Acta Neurochir (2010) 152:1419–1423
DOI 10.1007/s00701-010-0658-x

CASE REPORT

Acute intracranial hypertension and shunt dependency following treatment of intracranial arachnoid cyst in a child: a case report and review of the literature

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Received: 1 February 2010 / Accepted: 6 April 2010 / Published online: 21 April 2010

Abstract Arachnoid cysts are developmental anomalies that represent 1% of all intracranial space occupying lesions. Treatment of symptomatic cases may include shunting of the cyst or an open fenestration of it, among other less acceptable surgical procedures. Each procedure has its own pros and cons. We present a case of the development of an acute intracranial hypertension during cysto-peritoneal shunt malfunction in a child. We describe the possible mechanism of these phenomena and, based on this report and by reviewing other case series in the literature, we raise the possibility that acute increase in intracranial pressure and the development of shunt dependency, although rare, are important complications of shunting an arachnoid cyst.

Keywords Arachnoid cysts · Intracranial hypertension · Pediatric · Shunt

Introduction

Congenital arachnoid cysts (AC) are developmental anomalies in which cerebrospinal fluid (CSF) accumulates between the layers of the arachnoid membrane. The pathogenesis of this anomaly is not yet fully understood and may involve an abnormal genesis of the embryonic mesenchyme or an abnormal flow of CSF in the embryo.

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Most of the ACs discovered during the first two decades of life, usually as a result of an asymptomatic macrocephaly or as an incidental finding after imaging of the head.

ACs represents 1% of all intracranial space occupying lesions. They can remain asymptomatic for life, can undergo spontaneous regression or may enlarge and become symptomatic. The enlargement may be as a result of fluid accumulation or due to spontaneous or traumatic bleeding in the cyst. The symptoms may include seizures, signs of increased intracranial pressure, psychomotor retardation, or focal neurological signs. While it is generally accepted that asymptomatic cases do not require any intervention [4], controversy still remains regarding the preferred treatment for the symptomatic lesions. Currently, the two main surgical options that are widely in use are shunting of the cyst (mainly cysto-peritoneal shunt) or partial resection plus fenestration [3, 6, 8–11, 14, 17, 18, 21, 23]. Although placing a shunt is a minor procedure with little risks compared to fenestration, it still carries the risks of infection and malfunction with the latter may causing re-accumulation of fluids in the cyst. On the other hand, fenestration has the disadvantage of high percentage rate of recurrence of the cyst (up to 40% rate of recurrence) [7].

In this article, we will discuss another disadvantage of shunt placement—the possibility of developing an acute intracranial hypertension during shunt malfunction and, as a consequence, turning a non-shunt-dependent patient to a dependent one.

Case report

A 4.5-year-old boy first presented to another hospital at the age of 5 months with an asymptomatic increase in head circumference. Based on computed tomography (CT) of the

head, he was diagnosed with left sylvian fissure arachnoid cyst and a cysto-peritoneal (CP) shunt was inserted. The operative and post-operative courses were normal and there was normalization of the head circumference. Three years later, at the age of 3.5 years, 5 days after a minor head trauma, he presented to the same hospital with signs and symptoms of increased intracranial pressure (ICP). CT scan of the head revealed small residual of the arachnoid cyst and small ventricles. A measurement during a lumbar puncture revealed a pressure above 40 cm H2O. A lumbo-peritoneal (LP) shunt was inserted.

Eight months later, he was first presented to our hospital with a clinical picture of shunt over drainage (Figs. 1, 2, 3) and an antigravitation device was placed. Three months later, he returned to our institute with severe headaches and drowsiness due to shunt malfunction and was operated on once more for revision and repair of the LP shunt.

