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Case Report

Rare case of a rapidly enlarging symptomatic arachnoid cyst of the posterior fossa in an infant: A case report and review of the literature

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Abstract

Background: Intracranial arachnoid cysts are space-occupying lesions that typically remain stable or decrease in size over time. Cysts in infants younger than 1 year of age are remarkably different from those in older children and adults in terms of cyst localization and enlargement. Arachnoid cysts of the posterior fossa (PFACs) are very rare in infants and do not typically grow or present with clinical symptoms, such that surgical treatment is generally considered to be unnecessary. Here, we describe an extremely rare case of an infant with a rapidly enlarging symptomatic PFAC that was successfully treated with surgery.

Case Description: A 4-month-old boy presented with increasing head circumference and a rapidly enlarging arachnoid cyst in the left posterior fossa with ventriculomegaly, which was documented using serial imaging over the preceding 2 months. We performed a microscopic resection of the cyst membrane to remove the mass effect as soon as possible and facilitate normal development. To confirm dural closure and prevent cerebrospinal fluid leakage, we also performed short-term (7 days) percutaneous long-tunneled external ventricle drainage after the surgery. Magnetic resonance imaging over a 4-year follow-up period revealed adequate reduction of the ventricle and cyst. The patient no longer exhibited progressive macrocrania and showed normal development.

Conclusion: To our knowledge, this is the second successful case of surgical treatment of an enlarging symptomatic PFAC in an infant. Our surgical strategy for the treatment of this rare case can serve as a guide for surgeons in similar future cases.

Key Words: Arachnoid cyst, cyst enlargement, hydrocephalus, posterior fossa

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INTRODUCTION

Congenital arachnoid cysts are rare central nervous system malformations that represent only 1–2% of all intracranial masses.[23] These lesions are often clinically silent and rarely increase in size.[1,12,13,19] As such, arachnoid cysts seldom require surgical intervention.

The characteristics of arachnoid cysts vary remarkably between infants and older children or adults. In adults, the majority of intracranial arachnoid cysts arise in the supratentorial region (90%), with the middle cranial fossa being the most common site (60%) of origin and 10–20% of arachnoid cysts occurring in the posterior fossa.[4,7,22] In contrast, arachnoid cysts in infant patients commonly occur at the middle fossa (15%) and intraventicularly (14%), with very low percentages occurring in the posterior fossae (4.6%).[13] Lee *et al.* recently reported that cysts in infants can occasionally enlarge (16.2%), but that arachnoid cysts of the posterior fossa (PFACs) have an extremely low prevalence of enlargement in infant cases (2.3%)[13] and are generally asymptomatic such that they do not require treatment.

Here, we present an extremely rare case of a 4-month-old boy who presented with a rapidly enlarging symptomatic PFAC. After considering the current options for treating symptomatic PFACs in infants,[1,3,11,14,18,22,24] we performed a necessary microscopic membranectomy with post-surgical percutaneous long-tunneled external ventricle drainage (PL-EVD)[5,15] and obtained a good outcome. Because this is the second successful case of surgical treatment for an enlarging symptomatic PFAC in an infant,[3] the surgical strategy described here can guide surgeons in future clinical decisions.

CASE PRESENTATION

History

The patient in the present case was a boy that was born by vaginal delivery at 42 weeks of gestation. He had a birth weight of 2546 g, a normal head circumference (32.1 cm), and exhibited no neurological abnormalities. Based on the detection of an ultrasonographic abnormality, magnetic resonance imaging (MRI) was performed at 58 days after birth and a cystic lesion (1.2 cm × 2.9 cm at its largest diameter) was identified in the left retrocerebellar region of the posterior fossa [Figure 1]. The cerebellum was smaller than average, but there were no signs of significant compression. At this time, the circumference of the patient's head was slightly enlarged and measured in the upper limit for his age group (43.0 cm).

