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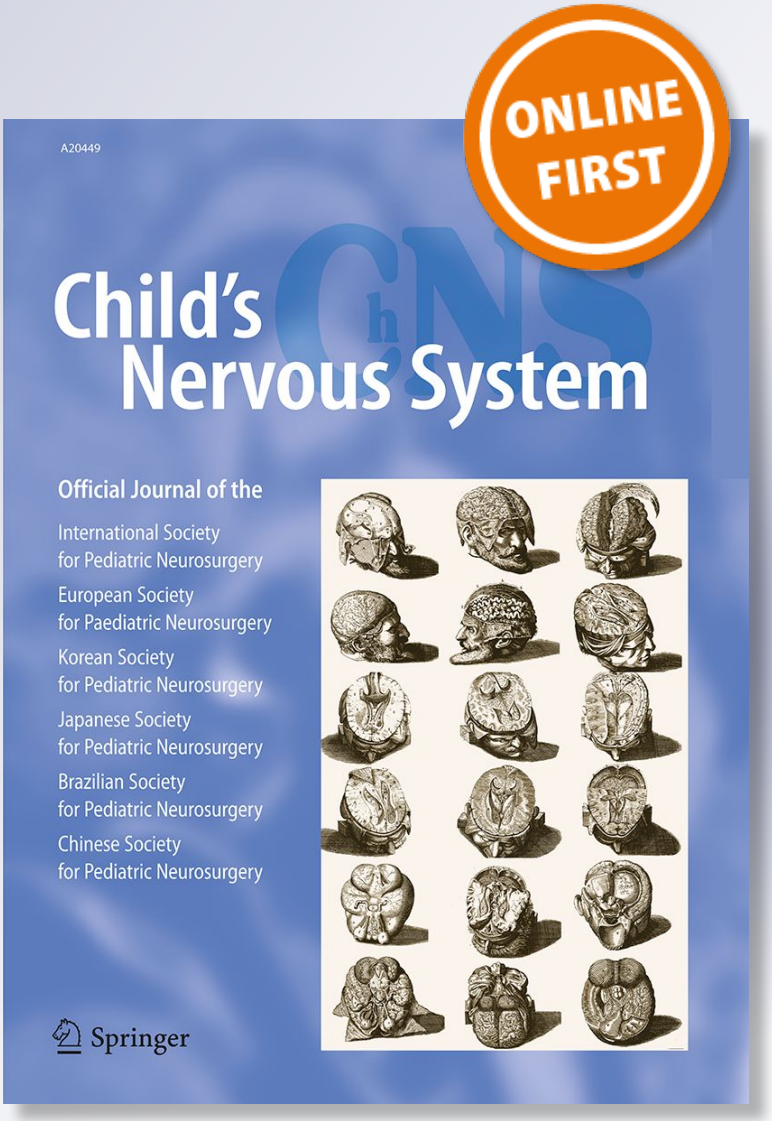

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CASE-BASED REVIEW

Acquired Chiari type I malformation managed by expanding posterior fossa volume and literature review

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Abstract

Purpose The acquired Chiari type I malformation is a rare late complication of supratentorial shunting in children which is often accompanied by abnormal cranial vault thickening. Several surgical treatments for this disease have been proposed including supratentorial skull enlarging procedures and subtentorial craniotomy. But there is still debate about the best treatment strategy for this disease.

Method and results We reviewed the current knowledge of this disease in the paper. We illustrated one patient of symptomatic acquired Chiari type I malformation who had cysto-peritoneal (C-P) shunting and ventriculoperitoneal (V-P) shunting. We observed the CSF flow dynamic of this patient at different periods. The acquired Chiari type I malformation disappeared on imaging after thinning the occipital planum combined with the standard surgical therapy of Chiari decompression. The symptoms of the patient were relieved after surgery.