In both times, there was no evidence of low level of the cerebellar tonsils on head CT. On April 2008, 3 months after his last surgery, he was referred to another hospital due to persistent subcutaneous fluid collections under his abdominal surgical wounds. CT scan of the head showed a residue of the arachnoid cyst with normal sized ventricles. A magnetic resonance venography (MRV) was done to rule out venous insufficiency as a possible cause for the increased ICP. The MRV was normal and both the CP shunt and the LP shunt were removed. Few days later, he presented again to the same hospital with headaches, drowsiness, and a CSF leak from the cranial surgical wound. This time, a left temporal craniotomy and fenestration of the arachnoid cyst were performed. Few days later, due to a persistent cranial pseudomeningocele, a new CP shunt was inserted (June 2008).

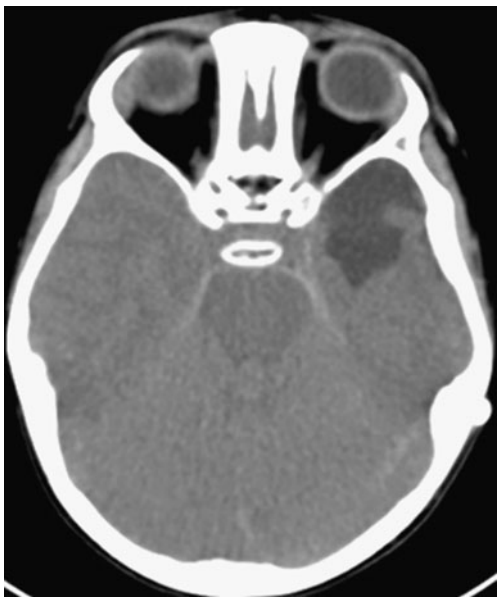

Fig. 1 Head CT scan taken 8 months after the insertion of an LP shunt. The patient was in clinical state of shunt over drainage (July 2007)

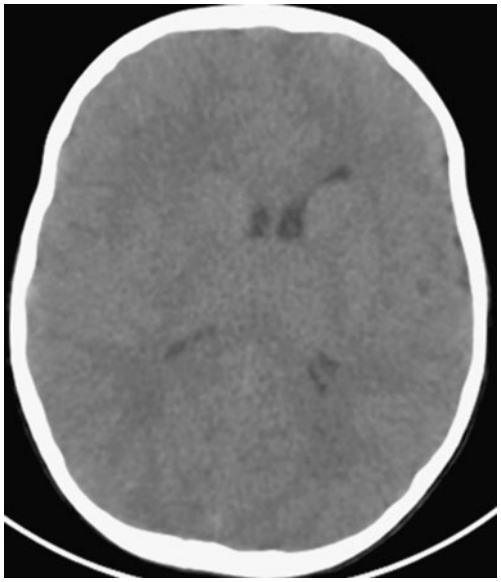

Fig. 2 Residue of left sylvian fissure arachnoid cyst

The patient returned to our department on August 2008 due to shunt infection. After a course of antibiotics, his CP shunt was replaced and he was discharge from the hospital in a generally good condition and as he is being well since then.

We would like to emphasize that during his last hospitalization, while he was with a temporary catheter in the cyst space that was draining outside; the amount of CSF that was accumulated in the draining bag was about 700 cc per day, almost equal to the amount of CSF that he produced per day. Furthermore, every time we attempted to

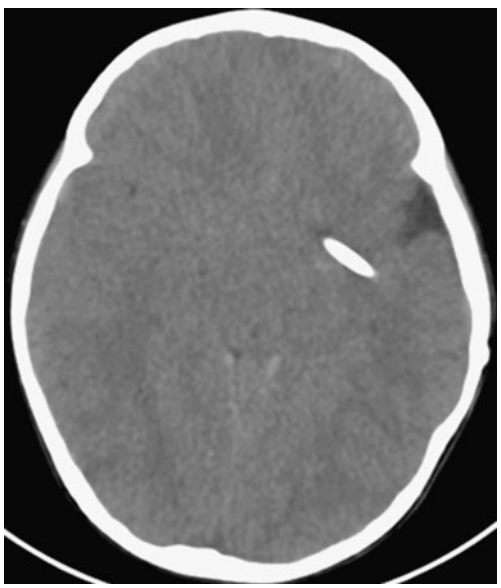

Fig. 3 Ventricles size and location of the CP shunt

close the catheter the child developed headaches, decrease in consciousness and bradycardia.

