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

Nobuyuki Takeshige, Tomoko Eto, Shinji Nakashima, Kiyohiko Sakata, Hisaaki Uchikado, Toshi Abe1, Motohiro Morioka

Departments of Neurosurgery and 1Radiology, Kurume University School of Medicine, Kurume, Japan

E-mail: *Nobuyuki Takeshige - takeshige_nobuyuki@med.kurume-u.ac.jp; Tomoko Eto - eto_tomoko@med.kurume-u.ac.jp; Shinji Nakashima - nakashima_shinji@med.kurume-u.ac.jp; Kiyohiko Sakata - kyo@med.kurume-u.ac.jp; Hisaaki Uchikado - uchikado@me.com; Toshi Abe - toshiabe@med.kurume-u.ac.jp; Motohiro Morioka - mmorioka@med.kurume-u.ac.jp

*Corresponding author

Received: 05 July 17  Accepted: 20 January 18  Published: 07 March 18

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

How to cite this article: Takeshige N, Eto T, Nakashima S, Sakata K, Uchikado H, Abe T, et al. Rare case of a rapidly enlarging symptomatic arachnoid cyst of the posterior fossa in an infant: A case report and review of the literature. Surg Neurol Int 2018;9:57.
http://surgicalneurologyint.com/Rare-case-of-a-rapidly-enlarging-symptomatic-arachnoid-cyst-of-the-posterior-fossa-in-an-infant:-A-case-report-and-review-of-the-literature/
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 intraventricularly (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%) 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.

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 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.
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. 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. 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.

The differential diagnosis of PFACs in infants mainly includes the Dandy–Walker malformation, mega cisterna magna, and Blake’s pouch cyst. It is also important to exclude other possible cystic lesions such as cerebellar cystic astrocytoma, cystic hemangioblastoma, and epidermoid or dermoid tumors. 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. 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. 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. 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.

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. To this
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. While some authors have reported that there is no significant difference between these surgical methods, studies evaluating the management of PFACs in a pediatric population demonstrated that craniotomy with excision was the first-line approach. 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. 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. 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. 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.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Al-Holou WN, Yew AY, Boomsma ZE, Garton HJL, Muraszko KM, Maher CO. Prevalence and natural history of arachnoid cysts in children. J Neurosurg Pediatr 2010;5:578-85.
2. Ali ZS, Lang SS, Bakar D, Storm PB, Stein SC. Pediatric intracranial arachnoid cysts: Comparative effectiveness of surgical treatment options. Childs Nerv Syst 2014;30:461-9.
3. Choi JW, Lee JY, Phi JH, Kim S-K, Wang KC. Stricter indications are recommended for fenestration surgery in intracranial arachnoid cysts of children. Childs Nerv Syst 2015;31:77-86.
4. Cincu R, Agrawal A, Eiras J. Intracranial arachnoid cysts: Current concepts and treatment alternatives. Clin Neurol Neurosurg 2007;109:837-43.
5. Collins CD, Hartley JC, Chakraborty A, Thompson DN. Long subcutaneous tunnelling reduces infection rates in paediatric external ventricular drains. Childs Nerv Syst 2014;30:1671-8.
6. Edinger P, Kaynar M, Bzdikus H, Qiplak N. Posterior fossa arachnoid cysts. Br J Neurosurg 1999;13:10-7.
7. Fulkerson DH, Vogel TD, Baker A, Patel N, Ackerman L, Smith JL, et al.

---

**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**

---

Table 1: Reported infant cases of enlarging symptomatic arachnoid cysts of the posterior fossa with surgical treatment
Cyst-ventricle stent as primary or salvage treatment for posterior fossa arachnoid cysts. J Neurosurg Pediatr 2011;7:549-56.

8. Gangemi M, Seneca V, Colella G, Ciolfi V, Imperato A, Maiuri F. Endoscopy versus microsurgical cyst excision and shunting for treating intracranial arachnoid cysts. J Neurosurg Pediatr 2011;8:158-64.

9. Gnanalingham KK, Hafuente J, Thompson D, Harkness W, Hayward R. Surgical procedures for posterior fossa tumors in children: Does craniotomy lead to fewer complications than cranioectomy. J Neurosurg 2002;97:821-6.

10. Halani H, Mina MS, Safain G, Heilman CB. Arachnoid cyst slit valves: the mechanism for arachnoid cyst enlargement. Report of 5 cases. J Neurosurg Pediatr 2013;12:62-6.

11. King AJ, Auguste KI, Halliday W, Drake JM, Kulkarni AV. Ventriculocystostomy and endoscopic third ventriculostomy/shunt placement in the management of hydrocephalus secondary to giant retrocerebellar cysts in infancy. J Neurosurg Pediatr 2010;5:403-7.

12. Kumagai M, Sakai N, Yamada H, Shinoda J, Nakashima T, Iwama T, et al. Postnatal development and enlargement of primary middle cranial fossa arachnoid cyst recognized on repeat CT scan. Childs Nerv Syst 1986;2:211-5.

13. Lee JY, Kim JW, Phi JH, Kim SK, Cho BK, Wang KC. Enlarging arachnoid cyst: A false alarm for infants. Childs Nerv Syst 2012;28:120311.

14. Marin-Sanabria EA, Yamamoto H, Nagashima T, Kohmura E. Evaluation of the management of arachnoid cyst of the posterior fossa in pediatric population: Experience over 27 years. Childs Nerv Syst 2007;23:535-42.

15. Matsumoto J, Koch M, Morioka M, Nakamura H, Makino K, Hamada J, et al. A long-term ventricular drainage for patients with germ cell tumors or medulloblastoma. Surg Neurol 2006;65:74-80.

16. Oberbauer RW, Haase J, Pucher R. Arachnoid cysts in children: A European co-operative study. Childs Nerv Syst 1992;8:281-6.

17. Parizek J, Mericka P, Nemecel S, Nemecelova J, Spacek J, Suba P, et al. Posterior cranial fossa surgery in 454 children. Comparison of results obtained in pre-CT and CT era and after various types of management of dura mater. Childs Nerv Syst 1998;14:426-38.

18. Raju S, Sharma RS, Meningi S, Momin J. Neuroendoscopy for Intracranial Arachnoid Cysts in Infants: Therapeutic Considerations. J Neurol Surg 2015;77:333-43.

19. Rao G, Anderson RCE, Feldstein NA, Brockmeyer DL. Expansion of arachnoid cysts in children. J Neurosurg 2015;102:314-7.

20. Sikorski CW, Yamani B, Frim DM. Endoscopic cyst fenestration outcomes in children one year of age or less. Pediatr Neurosurg 2004;40:59-63.

21. Srinivasan US, Lawrence R. Posterior fossa arachnoid cysts in adults: Surgical strategy: Case series. Asian J Neurosurg 2015;10:47-51.

22. Tsuboi Y, Hamada H, Hayashi N, Kurimoto M, Hirashima Y, Endo S. Huge arachnoid cyst in the posterior fossa: Controversial discussion for selection of the surgical approach. Childs Nerv Syst 2005;21:259-61.

23. Wilkinson CC, Wilson KR. Congenital arachnoid cyst and Dandy-Walker complex. In: Albright AL, Pollack IF, Adelson PD editors. Principles and Practice of Pediatric Neurosurgery. 2nd edition. Thieme: New York; 2008; pp 162-86.

24. Zada G, Krieger MD, McNatt S, Bowen I, McComb JG. Pathogenesis and treatment of intracranial arachnoid cysts in pediatric patients younger than 2 years of age. Neurosurg Focus 2007;22:E1.