Conclusion Overshunting manifestations require prompt recognition and management. Preventive measures should be taken which include making a stringent selection of cases being considered for surgery, avoiding C-P drainage, and placing of a programmable valve as initial treatment of intracranial arachnoid cysts (AC) if shunting is considered.

Keywords Chiari type I malformation · Decompression · Shunting · Arachnoid cyst · Hydrocephalus

Introduction

The Chiari type I malformation, also known as primary cerebellar ectopia, is characterized by a peg-like elongation of the cerebellar tonsils and medial aspect of the inferior vermis outside the cranial cavity into the cervical canal [1, 2].The acquired Chiari type I malformation is regarded as an iatrogenic form of hindbrain herniation [3, 4]. The tonsillar descent could be justifiable by the presence of a pressure gradient across the cranial and spinal compartments, such as spinal cerebrospinal fluid drainage. But recently, Chiari type I malformation has been reported as late complication of early treatment of hydrocephalus or supratentorial arachnoid cyst by means of shunting [2, 4, 5]. It is regarded as the most severe late complication of overshunting. Headache used to be the only symptom of this complication [1]. There is still debate about the surgical strategy to alleviate this symptom. We illustrate one patient with acquired Chiari type I malformation due to cysto-peritoneal (C-P) and following ventriculoperitoneal (V-P) shunting. The patient also had acquired craniocerebral disproportion (ACCD) which includes thick calvarial bones, small volume subarachnoid spaces, effacement of the arachnoid cisterns, and a crowded posterior fossa with cerebellar tonsillar herniation. We observed the CSF flow dynamic of the patient at different periods and adopted the surgical strategy suggested by Adriaan R. E. Potgieser which was proved useful in this patient [1].

Historical background

The acquired Chiari type I malformation was described for the first time in 1976 by Hoffman and Tucker [4], who reported it as a late complication of extrathecal shunt drainage in eight hydrocephalic patients. The incidence of this complication was about 1% in several reported series [1, 2]. The mechanism involved in the acquired Chiari type I malformation secondary to supratentorial shunting is still unknown. One possible explanation for the onset of the acquired disease is the modifications

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of skull growth induced by CSF shunting. It was reported that thickening of the cranial vault, with inward growth of the calvaria, was a usual finding in children with congenital hydrocephalus who was treated by means of extrathecal shunting [2]. The abnormal preoperative growth of head circumference will lead to a disproportion between the skull and its contents. Then, the correctly functioning of CSF shunting will result in the induction of inwards cranial growth to compensate for it [2]. Beside modifications of cranial vault, other changes were also observed at the cranial base with thickening of the sphenoidal plane, lesser sphenoidal wings and posterior clinoids, and with volume increase of the paranasal sinuses. All these modifications mentioned above are aimed at reducing the cranial-parenchymal disproportion which is brought about by CSF venting from the cranial cavity.

CSF hypotension seems to be the triggering factor involved in overdrainage in AC shunting [6]. At the initial stage, the ventricles will become enlarged and displaced towards the cyst along with the venting of CSF. Subsequently, the brain will also become displaced in order to fill up the gap left by the cyst. The venous system will become distended too. Meningeal swelling could be caused by venous engorgement which will contribute to dural and suture fibrosis. Decrease of the normal ICP and the cerebral pulse pressure are the other two contributing factors. In later stage, the paranasal sinuses will expand and the skull bones will become thicker by inward growth [6]. These osseous changes are also obvious at the posterior fossa. It will induce overcrowding of the neural structures and lead to tonsillar herniation. In addition, the reabsorption of CSF could probably be affected by all these changes above [6]. The changes are reversible initially. But, they will become permanent later on. They will produce cranioencephalic disproportion and hindbrain herniation.