Examinations

At 4 months of age, the patient's head circumference increased to 48.0 cm (>2 standard deviations above the mean for his age), and MRI showed significant enlargement of the cyst (4.0 cm × 3.5 cm at its largest diameter) with compression of the cerebellum and hydrocephalus caused by obstruction of the fourth ventricle outlets [Figure 2]. Intracranial hypertension was also recognized based on sunset phenomena in the patient's eyes, scalp varicosis, and separation of the coronal and sagittal cranial sutures. The patient did not exhibit any other abnormal neurological signs, and was diagnosed with an enlarging PFAC and obstructive hydrocephalus.

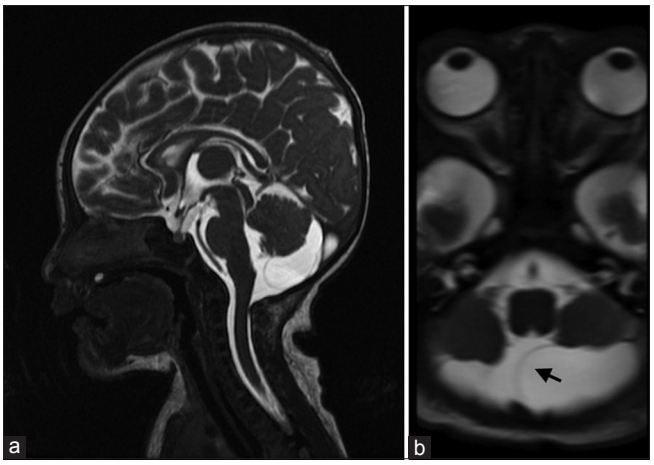

Figure 1: Initial sagittal (a) and axial (b) T2 magnetic resonance images of the male patient at 58 days of age.The image demonstrates a cystic lesion with the septum (arrow) in the posterior fossa

Surgical procedure

Clinical signs and symptoms of hydrocephalus were observed when the patient was 4 months of age. Thus, we performed a suboccipital craniotomy and direct microscopic treatments to resect the cyst membrane via a medial occipital approach with the patient in the prone position. This was done to restore normal communication between the Magendie and Lushka foramina and the subarachnoid space. Because the inner membrane of the cyst adhered to the rostral medullary velum and compressed the fourth ventricle, we carefully detached and resected both the outer and inner membranes of the cyst to open the fourth ventricle and cisterna magna [Figure 3]. During the surgery, we confirmed good cerebrospinal fluid (CSF) flow at the cerebral aqueduct and cisterna magna. A histopathological study of excised tissues stained with hematoxylin and eosin showed arachnoid mesothelial cells arranged among fibers of the connective tissue (data not shown), confirming our diagnosis. Immediately prior to the craniotomy, we performed PL-EVD to allow for external CSF drainage for 7 days after the surgery to prevent infection due to CSF leakage.[5] The procedure of PL-EVD was as follows. The posterior horn of the lateral ventricle was cannulated with a ventricular catheter. The peritoneal catheter was then subcutaneously tunneled with the proximal end connected to the distal end exiting at the lower anterior

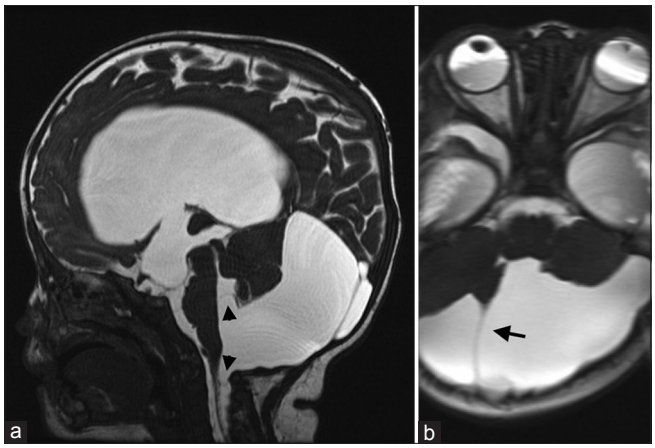

Figure 2: Preoperative sagittal (a) and axial (b) T2 magnetic resonance images of the patient at 4 months of age. An enlarged cystic lesion can be observed descending into the foramen magnum with the septum expanding beyond the midline (arrow). Blocked communication with the fourth ventricle, compression of the brainstem and fourth ventricle (arrowheads), and marked ventriculomegaly (stars) were evident