Discussion

In order to try to explain the development of shunt dependency and increased ICP in a child with presumed normal CSF dynamics and normal ICP prior to the insertion of the shunt, we need to understand the possible relationship between an arachnoid cyst and the subarachnoid space. Smith et al. [22] has postulated that there is an anatomical communication between the subarachnoid space and the arachnoid cyst and that CSF continues to enter the cyst from the subarachnoid space. According to his theory, a one-way valve prevents the fluid from going out of the cyst and thus enlarging the cyst. This anatomical communication was repetitively verified by CT after an intrathecal injection of metrizamide [13, 15, 19] and the slit-valve mechanism have been demonstrated by means of a preoperative cine-mode MRI, which was further confirmed during endoscopic intervention [20].

During the early 1990s, new reports on the development of shunt dependency in children treated for intracranial AC begun to emerge [2, 16, 24]. These articles described five cases in which children were treated with CP shunts for intracranial AC. In all cases, the children developed acute intracranial hypertension during shunt malfunction. In four of five cases, both the ventricles and the cysts were normal or small in their size at the time of the increase ICP, similarly to our case. In all five cases, no evidence was present of increased ICP prior to the insertion of the CP shunt. Interestingly, in one of the cases described by Maixner et al. [16] a temporary cystostome was inserted, which was noted to drain CSF in an amount equal to the one produced per day. This observation is similar to what we saw in our case and suggests that there was minimal, if any, natural absorption of CSF. As suggested by Maixner et al. [16], one way to explain the appearance of acute intracranial hypertension in the presence of CP shunt malfunction is that while the shunt was functioning, all the CSF drained through it. This caused the cyst to collapse (with the possibility of developing adhesions in the cyst) and to obstruct the natural drainage of CSF from the cyst to the subarachnoid space. Owing to this obstruction, the fluid accumulates in the cyst and cannot egress to the subarachnoid space. Instead, the fluid finds its way to the brain parenchyma, similar to the mechanism that was suggested in pseudotumor cerebri or in the slit ventricle syndrome.

Arai et al. [3] described 77 cases of AC in the middle fossa treated with a CP shunt. In all cases, after treatment, a major decrease in the size of the AC was seen. A 10% rate of shunt malfunction was noted, half of them (a total of four cases) presenting with acute intracranial hypertension. In all four cases, the AC was small and the ventricles were either small or normal sized. The average time, from shunt insertion to shunt malfunction, was 57 months. Based on these observations and on their experience with a small group of patients, which had their PC shunts removed electively after an average time of 3.3 years, this group suggested that all CP shunts should be remove once the AC has disappeared, mainly to avoid the complication of shunt dependency and the appearance of acute intracranial hypertension.

Moreover, Kim et al. [12] also supported the concept of avoiding, as much as possible, AC treatment with CP shunts. They described eight cases over a period of 10 years that were treated with CP shunts for intracranial AC, which developed the complication of shunt dependency and presentation of acute intracranial hypertension during shunt malfunction. In all cases, the size of the ventricles was normal prior to insertion of the shunt and during its malfunction; however, an ICP monitor demonstrated high pressure levels in all cases during the period of shunt malfunction. The average time from shunt insertion to shunt malfunction was 41 months. This group also experienced difficulty in explaining the reason for the appearance of shunt dependency. They opted for the theory that was first described by Ahn et al. [1] which supposed that there has to be some sort of connection between the subarachnoid space and the AC. After the insertion of the shunt tube to the cyst, the natural CSF absorption mechanism, which was normal before the insertion of the shunt, did not need to work. They referred to it as “chronic idling” of the CSF absorption mechanism which has now become impaired and is not able to compensate for the acute ceasing of CSF drainage through the shunt. Kim et al. [12] concluded that it is better to avoid inserting a CP shunt as a primary treatment for intracranial AC and, if non-the-less necessary, it is better to reach to the point where this shunt is no longer needed, as soon as possible.

They also recommended that in the case of CP shunt malfunction in the presence of acute intracranial hypertension the preferred treatment is insertion of LP shunt (as was performed in our case).