Post-shunting will cause cephalocranial disproportion and secondary craniosynostosis which will result in a small skull that was unable to accommodate for brain growth [2, 4, 7]. This theory was first proposed by Hoffman and Tucker in 1976 as the explanation for the acquired disease. Actually, some authors do not agree completely with this theory. They prefer the description of a "posterior cranial fossa disproportion" rather than "cephalocranial disproportion" [2, 8], because no significant differences were detected by comparing the supratentorial volume of shunted and nonshunted patients. The result above challenges the so-called cephalocranial disproportion. Thus, an arrested posterior cranial fossa growth seems to be the main pathology even in patients with extrathecal shunts [9]. They suppose that shunting will probably cause an alteration of CSF flow dynamic. It will impair the mechanism involved in the brain development consequently [2, 8]. Some authors hypothesize that the development of the posterior fossa crowding could probably be a result of a ventriculoperitoneal gradient [8]. The gradient is usually caused by the altered flow dynamics in the subarachnoid space. It will also lead to a negative pressure phenomenon with impairment in the stenting of the cranial sutures [8]. Subsequently, posterior cranial fossa disproportion will be caused.

The occurrence of different pressures between the medullar and cranial compartments after shunting was speculated by some authors. Re-expansion of the neural tissue inside the posterior fossa after drainage of the cyst is another suggested mechanism [10]. Jakob made a morphological and biochemical description of the brain, which pointed out that the cerebellum would be the most "hydric" portion of the central nervous system [10]. So, the cerebellum seems to be the most easily hydrated part of the central nervous system. The increased volume of the content inside the posterior fossa upon shunting could be explained by this theory.

Clinical presentation

The symptom of headache which used to be the only clinical manifestation of acquired Chiari type I malformation may have been due to the reduction in the volume of the posterior cranial fossa [8]. It will lead to elevated pressure and compression of the brainstem contents. In addition to this, there is another explanation that the relative movement of CSF from the compressed spaces within the subarachnoid space plays a role [8]. It will lead to accentuation in the brain movement within the cranium, as inadequate CSF volume will not be enough to dampen this movement. The symptoms of nausea and vomiting appear in part of the patients. The presence of the area postrema in the floor of the fourth ventricle may account for the symptoms of nausea and vomiting [8]. Some serious patients were also reported to have impaired consciousness.

Diagnosis

Chiari malformation (CM) is specifically characterized by the descent of the cerebellar tonsils > 5 mm below the foramen magnum into the spinal canal [11]. CM-I is generally considered as a congenital neurological condition, although in recent years, its acquired form has been identified as a complication of CSF diversion procedures or chronic CSF leakage.

The acquired Chiari type I malformation was originally observed in patients with lumboperitoneal (L-P) shunting, multiple lumbar punctures, or baclofen pump placement [1, 11–13]. It was later recognized that an acquired Chiari type I malformation can also occur after C-P or V-P shunting [1, 14, 15]. An early-onset acquired Chiari type I malformation after the L-P shunting could be explained by the pressure gradient theory [4, 13]. A craniospinal pressure gradient after L-P shunting will be generated which will suck the cerebellum into the cervical canal. The acquired Chiari type I malformation caused by L-P shunting could be successfully treated by converting it to a V-P shunting [1, 4]. The successful treatment supports the pressure gradient theory. An early-onset acquired

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Chiari type I malformation after supratentorial shunting should be explained by a different mechanism as no downward pressure gradient is generated. This may be caused by overdrainage of the ventricles [1]. Overdrainage of CSF will lead to sagging of the brain as a whole. A small (but previously sufficiently sized) posterior fossa may also play a role [1], while late-onset acquired Chiari type I malformation which is characterized by modifications of the skull induced by the shunting is more difficult to explain.

Management

Avoiding unnecessary surgery for AC treatment is the main measure of prevention [6]. At present, no tests have been proposed that can foresee a satisfactory outcome in the management of intracranial AC. Several diagnostic methods including ICP recording, SPECT studies, or neuropsychological testing have been reported to be useful in the decision-making [6, 16]. But, none of the above has been proved to be totally reliable. Nowadays, most investigators prefer using endoscopic techniques or open surgical treatment rather than C-P shunting placement. When shunting is deemed necessary, we recommend using a programmable and gravitational valve as a useful tool which will prevent the appearance of the abovementioned CSF overdrainage manifestations. It will improve conditions for proper shunting.