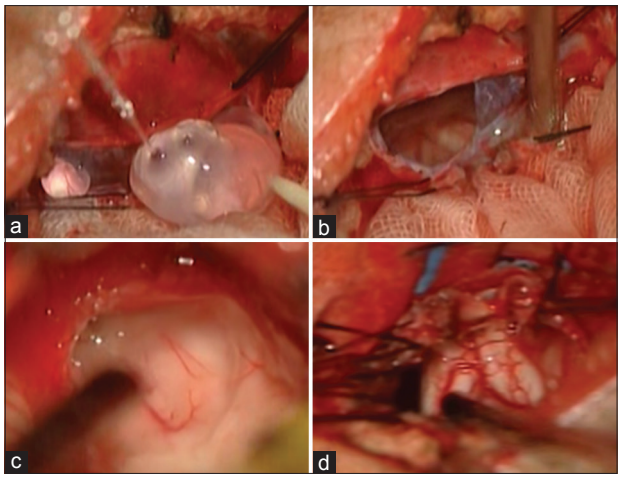

Figure 3: Intraoperative photographs. (a and b) The cyst wall was exposed and resected. (c) Opening of the cerebral aqueduct was confirmed. (d) Opening of the foramen magnum was confirmed

chest wall, 25 cm away from the proximal end. The distal catheter was then secured to the skin and connected to a ventricular drainage system.

Postoperative course

The postoperative course was uneventful. MRI performed 1 month after the surgery revealed that the cyst had disappeared, alleviating compression of the cerebellum and halting the patient's macrocrania. Further, the sulci of the cerebellum and the cerebellar pontine angle became easier to observe. At the last follow-up (performed 4 years postoperatively), the cyst wall was unidentifiable and the cerebellum had grown to near-normal size [Figure 4]. The patient was doing well and showed normal development.

DISCUSSION

An arachnoid cyst consists of a cavity lined with arachnoid cells and filled with fluid that closely resembles CSF.[23] Most arachnoid cysts are clinically silent and remain static in size; however, on rare occasions, these cysts can increase in size and produce symptoms due to mass effects.[1,12,13,19] Although the mechanisms that drive arachnoid cyst enlargement remain unknown, it has been postulated that cysts may expand due to (1) a ball-valve action that allows CSF entry into the cyst; (2) intracystic production of fluid by secreting cells occasionally found on the cyst walls; or (3) movement of fluids following an osmotic gradient.[10,23]

The differential diagnosis of PFACs in infants mainly includes the Dandy–Walker malformation, mega cisterna magna, and Blake's pouch cyst.[6,23] It is also important to exclude other possible cystic lesions such as cerebellar cystic astrocytoma, cystic hemanglioblastoma, and epidermoid or dermoid tumors.[6] MRI findings are very useful for the differential diagnosis of a PFAC, as demonstrated in our present case. Finally, postoperative histological studies are also useful for a definitive diagnosis.

Although very little data exist regarding the clinical characteristics of arachnoid cysts in infant cases, these characteristics have gradually become evident.[3,11,13,18,20] Lee *et al.* demonstrated differences in cyst localization between infants younger than 1 year of age and older children and adults, additionally indicating a much lower prevalence of PFACs in infants compared to adults.[13] Further, the authors found that even enlarging PFACs in infants did not necessarily warrant surgical treatment based on the absence of clinical symptoms. Accordingly, the present case of a 4-month-old boy with a rapidly enlarging symptomatic PFAC and successful surgical treatment is extremely rare and thus of clinical value.

To date, only 2 cases of enlarging symptomatic PFACs and surgical treatment have been reported in infant patients [Table 1].[3] Both patients in these cases were younger than 9 months of age and were treated by endoscopic fenestration, but clinical improvement was reported in only 1 case. Thus, our present case is the second report of successful surgical treatment of a rapidly enlarging symptomatic PFAC in an infant.[3]

The infratentorial space is much smaller than the supratentorial space; accordingly, mass effects related to arachnoid cysts are more probable in the infratentorial space than in the supratentorial space. Especially in infants with very small infratentorial spaces, mass effects must be removed as soon as possible to facilitate normal development.

