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CASE REPORT

Shunt dependency syndrome and acquired Chiari malformation secondary to cerebrospinal fluid diversion procedures: a 9-year longitudinal observation

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Abstract

Background Shunt dependency syndrome is a rare long-term complication of cystoperitoneal (CP) shunting for intracranial arachnoid cysts, which is characterized by acute intracranial hypertension with normal-sized or small ventricles. Additionally, acquired Chiari type I malformations (ACIM) could be infrequently secondary to extrathecal shunt drainage of cerebrospinal fluid.

Case report We described a 12-year-old boy who developed shunt dependency syndrome following a CP shunting for treating a temporal arachnoid cyst. To manage this rare complication, we placed a lumboperitoneal (LP) shunt. During the follow-up period, shunt-induced ACIM and concomitant syringomyelia were noted.

Conclusion Shunt dependency syndrome is a rare complication secondary to CP shunting in the treatment of temporal arachnoid cysts, and LP shunting is an effective option to relieve the intracranial hypertension. However, the clinicians should be alert to the ACIM as a rare late complication of cerebrospinal fluid diversion procedures, and the potential protecting effect of the programmable valve should be emphasized.

Keywords Shunt dependency syndrome · Chiari malformation · Tonsillar herniation · Arachnoid cyst · Cerebrospinal fluid shunt

Introduction

Shunt dependency syndrome is an extremely rare long-term complication of cystoperitoneal (CP) shunting for intracranial arachnoid cysts. This infrequent condition is characterized by acute intracranial hypertension during shunt malfunction; nevertheless, both the ventricles and the cysts are normal or small in size [8].

Chiari malformation, also known as tonsillar herniation, is generally considered a congenital neurological anomaly, in which the lower parts of the brain or cerebellum protrude through the magnum foramen into the spinal column. However, acquired forms of CM have been recognized in recent years. As reported, the development of "acquired" Chiari type I malformations (ACIM) could infrequently secondary to extrathecal shunt drainage of cerebrospinal fluid (CSF) in cases of hydrocephalus or pseudotumor syndrome [4–6, 11].

Shunt dependency syndrome occurring after CP shunting, as well as ACIM following CSF diversion procedures, is rare, and only a few literatures have been identified. Herein, we described the dynamic development of shunt dependency syndrome following placement of a CP shunt in the treatment of a temporal arachnoid cyst. Additionally, this rare complication required a lumboperitoneal (LP) shunt. During follow-up, shunt-induced ACIM and concomitant syringomyelia were noted. The 9-year longitudinal observation data were presented, and the relevant mechanisms were discussed.

Case report

A 12-year-old boy first presented to our department at the age of 3 years with headache and restlessness. Brain magnetic resonance (MR) imaging revealed an arachnoid cyst in the right temporal lobe, and there was no evidence of low level

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of the cerebellar tonsils (Fig. 1). A CP shunt was inserted, and the postoperative courses were uneventful.

Five years later, he presented to our department with a 2-year history of intermittent headache in the right frontal area, dizziness, nausea, and vomiting. Over the preceding week, the condition aggravated and he told a new-onset tinnitus. Computed tomography scan of the head demonstrated the arachnoid cyst was collapsed and the ventricles were narrowed (Fig. 2). A measurement during a lumbar puncture revealed a pressure above 350 mm H_2O. A diagnosis of increased intracranial pressure and shunt dependency syndrome was made. Accordingly, a LP cerebrospinal fluid diversion procedure was performed. The symptoms relieved postoperatively. The MR imaging 4 months thereafter showed small residual of the arachnoid cyst, mild cerebellar tonsillar herniation, and syringomyelia at C2 level (Fig. 3).

After a 27-month uneventful interval, he returned to our department with intermittent headache and dizziness. He also told that the pain alleviated in the supine position. MR imaging revealed an aggravated cerebellar tonsillar herniation; nevertheless, the arachnoid cyst and syringomyelia remained unchanged (Fig. 4a–c). Abdominal plain X-ray revealed a smooth continuity of the LP shunt. A measurement during a lumbar puncture revealed a pressure of 35–40 mm H_2O. A diagnosis of intracranial hypotension syndrome due to overdrainage was made. He underwent placement of a LP shunt with a programmable pressure valve, and the pressure was started at 150 mm H_2O. The patient was uneventfully discharged. Given the inapparent clinical manifestations, the shunt-induced Chiari malformation was not surgically treated. On the latest MR images 18 months after discharge, we noted there was no remarkable change of the arachnoid cyst, the cerebellar tonsillar herniation, and the syringomyelia (Fig. 4d–f). During a 25-month follow-up period following the last operation, he was asymptomatic.