In patients without ACCD, limiting the presumed overdrainage of the valve apparatus through the upgrading of the opening pressure or adding an anti-siphoning device to the CSF shunt system could be performed to impede further progression and re-establish a normal ventricular volume. When modifications of the CSF shunting fail or ACCD is already established, surgical treatment which aims at increasing the intracranial volume seems to be appropriate. A continuing debate about the optimal surgical treatment of the acquired Chiari type I malformation has always been going on. The subtemporal craniectomy introduced to offer an extra-volume for the intracranial content has nowadays been abandoned. As this procedure will result in impact of the atmospheric pressure on the open skull which actually favors CSF overdrainage, then, some authors introduce the supratentorial skull enlarging procedures which include anterior cranial vault advancement and biparietal expansion [1, 2], while some other doctors advocate a suboccipital craniectomy with or without C1 laminectomy and dural patch grafting [1, 10, 15]. This surgery is considered as a standard procedure for the "normal" Chiari type I malformation. It was first suggested by Adriaan R. E. Potgieser in 2016, which has been proved useful in three cases with acquired Chiari type I malformation [1]. However, the procedure may probably increase the risk of further sagging of the cerebellum. In our surgery, we further reduced the size of the occipital flap to avoid this complication.

Prognosis and outcomes

Supratentorial craniotomy, which is recommended by Professor Di Rocco, was proved to be useful in patients with acquired Chiari type I malformation. The combined surgical procedure of Chiari decompression and internal volume expansion of the posterior fossa by thinning of the occipital planum appeared to be effective in three cases reported by Adriaan R. E. Potgieser. It relieved the patient's symptoms and did not create new problems of a symptomatic descending cerebellum. This surgery was also proved useful in our patient. The symptom of headache was relieved significantly. MRI also showed that the Chiari type I malformation disappeared after surgery.

Exemplary case description

The patient was a 10-year-old girl with a left temporal arachnoid cyst (AC) when she was born (Fig. 1a). The surgery of C-P shunting was performed with a PS medical low valve in Children's Hospital of Nanjing when she was about 1 year old in 2009. The AC got smaller and disappeared eventually 5 years later after the procedure (Fig. 1b). The girl did well until 2018 when she reported the onset of episodic headaches, typically associated with an orthostatic posture. Neuroradiological investigation demonstrated smaller ventricles and an acquired Chiari type I malformation (Figs. 1c, d and 2b). We thought this was due to excessive drainage of CSF. Then, placement of a V-P CSF shunting was carried out in our hospital in 2018 (Fig. 1e). Valve pressure of the V-P shunting was set at 140 mm H_2O. At the same time, C-P shunting ligation was performed. The patient's headaches diminished after this surgical procedure. Two months later, the patient came to our hospital because of short paroxysmal non-postural headaches. We ensured that the V-P shunting was working properly. The CT scan showed that the ventricles became smaller than before (Fig. 1f). Then, the valve pressure of V-P shunting was gradually upgraded to 200 mm H_2O. The symptom of the patient was temporarily relieved. With the increase of valve pressure, the severity of the cerebellar tonsil hernia was improved on imaging and arachnoid cisterns around the brainstem became enlarged (Fig. 2c, d). With the increase of valve pressure, excessive drainage of CSF may be partially improved and CSF flow dynamic could probably be changed. It will create the increase of CSF in the subarachnoid space. Then, the increased CSF will alleviate the downward pressure as the brain tissue pulsates. At the same time, sagging of the brain will be alleviated with the increase of buoyancy created by intracranial CSF. But, the patient's headache returned one month later. We believe that this is because the limited volume of the cranial cavity is unable to meet the increased CSF and the ascending cerebellar tonsillar. Fundus examination revealed