PFACs can be particularly challenging to treat depending on the cyst size, cyst location, and the presence of obstructive hydrocephalus.[6,21] To this

Table 1: Reported infant cases of enlarging symptomatic arachnoid cysts of the posterior fossa with surgical treatment

Authors	Age/sex	Age at diagnosis	Symptoms and neurological findings	Surgical treatment	Outcome
Choi[3]	3 m uk	Uk	Uk	Endoscopic fenestration	No clinical improvement, subdural fluid collection
Choi[3]	9 m/uk	Uk	Hydrocephalus	Endoscopic fenestration	Clinical improvement
Present report	4 m/M	58 days	Hydrocephalus, intracranial hypertension	Open cyst excision + PL-EVD	Clinical improvement

M: Male, m: Month, PL-EVD: Percutaneous long-tunneled external ventricle drainage, uk: Unknown

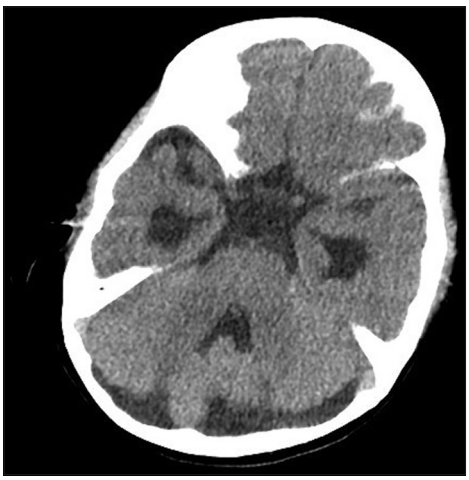

Figure 4: Computed tomography performed 4 years after the surgery. The image demonstrates absence of the septum as well as reduced compression of the brainstem and fourth ventricle. Furthermore, the image clearly depicts the sulci of the cerebellum, the cerebellopontine angle, and amelioration of the patient's obstructive hydrocephalus

end, there is no consensus regarding a standard or optimal surgical treatment for PFAC. Various treatment approaches exist, including microsurgical excision or fenestration by craniotomy, cyst shunting, and endoscopic fenestration.[1,3,11,14,18,22,24] While some authors have reported that there is no significant difference between these surgical methods,[2,8] studies evaluating the management of PFACs in a pediatric population demonstrated that craniotomy with excision was the first-line approach.[14,16] We selected to perform a microscopic membranectomy of both the inner and outer membranes because the resolution of obstructive hydrocephalus was a high priority and it was necessary to remove the mass effect to facilitate normal development of the infant.

There is relatively high risk of postoperative complications such as CSF leakage, subdural fluid collection, and meningitis after surgery on the posterior fossa, especially in infants.[3,9,17,18] In the present case, we performed PL-EVD and continued drainage for 7 days after surgery to confirm dural closure and prevent CSF leakage, which may have led to an infection such as meningitis.[5,15] After removal of the drainage tube, no CSF leakage or meningitis was observed and the skin wound healed completely. The reasons for performing PL-EVD are as follows: (1) it is very difficult to stitch and effectively close an incision of the dura mater in the posterior fossa region in infants; (2) as the skin, subcutaneous fat, and muscles of infants are relatively thin, CSF tends to leak externally when the dura mater is breached; (3) it is difficult to evaluate whether infant patients have sufficient ability to reabsorb leaked CSF; and (4) PL-EVD is a simple and effective method for lowering the risk of infection after surgical treatment of a PFAC.[15] To our knowledge, this is the first case in which PL-EVD performed in an infant with a PFAC. Another necessary precaution during surgical intervention for a PFAC is the protection of important neurovascular structures such as the brainstem. In summary, these surgical considerations and techniques provide important and useful insight for neurosurgeons planning surgical treatment for PFACs in infant patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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