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Shunt Dependency Syndrome Combined with Sinus Stenosis after Cyst-peritoneal Shunting of Arachnoid Cyst: A 20-year Rare Complication

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Abstract

Shunt dependence syndrome is a serious long-term complication characterized by symptoms and signs of increased intracranial pressure with normal-sized lateral ventricles after several years of arachnoid cyst-peritoneal shunting. It is easy to misdiagnose and overlook when combined with sinus stenosis, thus delaying treatment. Here, we present a 35-year-old man with an unexplained headache and binocular horizontal diplopia with high intracranial pressure. Based on stenosis of the right transverse sinus and absent filling of the left transverse sinus found by cerebral angiography, we first suspected cerebral venous sinus thrombosis (CVST). However, the treatment of anticoagulation and enhanced dehydration did not work. Twenty years ago, he had a cyst-peritoneal (CP) shunt performed for an asymptomatic arachnoid cyst. Finally, neurosurgeons ruled out CVST by direct retrograde cerebral venography and diagnosed shunt dependence syndrome. All his symptoms resolved well after ventriculoperitoneal (VP) shunt treatment. This case reminds pediatrics and neurology colleagues to consider shunt dependence syndrome first when someone has a history of CP shunting of arachnoid cysts presented with a severe headache, even if imaging shows a thin or occlusive venous sinus. Once high intracranial pressure secondary to shunt dependence is diagnosed, timely reconstruction of the cerebrospinal fluid (CSF) pathway is recommended rather than osmotic therapies because cerebral herniation formation will be life-threatening if ongoing increased intracranial pressure is not relieved.

Keywords: shunt dependency syndrome, arachnoid cyst, cyst-peritoneal shunt, acute intracranial hypertension, ventriculoperitoneal shunt

Introduction

Arachnoid cyst is a congenital malformation, and its global prevalence is 0.3%-2.6%.[1)] It is more common in children, most of whom are asymptomatic.[2)] These cysts contain colorless and transparent fluid similar to cerebrospinal fluid (CSF) and communicate with the subarachnoid space.[3,4)] Intracranial arachnoid cysts are located in the lateral fissure, mostly reaching 1/2-2/3. Many cysts are discovered incidentally after head imaging.[5)]

Various surgical methods for arachnoid cysts have been developed over nearly 100 years. Cyst-peritoneal (CP) shunt surgery is developed based on traditional craniotomy cystectomy,[6)] which was widely used in China around 2000 and became popular as the first choice for surgical treatment of arachnoid cyst due to its simplicity and visible effect to reduce the cyst rapidly.[7-9)] However, more drawbacks of shunt surgery emerged over time in addition to the inherent defects of shunt surgery, that is, foreign body implantation in vivo for life.[10)] Shunt dependency syndrome relevant to CP has gradually emerged and increased, troubling neurologists. Whenever a shunt fails, even minimal CSF volume fluctuation can contribute to increased intracranial pressure for decreased cerebral elasticity and com-

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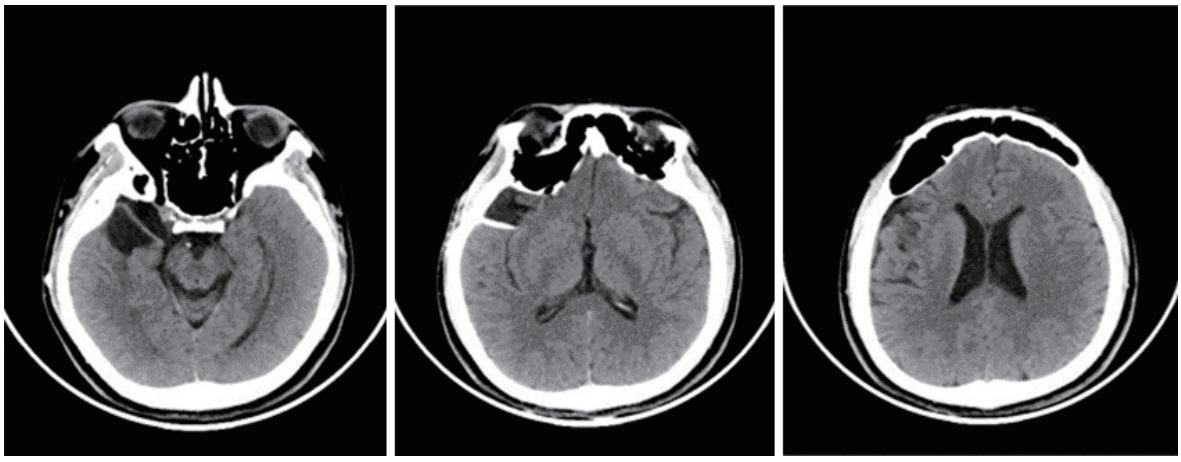

