Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

A Cyst in Disguise: Fourth Ventricular Arachnoid Cyst Masquerading as Congenital Hydrocephalus in Infancy: A Case Report

DOI: 10.70780/medpeer.000QGUH

AUTHOR AND AFFILIATION

Ibtissam El bqaq[1], Zaineb Ehirchiou[1], Lina Belkouchi[1], Nazik Allali[1], Latifa Chat[1], Siham El Haddad[1]

[1] Department of Radiology,Rabat Children's Hospital, Ibn Sina University Hospital Center,Rabat

Corresponding author: Ibtissam el bqaq, email: ibtissamelbqaq@gmail.com

ABSTRACT

Arachnoid cysts of the fourth ventricle are exceedingly rare, benign, cerebrospinal fluid–filled developmental lesions accounting for only a small fraction of intracranial arachnoid cysts, with occurrence in early infancy being particularly uncommon. We report the case of a 5-month-old infant with hydrocephalus present since birth, who presented with hypotonia and feeding refusal. Standard brain MRI identified a large, well-defined, thin-walled cystic formation of the fourth ventricle with regular margins, measuring approximately 67 x 64 x 77 mm (AP x H x T), hypointense on T1-weighted images, hyperintense on T2-weighted images, and suppressed on FLAIR sequences, without diffusion restriction or enhancement following contrast administration, responsible for upstream triventricular dilatation and mass effect on the adjacent cerebellum, pontocerebellar space, and brainstem. This case illustrates an unusual presentation and age of onset for this rare entity and discusses the differential diagnosis of posterior fossa cystic malformations in infancy, as well as the diagnostic and management considerations specific to this age group.

KEYWORDS

Arachnoid cyst, Fourth ventricle, Infant, Hypotonia, Posterior fossa

MAIN ARTICLE

INTRODUCTION

Arachnoid cysts are benign, congenital, cerebrospinal fluid–filled lesions arising from splitting or duplication of the arachnoid membrane, accounting for approximately 1% of intracranial space-occupying lesions [1,2]. They most commonly occur in the sylvian fissure, the suprasellar and quadrigeminal cisterns, and the cerebellopontine angle [2]. An intraventricular location is rare, and involvement of the fourth ventricle is particularly uncommon, with the first paediatric case reported only in 1979 [1]. Since then, only a limited number of cases have been described in the literature, the large majority in older children and adults [1]. When they occur in the fourth ventricle, arachnoid cysts typically present with signs of obstructive hydrocephalus and cerebellar or brainstem compression, most often described in older children and adults [1].

Presentation in early infancy is distinctly unusual, and clinical signs at this age are often nonspecific, including hypotonia, poor feeding, and delayed motor milestones, which can make the diagnosis challenging without dedicated neuroimaging. We report a case of fourth ventricular arachnoid cyst diagnosed in a 5-month-old infant with hydrocephalus present since birth, presenting with hypotonia and feeding refusal, and discuss its imaging features and differential diagnosis.

CASE REPORT

A 5-month-old infant, known to have hydrocephalus since birth, presented with hypotonia and refusal to feed, prompting neurological evaluation and brain MRI.

Standard brain MRI was performed, which identified a large, well-defined, thin-walled cystic formation of the fourth ventricle with regular margins, measuring approximately 67 x 64 x 77 mm (AP x H x T) (Figure 1). The lesion was hypointense on T1-weighted images, hyperintense on T2-weighted images, and showed signal suppression on FLAIR sequences, consistent with a cerebrospinal fluid–like content. There was no diffusion restriction and no enhancement following contrast administration (Figure 2).

This cystic formation was responsible for upstream triventricular dilatation (lateral and third ventricles), consistent with obstructive hydrocephalus secondary to fourth ventricular outlet obstruction. A mass effect was noted on the adjacent structures, namely the cerebellum, the pontocerebellar space, and the entire brainstem.

On examination, the infant displayed hypotonia, hyporeflexia, macrocephaly, and a "setting sun" sign (downward deviation of the eyes with visible sclera above the iris), findings consistent with raised intracranial pressure in the context of the underlying hydrocephalus.