It is important to remember that shunting an arachnoid cyst may induce the formation of an acquired Chiari I syndrome, as was described in the work of Caldarelli et al. [5]. Their work included two cases in which symptomatic tonsilar herniation were diagnosed few years after shunting an intracranial AC. Both cases were presented with headache and bilateral papilledema. As we mentioned above, there was no evidence of low-lying tonsils during the decompensations of the patient described in our case.

In summary, so far less than 20 cases of intracranial arachnoid cysts with presentation of acute intracranial

hypertension in the presence of CP shunt malfunction have been published. Our case represents these set of circumstances and what we believe to be an established possible complication of treatment of intracranial AC with CP shunt. From our own experience and based on the works mentioned above, we believe that this manifestation is a rare complication in clinical practice worldwide. However, turning a non-shunt dependent patient into a dependent one is a poor consequence as was shown in the prolonged morbidity of our patient.

Other studies, such as the one of Zada et al. [25] also supported the notion of craniotomy and fenestration as first line treatment in case of intracranial AC, especially in the subgroup of children under the age of 2 years who present with macrocephaly or another neurological signs and without hydrocephalus. They propose that the insertion of CP shunt should only be applied to failed cases. Regarding the possibility of removing the shunt electively, more studies need to be done in order to prove its efficacy.

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Comments

There is a general belief in the neurosurgical community that shunt dysfunction is mainly related to CSF diversion procedure of the ventricular space, that is to say complications of the treatment of hydrocephalus. Laliv and Michowitz present a new case of shunt dysfunction occurring after the insertion of a cysto-peritoneal shunt (CPS) in a temporo-sylvian AC. Looking at previous report about this complication, they were able to identify less than 20 cases of intracranial AC complicated with acute intracranial pressure at the time of CPS malfunction. This complication probably reflects a sort of decreased intracranial compliance due to the chronic drainage of CSF with a low pressure shunt. These patients behave almost like those who have a slit ventricle syndrome and that are prone to develop acute intracranial hypertension in case of shunt blockage. Although the authors mention in their last sentence the possibility of elective shunt removal in their patient, we would advise to keep the patient for several days in the ward to be sure that he would not develop a delayed raised ICP or to be prepared to restore the intracranial compliance with an expansion craniotomy.

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This paper describes the development of shunt dependency and the need for multiple procedures following placement of a cystoperitoneal shunt in the treatment of a temporal arachnoid cyst. This case highlights again the long-standing debate concerning the treatment of arachnoid cysts in childhood. It also touches on some of the other questions raised by this topic, what symptoms or signs constitute an indication for treatment? What is the optimal age for treatment? How should treatment success be determined?

In contrast to the suggestion that this is an exceptional case, I suspect that the scenario presented is one that is all too familiar to a number of pediatric neurosurgeons. The pathophysiology of arachnoid cysts, treated and untreated remains poorly understood though most would agree that managing the complications of the failed shunt or shunt malfunction in the context of arachnoid cysts can be a formidable task typified by repeated interventions, prolonged hospital admissions not to mention significant morbidity.

The statement "placing a shunt is a minor procedure with little risks compared with fenestration" is one that would be questioned by most pediatric neurosurgeons and probably not supported by current literature; whilst there is indeed no universal consensus of treatment the prevailing mood seems to be toward fenestration (either open or endoscopic) at least in the first instance. Whether or not fenestration of this temporal arachnoid cyst into the basal cisterns as an initial treatment in this case would have had a better outcome remains unknown though whenever there is a treatment alternative to placement of a shunt this is surely worthy of consideration.

Certainly, if reduction in cyst size is the end point by which success is to be measured then fenestration is likely to be judged unfavorably. Whilst it is always satisfying to see a reduction in cyst size and re-expansion of adjacent brain on the post-treatment imaging, this is by no means essential for success, the equilibration of pressure between the cyst and the rest of the CSF pathway should perhaps be considered the more appropriate goal, a goal more likely to be achieved via fenestration than drainage

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