Fig. 1 CT demonstrated a collapsed intracranial arachnoid cyst, simple cystic lesions, and isodense to CSF. The intra-cystic catheter of the CP shunt was in it. The ventricle was normal-sized.

pliance. This is a case report of a patient presenting with a headache and suspected CP shunt malfunction/shunt dependency or cerebral venous sinus stenosis as the cause of the headache, which was difficult to determine.

Case Report

A 35-year-old man presented to our hospital with both intermittent and oppressive pain throughout the head lasting for 5 days accompanied by backache and excessive perspiration, episodes of which lasted from a few minutes to hours, and subsequently binocular horizontal diplopia for 3 days, as well as an unsteady gait. Despite smoking and drinking, he had no history of hypertension, diabetes mellitus, coronary heart disease, myocarditis, arrhythmia, arterial aneurysm, trauma, or cluster headaches. He did have an asymptomatic arachnoid cyst as an incidental finding after imaging of the head twenty years earlier. He underwent placement of a right cyst-peritoneal (CP) shunt, but he failed to follow up regularly.

Except for neck toughness, restricted abduction of the left eye, and binocular horizontal diplopia, the remaining physical examinations were normal, including vital signs, pupil examination, limb muscle strength testing, and reflexes.

On admission, brain computed tomography (CT) revealed a collapse of the right temporal lobe arachnoid cyst after CP shunt catheter placement and showed normal ventricle size (Fig. 1). However, the patient underwent a lumbar puncture after admission. Clear and transparent cerebrospinal fluid (CSF) spurted out, with a pressure measurement exceeding 400 mmH_2O. The routine and biochemical examinations of CSF were normal. The fundus examination did not reveal significant papilledema (Fig. 2). A life-threatening report of critical value from a standard 12-lead ECG soon after the lumbar puncture reported junction escape rhythm and sinus arrest (Fig. 3), which may be due to acute high intracranial pressure via "Cushing response" simultaneously with high blood pressure and slow breathing rate.

Given the situation, atropine was pumped slowly to maintain the patient's heart rate. He received a combination of mannitol (25 g every 6 hours), glycerol fructose (250 mL every 12 hours), and furosemide (20 mg every 12 hours) to reduce the intracranial pressure, yet with little effect. We performed a cerebral angiography and identified a stenosis of the right transverse sinus and absent filling of the left transverse sinus (Fig. 4), which seemed the most likely cause of his increased intracranial pressure. Anticoagulation therapy (low molecular weight heparin 5000 IU/day by subcutaneous injections) was performed because of concerns about stenosis caused by venous thrombosis for its probable poor prognosis without timely treatment. However, there were no obvious abnormalities in the testing of the coagulation cascade, thrombophilia screening, D-dimer, routine items of rheumatic disease, autoantibody, and erythrocyte sedimentation rate.

Besides the venous sinus stenosis, shunt malfunction/shunt dependency could not be ruled out, given the history of CP shunting and the poor effectiveness of conservative treatment. Thus, the patient was transferred to the neurosurgical department. A diverter valve inspection indicated that the elasticity, pressure, and rebound were acceptable. Abdominal CT showed a drainage tube in the abdominal cavity with an unwrapped peritoneal catheter tip. The patient underwent two lumbar punctures in the following days, with CSF pressure showing 300 mmH_2O and 320 mmH_2O, respectively. The examination performed during the two lumbar punctures with direct retrograde cerebral venography and manometry evaluated whether he was suitable for venous stent placement. The findings demonstrated moderate venous sinus stenoses and a pressure difference of 2 mmHg in venous pressure between the distal and proximal parts of stenosis. Therefore, stent placement