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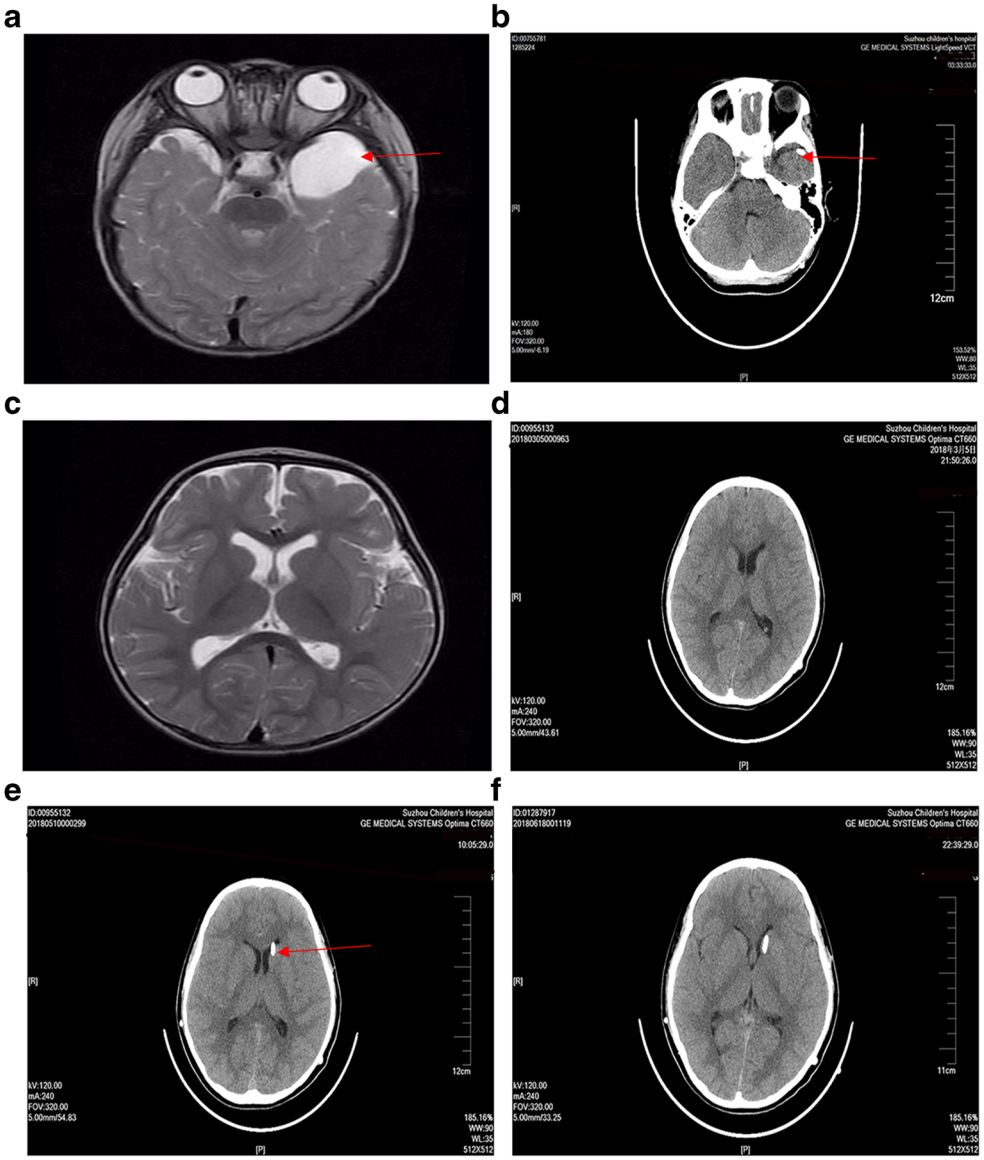