Given the size of the lesion, the degree of upstream ventricular dilatation, and the associated mass effect on the brainstem and cerebellum, the infant was referred to a specialised paediatric neurosurgical centre for further evaluation and management.

DISCUSSION

The pathophysiology of fourth ventricular arachnoid cysts remains incompletely understood. The most widely accepted hypothesis proposes entrapment of arachnoid membrane within the choroid plexus mesenchyme during embryonic development [2], while an alternative theory suggests extension of a cyst from the supracerebellar or cisternal subarachnoid space into the ventricle through the choroidal fissure [3]. Regardless of the precise mechanism, arachnoid cysts are lined by a thin membrane of arachnoid cells and contain fluid essentially identical to cerebrospinal fluid, without free communication with the surrounding subarachnoid space [1].

Clinically, the presence of a fourth ventricular arachnoid cyst can obstruct cerebrospinal fluid outflow through the foramina of Luschka and Magendie, producing obstructive hydrocephalus, and can exert direct mass effect on the cerebellum and brainstem [1]. Most reported cases present with signs of raised intracranial pressure, truncal ataxia, or cerebellar dysfunction, typically in older children or adults [1]; presentation in early infancy, as in the present case, is distinctly rare. A comparable case was reported by Takeshige et al., describing a rapidly enlarging posterior fossa arachnoid cyst in an infant who developed progressive macrocrania and obstructive hydrocephalus by four months of age, ultimately requiring surgical fenestration [4]. With a largest diameter of 77 mm, the cyst in our patient was considerably larger than the 40 x 35 mm lesion reported in that case, suggesting that the volume attained by symptomatic fourth ventricular arachnoid cysts in infancy may be more variable than previously described. In infants, clinical signs of a posterior fossa cystic lesion are often nonspecific — hypotonia, poor feeding, irritability, or delayed motor development — and may precede more overt signs such as macrocephaly or a bulging fontanelle, underscoring the value of early neuroimaging in infants presenting with unexplained hypotonia or neurological signs.

Differentiating a fourth ventricular arachnoid cyst from its principal mimics is critical, as management and prognosis differ substantially between entities; phase-contrast cine MRI for cerebrospinal fluid flow evaluation has been shown to improve diagnostic specificity in this setting, as each posterior fossa cystic malformation displays a distinct flow pattern [5]. Mega cisterna magna, the most common differential diagnosis, is defined by an enlarged cisterna magna (>10 mm on midsagittal imaging) with an intact cerebellar vermis, a normal fourth ventricle, and free communication with the surrounding subarachnoid space, without mass effect [6]; the presence of a visible falx cerebelli and normal posterior dural folds favours this diagnosis over an arachnoid cyst [7]. A persistent Blake's pouch cyst presents as an infravermian cyst communicating with the fourth ventricle, typically associated with hydrocephalus but without cerebellar hypoplasia [6]. Dandy-Walker malformation, by contrast, is characterised by vermian hypoplasia or agenesis, torcular-lambdoid inversion, and a posterior fossa cyst in direct continuity with a dilated fourth ventricle [8]. While arachnoid cysts, mega cisterna magna, and Blake's pouch cysts share an identical cerebrospinal fluid signal on conventional sequences, arachnoid cysts are distinguished by their mass effect — including scalloping of the adjacent calvarium — and by restricted, often absent, internal fluid motion [6,7].

Management options for symptomatic fourth ventricular arachnoid cysts include cerebrospinal fluid diversion, endoscopic or open cyst fenestration, and, in select cases, third ventriculostomy. Shunting procedures alone have been reported to provide inadequate long-term symptom control in several series [1], with definitive treatment more often achieved through direct fenestration or resection of the cyst wall to restore physiological cerebrospinal fluid pathways [1].