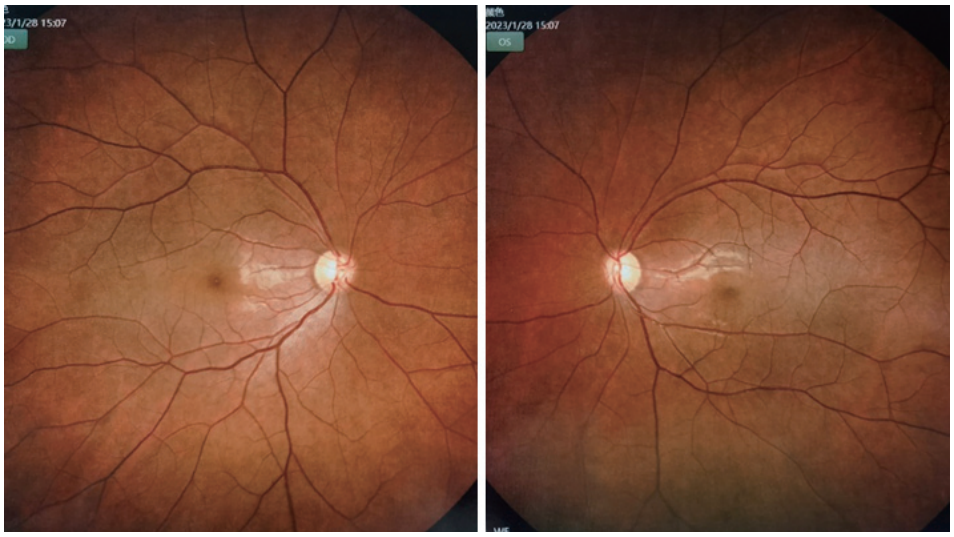

Fig. 2 Fundus examination did not reveal significant papilledema.

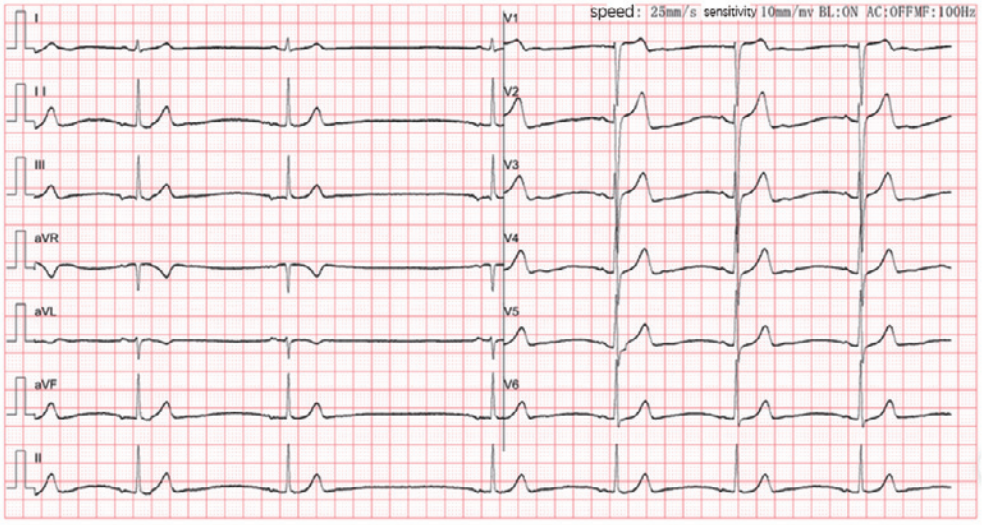

Fig. 3 ECG reported junction escape rhythm and sinus arrest.

was not performed. At this time, shunt dependence syndrome was highly suspected. Since subsequent cardiac ultrasound, ambulatory electrocardiogram, and other comprehensive evaluations showed no contraindications for surgery, the patient underwent a ventriculoperitoneal (VP) shunt with a catheter tip in the left lateral ventricle (Supplementary Figure 1). Ultimately, the patient achieved good results compared with osmotic and anticoagulation therapies; that is, the patient's headache and diplopia were visibly relieved at discharge, supporting the final diagnosis of shunt dependency syndrome. Furthermore, the patient did not have any sequelae during the 8-month follow up.