Fig. 1 Imaging manifestations of the AC and ventricles at different times. **a** The MRI showed a left temporal arachnoid cyst (AC) when she was born in 2009. **b** The AC got smaller and disappeared 5 years later after C-P shunting in 2014. **c** Neuroradiological investigation in 2009 demonstrated normal ventricles. **d** Neuroradiological investigation in 2018 demonstrated small ventricles when she got headaches. **e** Placement of a V-P CSF shunt was performed in 2018. **f** The CT scan showed smaller ventricles when the headaches reappeared two months after V-P shunting

papillary edema which means intracranial hypertension. And, eye examination showed decreased vision. These clinical manifestations mean that an urgent surgery is needed. Imaging results also showed thick calvarial bones (Fig. 3a, b), small volume subarachnoid spaces (Fig. 2d), effacement of the arachnoid cisterns, and a crowded posterior

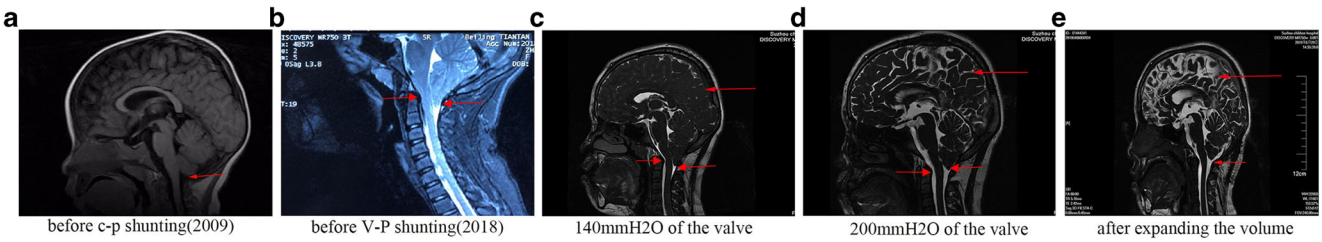

Fig. 2 The severity of the cerebellar tonsil hernia and the volume of the posterior fossa volume at different times. **a** The MRI showed that the patient had no cerebellar tonsil hernia before C-P shunt in 2009. **b** The MRI before V-P CSF shunt showed the cerebellar tonsil hernia in 2018. **c**, **d** With the increase of valve pressure, the severity of the cerebellar tonsil hernia was improved on imaging and arachnoid cisterns around the brainstem became enlarged. **e** MRI which was performed 9 months after expanding posterior fossa volume showed an adequate decompression and regression of the caudal tonsillar herniation. The volume of subarachnoid space also became increased than before

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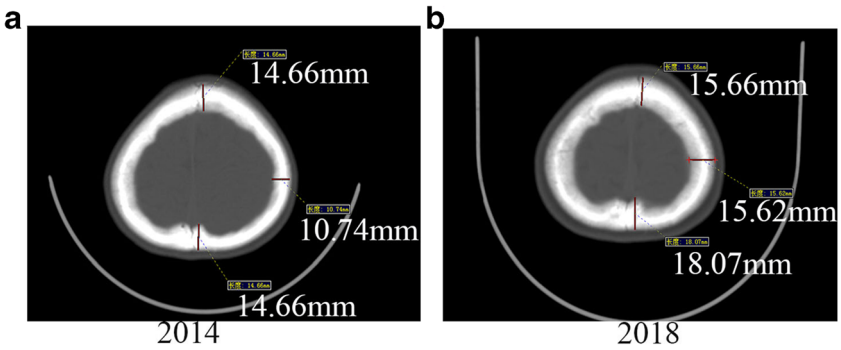