Discussion

Since 1990, scholars noted that the children could develop acute intracranial hypertension after CP shunting for temporal arachnoid cysts as a result of shunt malfunction; nevertheless,

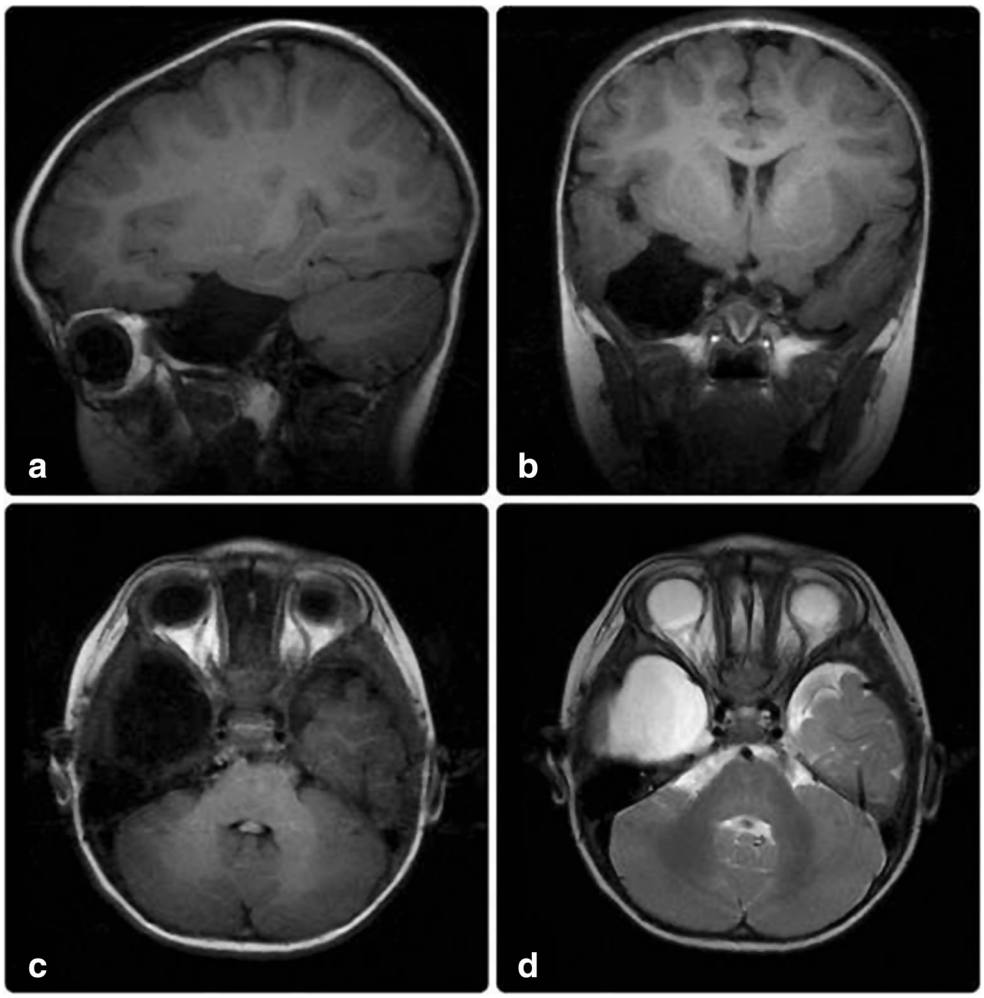

Fig. 1 Sagittal (**a**) and coronal (**b**) T1-weighted MR images and axial T1-weighted (**c**) and T2-weighted (**d**) images demonstrate an arachnoid cyst in the right temporal lobe