Discussion

Shunt dependency syndrome is a long-term complication after CP shunt surgery.[11] The development of shunt dependence has no significant correlation with age at the time of the first shunt operation, the cyst location or size, or the number of previous CSF shunt operations.[6,12] Shunt dependency syndrome mainly presents with symptoms and signs such as unexplained severe headache, tinnitus, decreased vision, and papilledema gradually after several years of shunt surgery, often combined with ventricular fissure reduction, reactive skull hyperplasia, decreased cranial volume, cerebellar tonsillar hernia, etc.[12,13] The characteristic was that although there were no obvious signs of intracranial hypertension on neuroimaging, intracranial pres-

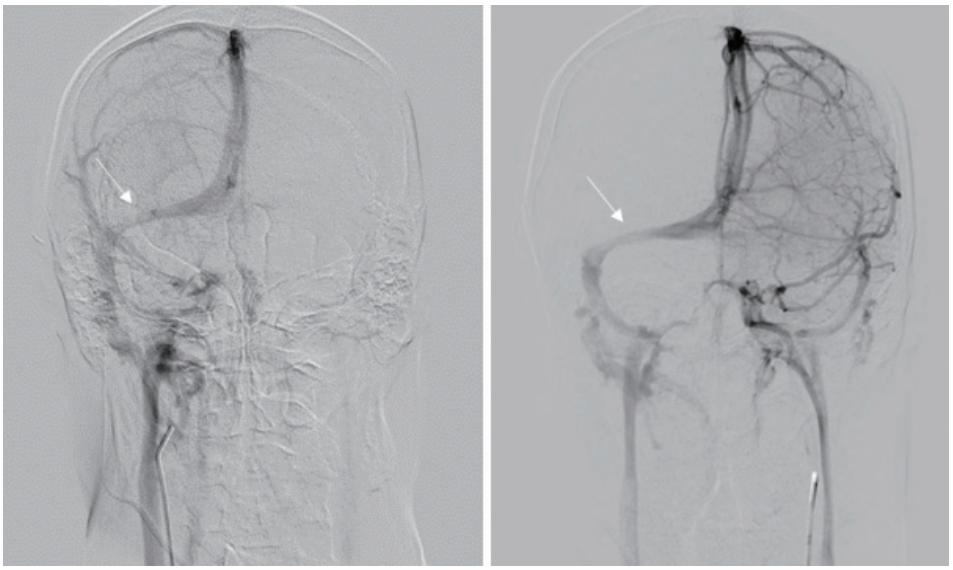

Fig. 4 Cerebral angiography revealed right transverse sinus stenosis and absent filling of the left transverse sinus.

sure (ICP) monitoring shows obvious hypertension in all patients,[9] which was also different from shunt malfunction.[8,14]

The etiology and pathogenesis of shunt dependence syndrome remain unclear.[9] Arachnoid cyst is involved in CSF circulation, considering it communicates with the subarachnoid space, as verified by CT after an intrathecal injection of metrizamide.[3,4] Ahn et al.[15] hold that implementing the shunt affects the original circulation pathway of CSF, and the excessive drainage of CSF destroys the original mechanism of ICP regulation, resulting in shunt dependency syndrome. We suggest that the probable mechanism of shunt dependence syndrome comes as follows: Step 1. The arachnoid cyst becomes smaller as the CSF drains in large quantities after shunt tube implantation, sometimes along with the ventricles becoming smaller, and the cyst's collapse leads to obstruction of the shunt tube head. Step 2. As CSF drainage is obstructed, CSF accumulation again causes the arachnoid cyst and ventricles to expand, thereby relieving the obstruction and restoring the shunt function. The patient's ICP regulation mechanism still works at this time, only with intermittently increased ICP and intermittent symptoms. Step 3. However, the above process occurs repeatedly until the CSF circulatory mechanism is destroyed, resulting in complete reliance on shunt drainage of cerebrospinal fluid based on ependymal fibrosis, paraventricular edema and gliosis, decreased ventricle and brain compliance.[16] Step 4. Once the shunt tube, whichever part including the shunt head, pump, or peritoneal end, is obstructed, ICP continues to increase sharply, and there is no spontaneous remission between the patient's symptoms because remaining regulation for ICP cannot compensate. In our case, it is twenty years from step 1 to step 4, turning a non-shunt-dependent patient into a dependent one. In Kim's work, eight patients (7 boys and 1 girl) were diagnosed as shunt dependent following CP shunting, and the time range from 17 to 80 months.[9]

