patients with CSF shunt systems (5). The term derives from the concept that, once a shunt is placed, normal mechanisms of CSF absorption and drainage are impaired, causing CSF drainage to depend almost exclusively on the shunt (4). This term is analogous to "slit ventricle syndrome," which refers to the absence of ventricular dilation despite significant changes in intracranial pressure.

Five headache entities are currently included under shunt dependency syndrome: (1) extreme reduction in intracranial pressure due to CSF overdrainage through the shunt system; (2) transient intracranial hypertension secondary to intermittent proximal catheter obstruction associated with collapse of the ventricular wall or cyst; (3) intracranial hypertension due to shunt

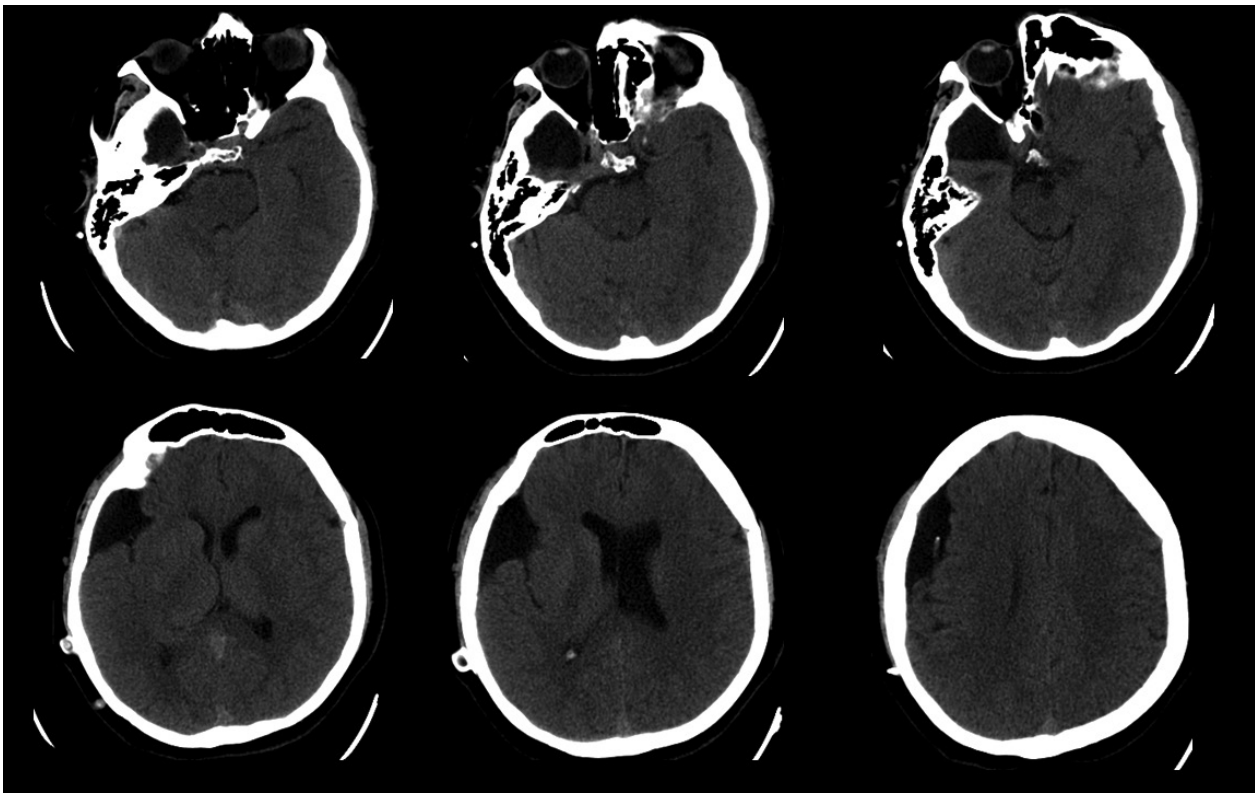


Fig. 1. Brain computed tomography shows a right temporal arachnoid cyst with the cystoperitoneal shunt and normal-sized ventricles.

failure without ventricular dilation; (4) intracranial hypertension associated with a functioning shunt due to encephalocranial disproportion; and (5) headache of primary origin with a normally functioning shunt (6,7).

These headache syndromes have been reported almost exclusively in children. However, our case illustrates that shunt dependency syndrome can also affect adults, in whom diagnosis is often delayed, possibly because of transient clinical manifestations, the absence of hydrocephalus on diagnostic imaging, and limited awareness among medical personnel.

Our patient presented with classic signs of intracranial hypertension, including headache that worsened in the supine position, nausea, vomiting, and visual loss (8). Furthermore, these symptoms improved with CSF drainage through lumbar puncture, and opening pressure was normal or elevated. Neither brain CT nor MRI showed dilation of the ventricular system, changes in the arachnoid cyst or collapse near the proximal end of the catheter. Therefore, we classified the condition within the third category: shunt failure without signs of hydrocephalus (pseudotumor cerebri syndrome).

The mechanism by which the arachnoid cyst or ventricular system fails to expand despite increased intracranial pressure has not been clearly established. It has been postulated that progressive reductions in cerebral elasticity and compliance due to fibrosis of the surrounding tissue may be responsible (9).

Surgical exploration revealed a distal catheter obstruction, and symptoms resolved after shunt replacement. We believe that the recurrent headache episodes were caused by shunt malfunction due to intermittent distal catheter obstruction in the peritoneum. During each headache episode, clinical manifestations developed progressively as intracranial pressure increased until it overcame the obstruction in the shunt system, allowing CSF outflow and, consequently, symptom remission.

Notably, the headaches were associated with transient unilateral amaurosis and mydriasis, which eventually led to a permanent decrease in visual acuity due to optic nerve atrophy. Although the exact mechanism of these findings is difficult to explain, we believe that it may be related to the location of the arachnoid cyst in the middle cranial fossa. This location could cause nearby structures, including the right optic and oculomotor nerves, to be predominantly affected by intermittent intracranial hypertension, potentially leading to secondary optic nerve atrophy (10).

CONCLUSION

In conclusion, careful evaluation of patients with arachnoid cysts is important to avoid unnecessary cystoperitoneal shunting because shunt dependency syndrome is a rare but potentially serious complication. As our case illustrates, this condition is not exclusive to children. Therefore, physicians should consider this possibility in adults with cystoperitoneal shunts who present with clinical signs of elevated intracranial pressure, even if neuroimaging does not reveal morphological changes in ventricular or arachnoid cyst volume (11). These patients should undergo prompt neurosurgical evaluation to determine whether surgical exploration is warranted and to prevent permanent neurological sequelae (12).

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