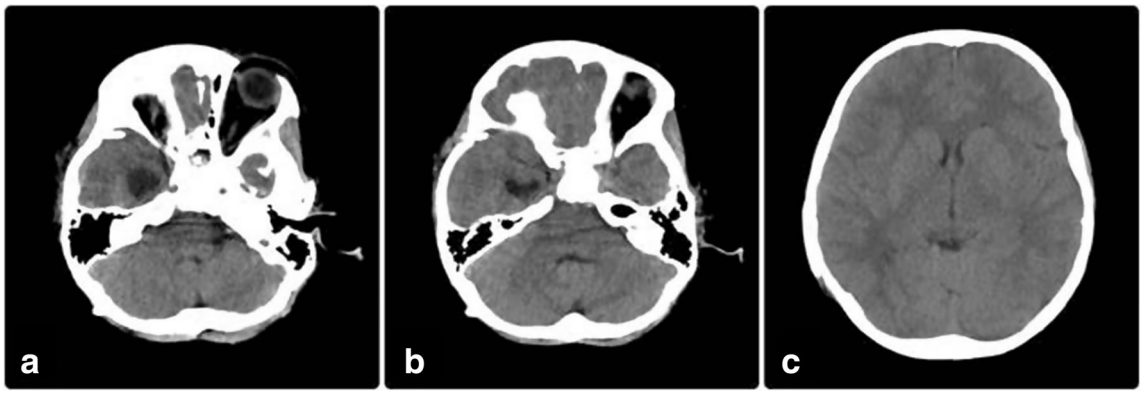

Fig. 2 Computed tomography scan of the head demonstrate a collapsed arachnoid cyst (**a** and **b**) and the ventricles were small (**c**)

both the ventricles and the cysts were normal or even small in size [2, 9]. They consider this condition similar to slit ventricle syndrome (SVS) which is a common long-term complication of a functioning cerebrospinal fluid shunt due to overdrainage [2]. In recent years, some scholars suggested renaming this condition to "shunt dependency syndrome" [8].

The etiology and pathogenesis of shunt dependency syndrome has not yet fully understood. There are two mainstream