The prognosis of surgically managed fourth ventricular arachnoid cysts is generally favourable when treatment is undertaken promptly. In the case reported by Takeshige et al., surgical fenestration of an enlarging posterior fossa arachnoid cyst in an infant resulted in resolution of hydrocephalus and a decompressed fourth ventricle at follow-up [4]. Similarly, direct surgical resection has been associated with uneventful recovery and sustained symptom resolution at one-year follow-up in a paediatric case [2]. Given the referral of the present infant to a specialised neurosurgical centre in view of the significant ventricular dilatation and brainstem compression identified on imaging, a comparable favourable outcome may be anticipated with timely surgical management, underscoring the importance of prompt recognition and referral once the diagnosis is suspected on imaging.

CONCLUSION

This case illustrates a rare presentation of a fourth ventricular arachnoid cyst manifesting with congenital hydrocephalus, hypotonia, and feeding refusal in early infancy, a presentation and age group only exceptionally described in the literature. It underscores the importance of considering posterior fossa cystic lesions in infants presenting with unexplained hypotonia and highlights MRI as the key diagnostic tool for their characterisation and differentiation from other posterior fossa malformations.

FIGURES :

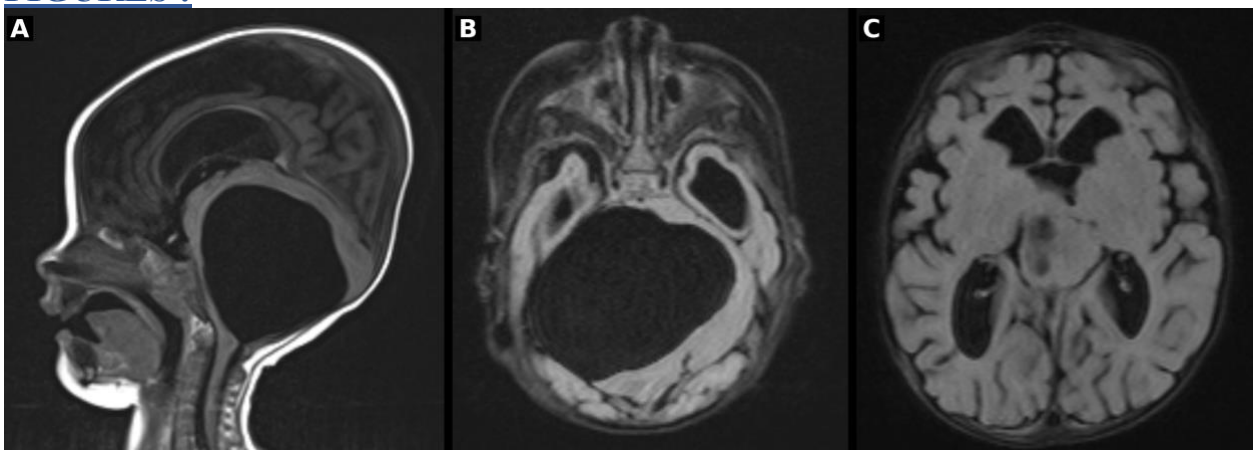

Figure 1. Brain MRI (T1-weighted sequences) of the 5-month-old infant showing a large, well-defined, thin-walled cystic formation of the fourth ventricle. (A) Midsagittal T1-weighted image demonstrating the hypointense cystic lesion occupying the posterior fossa, with mass effect on the cerebellum and brainstem and upstream dilatation of the third ventricle. (B) Axial T1-weighted image at the level of the posterior fossa showing the cystic formation with regular margins, exerting mass effect on the adjacent cerebellar and pontocerebellar structures. (C) Axial T1-weighted image at the level of the lateral ventricles showing triventricular dilatation, consistent with obstructive hydrocephalus secondary to fourth ventricular outlet obstruction.