Fig. 3 The thickness of the calvarial bones at different times. **a** The thickness of calvarial bones in 2014. **b** The thickness of calvarial bones in 2018

fossa with cerebellar tonsillar herniation (Fig. 2d), all of which could be observed in a patient with ACCD. So, we decided to augment the intracranial volume. There are two surgical options available, i.e., supratentorial craniotomy and subtentorial craniotomy. As the decrease in the volume of subarachnoid space was most pronounced around the brainstem, we thought that we could improve local CSF flow around the brainstem by enlarging the posterior fossa volume. Then, the downward pressure will be partly alleviated as the brain tissue pulsates. But standard surgical therapy for Chiari decompression will probably increase the risk of further sagging of the cerebellum. In our surgery, we further reduced the size of the occipital flap to avoid this complication. We modeled the thickened occipital planum to a normal size and subsequently placed back the thinned bone (Fig. 4a). During the same procedure, a standardized C0 augmentation and C1 laminectomy were given (Fig. 4a, b); the circumoccipital fascia was also cut off. The patient's headaches diminished 3 days after the operation. MRI which was performed 9 months after the surgery showed an adequate decompression with an obvious increase in the volume of subarachnoid spaces around the brainstem and regression of the caudal tonsillar herniation (Fig. 2e). The lateral ventricles become enlarged (Fig. 4c). However, the cyst appeared again (Fig. 4d).

Conclusions

Overshunting manifestations require prompt recognition and management. Preventive measures should be taken before surgery. Thinning the occipital planum combined with the standard surgical therapy of Chiari decompression seems to be an effective method in expanding posterior fossa volume which will relieve the symptoms of the acquired Chiari type I malformation.

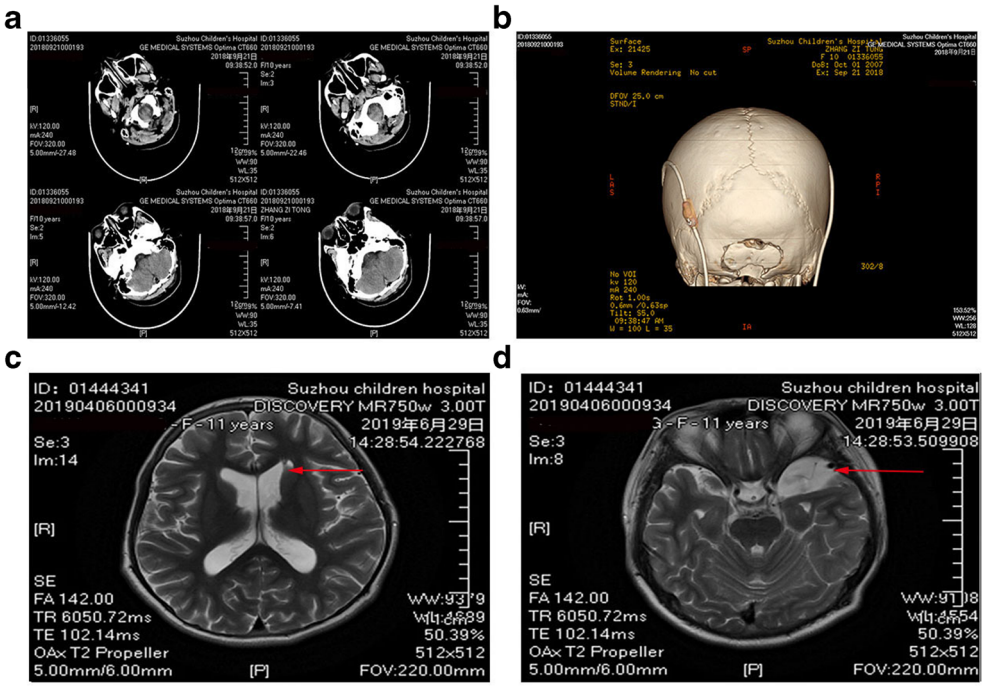

Fig. 4 Imaging manifestations after expanding posterior fossa volume. **a**, **b** The CT showed the thinned occipital planum, a standardized C0 augmentation and C1 laminectomy. **c** MRI which was performed 9 months after the surgery showed the enlarged lateral ventricles. **d** The cyst appeared again after surgery

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Compliance with ethical standards

Conflict of interest The authors received no financial and/or material support for the research reported in this paper. The authors state that there are no conflicts of interest arising from the research reported in this paper.