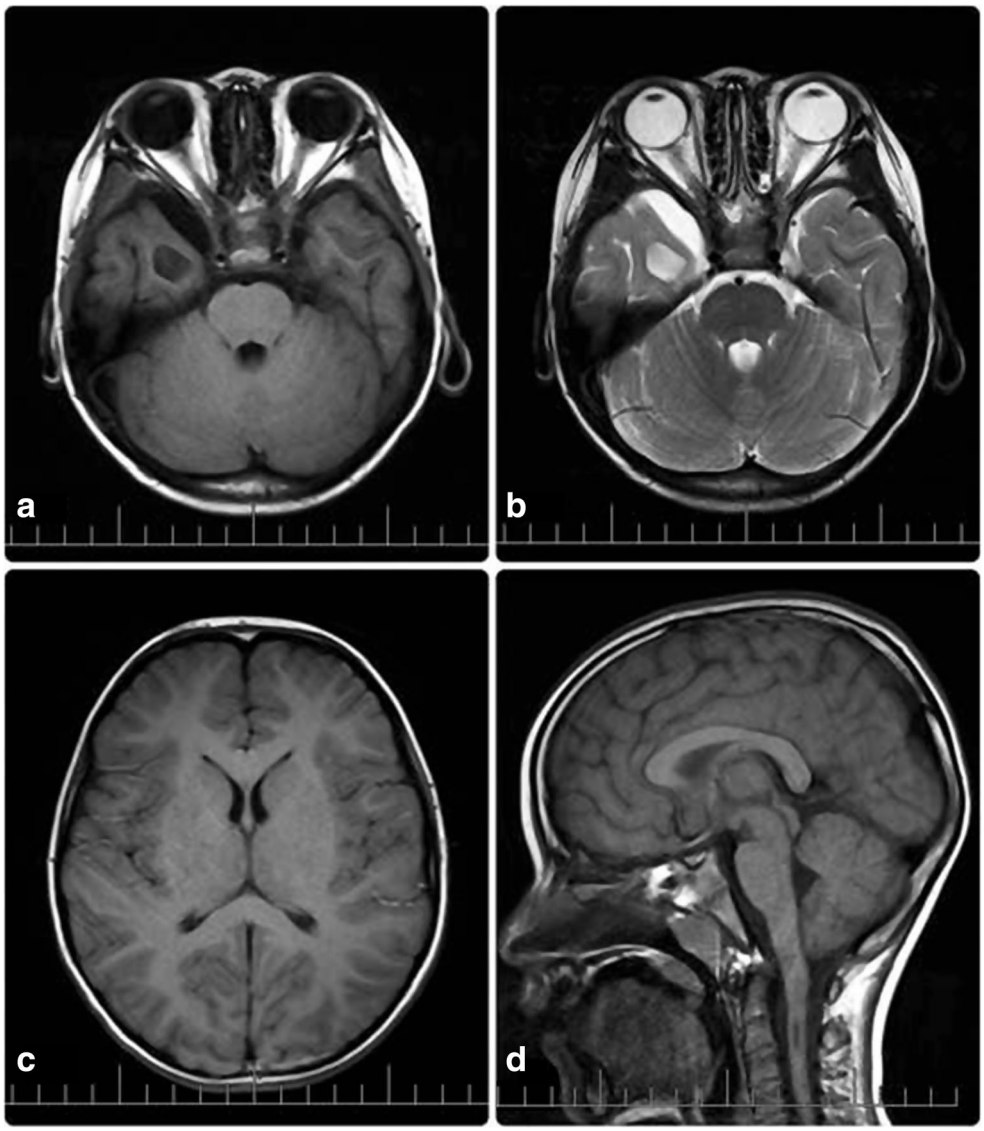

Fig. 3 Axial T1-weighted (**a**) and T2-weighted (**b**) MR images reveal small residual of the arachnoid cyst. Coronal T1-weighted images (C) show a narrowed ventricle. Sagittal T1-weighted images (**d**) demonstrate mild cerebellar tonsillar herniation and syringomyelia at C2 level