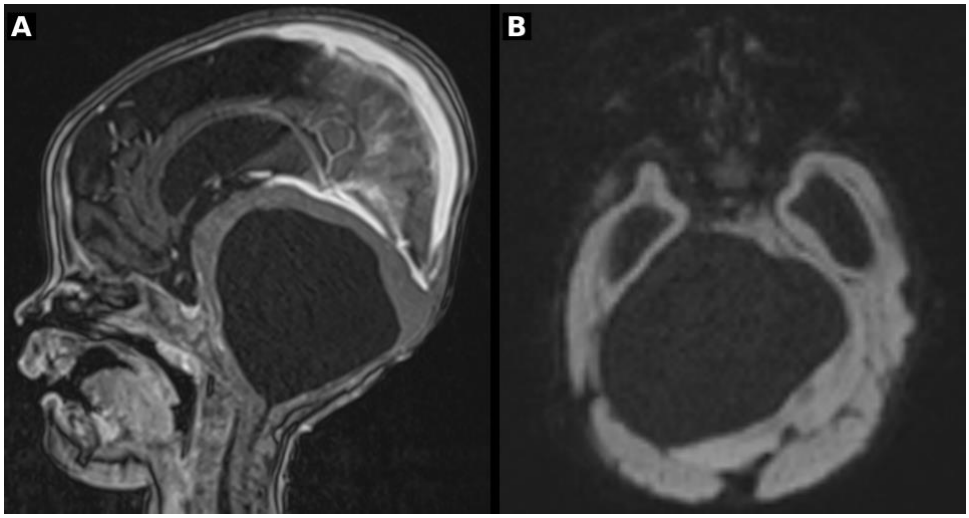

Figure 2. Additional MRI sequences of the same infant. (A) Midsagittal T1-weighted image following gadolinium administration, showing no abnormal enhancement of the cyst wall or content, in keeping with the benign nature of the lesion. (B) Axial diffusion-weighted image (DWI) at the level of the posterior fossa showing no diffusion restriction within the cystic lesion, consistent with cerebrospinal fluid content and helping exclude an epidermoid cyst.

ACKNOWLEDGEMENTS

None.

Conflict of Interest: The authors declare that they have no conflicts of interest to disclose.

Funding: This case report received no specific grant or financial support from any funding agency in the public, commercial, or not-for-profit sectors.

Ethical Approval: Formal ethical approval was not required for this case report in accordance with institutional and journal policy, as it did not involve any deviation from standard clinical care.

Consent: Written informed consent was obtained from the patient's legal guardian(s) for publication of this case report and the accompanying images.

REFERENCES

1. Westermaier T, Vince GH, Meinhardt M, Monoranu C, Roosen K, Matthies C. Arachnoid cysts of the fourth ventricle: short illustrated review. Acta Neurochir (Wien). 2010;152(1):119-124. doi:10.1007/s00701-009-0309-2
https://doi.org/10.1007/s00701-009-0309-2

2. Kanjilal S, Maurya VP, Das KK, Verma PK, Jaiswal AK, Pal L, Behari S. Intra-fourth ventricular arachnoid cyst: the case for a direct surgical approach. J Pediatr Neurosci. 2022;17(2):148-154. doi:10.4103/jpn.JPN_106_21
https://doi.org/10.4103/jpn.JPN_106_21

3. Park SW, Yoon SH, Cho KH, Shin YS. A large arachnoid cyst of the lateral ventricle extending from the supracerebellar cistern - case report. Surg Neurol. 2006;65(6):611-614. doi:10.1016/j.surneu.2005.07.069
https://doi.org/10.1016/j.surneu.2005.07.069

4. Takeshige N, Eto T, Nakashima S, Sakata K, Uchikado H, Abe T, Morioka M. 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. doi:10.4103/sni.sni_245_17
https://doi.org/10.4103/sni.sni_245_17

5. Yildiz H, Yazici Z, Hakyemez B, Erdogan C, Parlak M. Evaluation of CSF flow patterns of posterior fossa cystic malformations using CSF flow MR imaging. Neuroradiology. 2006;48(9):595-605. doi:10.1007/s00234-006-0098-8
https://doi.org/10.1007/s00234-006-0098-8

6. Weisbrod LJ, Thorell W. Mega Cisterna Magna. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2023.

7. Regmi PR, Amatya I. CSF flow study in differentiation between the arachnoid cyst and mega cisterna magna. Eurorad. 2020. Case 16736. doi:10.35100/eurorad/case.16736

8. Karsonovich T, Das JM, Ahmad T. Dandy-Walker Malformation. StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2026.