Ethics approval and consent to participate Not applicable.

Consent for publication Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

A copy of the written consent is available for review by the Editor-in-Chief of this journal.

References

1. Potgieser ARE, Hoving EW (2016) A novel technique to treat acquired Chiari I malformation after supratentorial shunting. Childs Nerv Syst 32:1721–1725
2. Caldarelli M, Novegno F, Di Rocco C (2009) A late complication of CSF shunting: acquired Chiari I malformation. Childs Nerv Syst 25:443–452
3. Chumas PD, Armstrong DC, Drake JM, Kulkarni AV, Hoffman HJ, Humphreys RP, Rutka JT, Hendrick EB (1993) Tonsillar herniation: the rule rather than the exception after lumboperitoneal shunting in the pediatric population. J Neurosurg 78:568–573
4. Di Rocco C, Velardi F (2003) Acquired Chiari type I malformation managed by supratentorial cranial enlargement. Childs Nerv Syst 19:800–807
5. Di Rocco C, Tamburrini G (2003) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 38:164
6. Martínez-Lage JF, Ruíz-Espejo AM, Almagro M-J, Alfaro R, Felipe-Murcia M, López-Guerrero AL (2009) CSF overdrainage in shunted intracranial arachnoid cysts: a series and review. Childs Nerv Syst 25:1061–1069
7. Fagan LH, Ferguson S, Yassari R, Frim DM (2006) The Chiari pseudotumor cerebri syndrome: symptom recurrence after decompressive surgery for Chiari malformation type I. Pediatr Neurosurg 42:14–19
8. Osuagwu FC, Lazareff JA, Rahman S, Bash S (2006) Chiari I anatomy after ventriculoperitoneal shunting: posterior fossa volumetric evaluation with MRI. Childs Nerv Syst 22:1451–1456
9. Nishikawa M, Sakamoto H, Hakuba A, Nakanishi N, Inoue Y (1997) Pathogenesis of Chiari malformation: a morphometric study of the posterior cranial fossa. J Neurosurg 86:40–47
10. Mendonça R, Lima TTF, Oppitz PP, Raupp SF (2006) Herniation of the cerebellar tonsils after suprasellar arachnoid cyst shunt: case report. Arq Neuropsiquiatr 64:523–525
11. Liu H, Yang C, Yang J, Xu Y (2017) Pediatric Chiari malformation type I: long-term outcomes following small-bone-window posterior fossa decompression with autologous-fascia duraplasty. Exp Ther Med 14:5652–5658
12. Hoffman HJ, Tucker WS (1976) Cephalocranial disproportion. A complication of the treatment of hydrocephalus in children. Childs Brain 2:167–176
13. Payner TD, Prenger E, Berger TS, Crone KR (1994) Acquired Chiari malformations: incidence, diagnosis, and management. Neurosurgery 34:429–434 discussion 434
14. Hassounah MI, Rahm BE (1994) Hindbrain herniation: an unusual occurrence after shunting an intracranial arachnoid cyst. Case report. J Neurosurg 81:126–129
15. Kim S-K, Cho B-K, Chung Y-N, Kim H-S, Wang K-C (2002) Shunt dependency in shunted arachnoid cyst: a reason to avoid shunting. Pediatr Neurosurg 37:178–185
16. Di Rocco C, Tamburrini G, Caldarelli M, Velardi F, Santini P (2003) Prolonged ICP monitoring in Sylvian arachnoid cysts. Surg Neurol 60:211–218

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