The possibility of shunt dependence syndrome should be considered when patients with arachnoid CP shunt surgery have the following conditions: (1) clinical symptoms related to high ICP (headache, nausea, vomiting, vision loss, etc.) or other unexplained symptoms such as epilepsy, etc.; (2) significantly high intracranial pressure monitored by lumbar puncture or insertion of an ICP monitor; (3) slow filling of the shunt pump; (4) significantly reduced arachnoid cyst along with normal-sized or small ventricles, with or without paraventricular edema. Shunt dependence syndrome should be highly suspected as long as 3 of the above 4 diagnostic criteria are met.[7]

Patients with shunt-dependent syndrome are often misdiagnosed as having a venous sinus thrombosis because of impaired venous sinus drainage under high ICP. Cerebral venous sinus thrombosis (CVST) is mainly secondary to intracranial infection, head trauma, and brain tumors, which also have symptoms and signs of high cranial pressure.[17] CTV or MRV imaging, fibrin degradation products, and other examinations can further identify it. However, all those examinations help identify CVST but are unhelpful in confirming the diagnosis and distinguishing it from shunt dependence syndrome. It is necessary to remind pediatrics and neurology colleagues to consider shunt dependence syndrome first in patients with a history of CP shunting of arachnoid cysts who present with severe headache and with imaging showing thin or occlusive venous sinus. Keep in mind not to merely follow the treatment of venous sinus thrombosis because cerebral herniation formation will be life-threatening if ongoing increased ICP is not relieved.

Distinguishing features also include the fact that children with shunt dependence syndrome can temporarily relieve headaches by releasing small amounts of CSF after lumbar puncture, but they have little relief with mannitol infusion. Since mannitol decreases intracerebral pressure mainly by drawing fluid into blood vessels from brain parenchyma through osmotic pressure difference, it is more effective for cerebral parenchymal edema. People with shunt dependence syndrome are often insensitive to mannitol, for their increased ICP is more likely due to reduced cranial volume but not cerebral edema. In our case, we promptly requested a neurosurgical consultation when anticoagulation and enhanced dehydration did not work, and finally, the patient underwent an effective VP shunt.

More evidence suggested that a programmable shunt could avoid complications from overdraining. The lateral ventriculoperitoneal shunt is the preferred treatment to relieve the high ICP instead of mannitol infusion in shunt-dependent syndrome.[13)]

One regret of this case is that a shuntgraphy should have been performed before cerebral angiography for an earlier diagnosis of shunt problems.[18)] It is possible to identify the exact level of malfunction (ventricular catheter, valve device, distal catheter), thus allowing for the tailoring of surgical VP shunt revision and avoiding unnecessary complete system replacement.[19)]

Conclusion

In summary, shunt dependency should be suspected if a patient with a CP shunt complains of intracranial hypertension, such as headache, vomiting, or abducens nerve palsy, even if there is no change in cyst size. When imaging shows the venous sinus is thin or occluded, we should think more to prevent misdiagnosis for VSS or CVST. Timely surgery, especially the lateral ventriculoperitoneal shunt, is preferred to relieve the high ICP instead of mannitol infusion or anticoagulation drugs in shunt-dependent syndrome.

Supplementary Material

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Ethics Declarations

The authors confirm that written consent for submitting and publishing this case report, including images and associated text, was obtained from the patient per COPE guidelines. The participants provided informed consent. Review and/or approval by an ethics committee was not required for this study.

Data Availability Statement

The original contributions presented in the study are included in the article; further inquiries can be directed to the corresponding author.

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CRediT Authorship Contribution Statement

Mengying Chen: Writing - original draft, Validation, Investigation, Formal analysis, Conceptualization. Mengyuan Zhang: Writing - original draft, Validation, Methodology, Formal analysis. Hong Sun, Huiyang Qu, Yuxuan Cheng, Jiaxin Fan, Qingling Yao, Xiaodong Zhang, Shuyin Ma: Writing - review & editing, Methodology, Investigation, Formal analysis. Shuqin Zhan: Writing - review & editing, Project administration, Data curation, Funding acquisition.

Conflicts of Interest Disclosure

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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