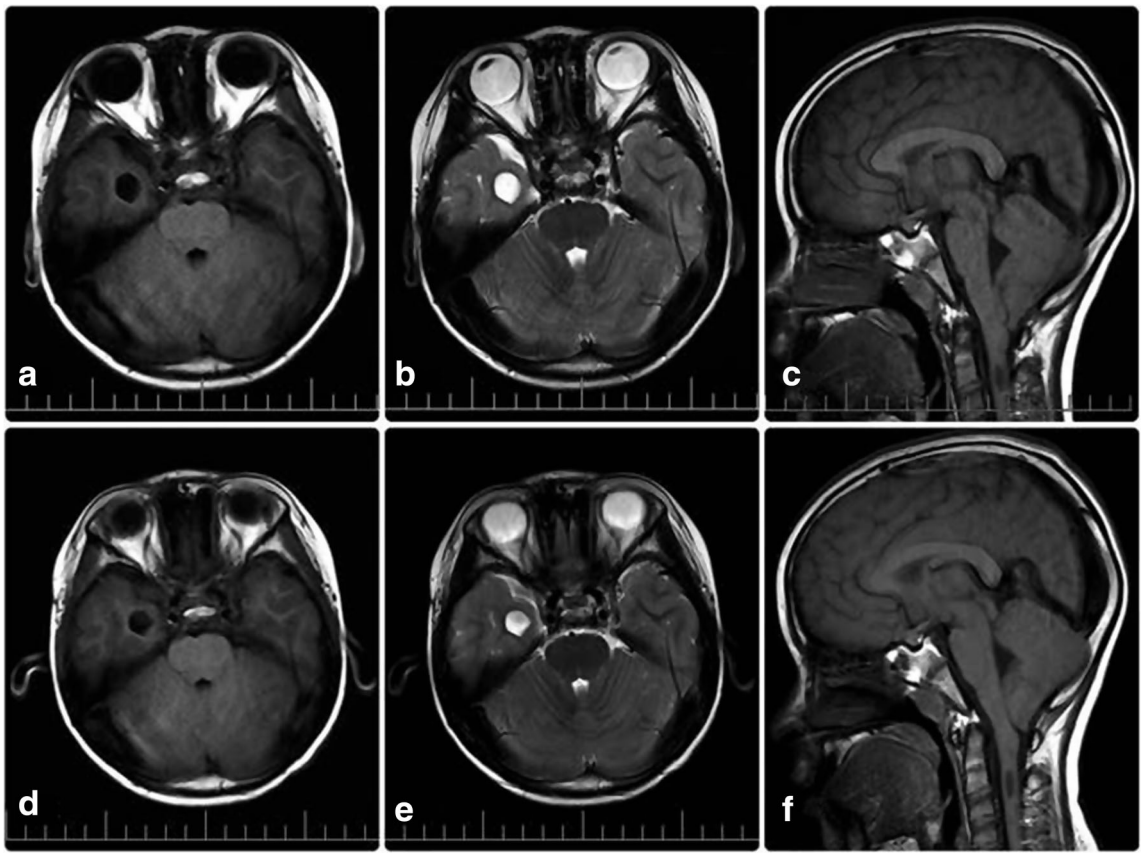

Fig. 4 Axial T1-weighted (**a**) and T2-weighted (**b**) MR imaging reveal the arachnoid cyst and syringomyelia are similar to the previous examination. Sagittal T1-weighted images (**c**) show an aggravated cerebellar tonsillar herniation. On the latest MR images (**d**–**f**), the cerebellar tonsillar herniation and syringomyelia show no significant progression

hypotheses: a volume-reservoir mechanism postulated by Maixner et al. and a "chronic idling" of the CSF absorption mechanism proposed by Ahn et al. [3, 9].

Some scholars recommended that the preferred treatment for shunt dependency syndrome or for relieving "benign" intracranial hypertension is insertion of LP shunt, considering the following two reasons: inserting the ventricular catheter into a small ventricle is difficult; LP shunting is less invasive [1, 8]. However, as Caldarelli et al. warned, this procedure has a risk of the formation of an acquired Chiari type I malformation [11].

Chiari type I malformation is generally considered a congenital neurological disorder; however, in recent years, its acquired form (ACIM) has been identified as a complication of CSF diversion procedures or the chronic spinal leakage [4, 11].

A reasonable explanation to account for this condition is that CSF diversion can lead to an abnormal craniospinal pressure gradient iatrogenically [4, 11]. Normally, CSF can be absorbed via cranial and spinal pathways. Due to the overdrainage in LP shunting, the normal cephalic absorptive pathways are disrupted, and CSF can be drained via the shunt, which may lead to a downward pressure gradient. There is even a case reporting development of ACIM after multiple lumbar punctures [12].

However, some scholars noted that ACIM could not only occur as a specific complication of LP shunts, but it has also been observed in children with ventricular shunts or in those with shunts draining the intracranial cavity [5, 6]. The "craniospinal pressure gradient theory" cannot explain this phenomenon, and thus, the "cephalocranial disproportion theory" seems to provide a reasonable hypothesis [5]. The extrathecal shunt drainage may lead to an arrest of cranial growth resulting in a relatively small skull, and further brain growth can no longer be accommodated; this secondary cephalocranial disproportion may be also associated with the caudal descent of the cerebral tonsils.

According to literatures, the incidence of ACIM secondary to LP shunt is less than 1% [1]. Riffaud et al. pointed out that the risk of symptomatic ACM and syringomyelia was closely associated with the valveless LP shunt, and they suggested a LP shunt with an adjustable valve to prevent such complications [11]. Rekate et al. used the valve system in LP shunting and did not come across any risk of ACIM [10]. Lam et al. observed two cases with ACIM after LP shunting, and they elected to alter the construct of the LP shunt by inserting a programmable valve, eventually achieving significant clinical and radiological reversal of the tonsillar herniation and the

associated syringomyelia [7]. More evidence suggested that the overdrainage complications including ACIM could be avoided by a programmable shunt. There are still some scholars suggesting that posterior fossa cranial decompression is the effective therapeutic procedure to resolve the ACIM.

Conclusion

Shunt dependency syndrome is a rare complication secondary to CP shunting in temporal arachnoid cysts, and LP shunting can effectively relieve the intracranial hypertension. The clinicians should be alert to the ACIM as a rare complication of CSF diversion procedures, and the potential protecting effect of the programmable valve in LP shunting should be emphasized.

Compliance with ethical standards

Conflict of interest The authors have no conflicts of interest to disclose.

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