Diagnosis and Management of Complex Cases

ASSOCIATION OF A VERMIAN DERMOID CYST AND A CEREBELLOPONTINE ANGLE ARACHNOID CYST IN A CHILD: A Case Report and Literature Review

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Received: *September05, 2025 —* ***Accepted:****December 30, 2025 —* ***Published:*** *January 30, 2026*

Citation : BEKRALAS Houria. Association of a Vermian Dermoid Cyst and a Cerebellopontine Angle Arachnoid Cyst in a Child: A Case Report and Literature Review. ***OLCIAS Vol.3, Issue 1***

ABSTRACT :

We report an exceptional case of an association between a vermian dermoid cyst and a cerebellopontine angle arachnoid cyst. The patient is a 5-year-old girl presenting with intracranial hypertension syndrome, static cerebellar syndrome, and lateral deviation of the eyes. CT and MRI investigations revealed a vermian dermoid cyst, a compressive cerebellopontine angle arachnoid cyst, and active triventricular hydrocephalus. The patient underwent neurosurgical management in three stages: ventriculoperitoneal shunt as a first step, surgical approach to the dermoid cyst as a second step, and surgical treatment of the arachnoid cyst as a third step. Postoperative clinical outcome was satisfactory. Follow-up imaging confirmed total removal of the dermoid cyst and collapse of the arachnoid cyst. The outcome at 3 years was satisfactory.

Keywords: *arachnoid cyst; dermoid cyst; hydrocephalus; posterior cranial fossa.*

INTRODUCTION

An arachnoid cyst is a cystic lesion whose wall is arachnoid and whose content is cerebrospinal fluid (CSF). [10, 11, 12] Most authors agree on its congenital origin, resulting from abnormal development of arachnoid tissue. [10, 11]

Symptomatic arachnoid cysts represent about 1% of intracranial space-occupying lesions,

with children accounting for 60% to 90% of cases. [9, 10, 11, 13]

A dermoid cyst is a rare benign tumor of the central nervous system, representing 0.1% to 0.7% of all intracranial lesions. [2, 3, 5, 6, 7, 8, 14, 16] It consists of a thick wall lined by keratinizing stratified squamous epithelium, and its content is composed of dermal elements such as hair, teeth, and sebaceous glands. [2, 3, 5] It is a congenital tumor due to an abnormality of development between the third and fifth week of embryonic life, and its clinical presentation most often occurs in adolescents. [5, 14]

The association of an arachnoid cyst with a dermoid cyst is very rare. We found only one case described in the literature, reported by Chang WH in 1989 [1], consisting of a middle fossa arachnoid cyst associated with a suprasellar dermoid cyst.

We report a very rare case of a cerebellopontine angle arachnoid cyst associated with a vermian dermoid cyst. [3, 9, 14, 16]

CASE REPORT

This is a 5-year-old girl born at term, the fourth child in a family of four, admitted with intracranial hypertension manifested by somnolence and repeated vomiting. Clinical examination showed mild macrocephaly (unusual in the family) and a static cerebellar syndrome with an unsteady gait. History revealed mild but daily headaches for six months, relieved by analgesics. An emergency brain CT scan showed two cystic lesions of the posterior fossa with active triventricular hydrocephalus (Figure 1).

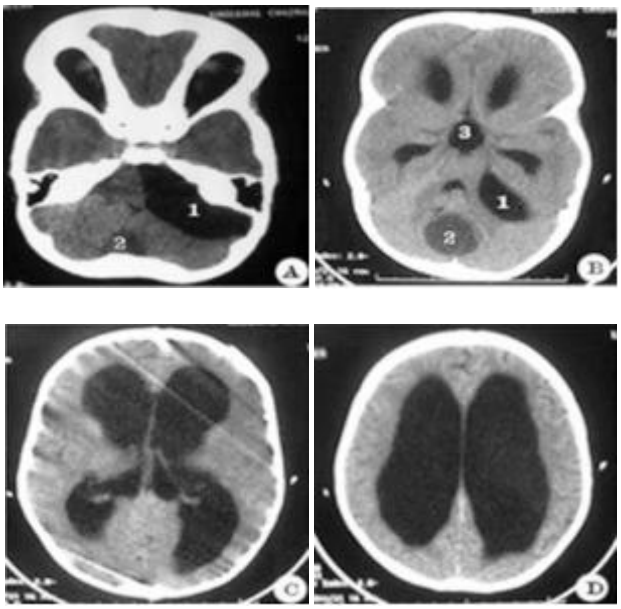

Figure 01: Axial **non-contrast** brain CT scan showing a **left cerebellopontine angle (CPA)**

arachnoid cyst (1) (**a, b**), a **vermian dermoid cyst** (2) (**a, b**), **dilatation of the third ventricle** (3) (**b**), and **dilatation of the lateral ventricles** (**c, d**)

Because of somnolence, an emergency ventriculoperitoneal shunt was placed, leading to normalization of consciousness. A subsequent brain MRI characterized a cerebellopontine angle arachnoid cyst displacing the brainstem, as well as a vermian dermoid cyst (Figure 2).

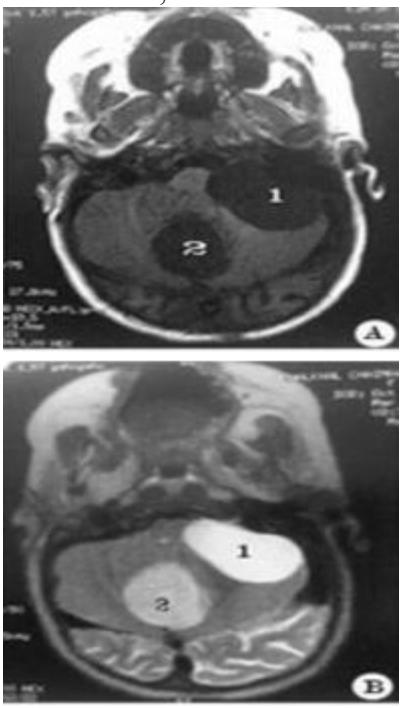

Figure 02: Axial brain MRI performed in **T1-weighted** (a) and **T2-weighted** (b) sequences: a **left cerebellopontine region arachnoid cyst** (1) showing **hypointense signal on T1** (a) and **hyperintense signal on T2** (b), and a **vermian dermoid cyst** (2) showing **hypointense signal on T1** (a) and **slightly hyperintense signal on T2** (b).

A second surgical procedure was performed consisting of total excision of the vermian dermoid cyst through a midline suboccipital craniectomy (Figure 3). The unsteady gait resolved completely.

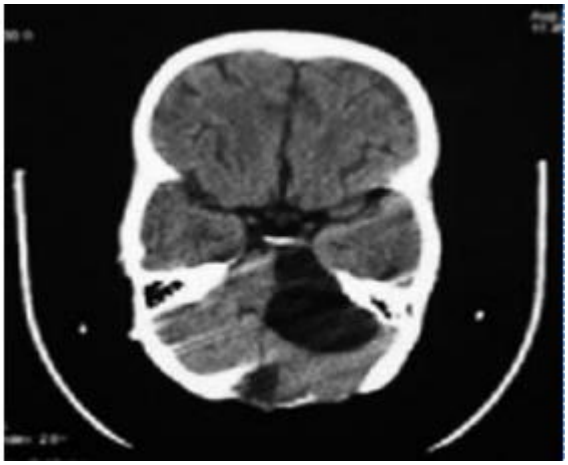

Figure 3: Postoperative brain CT scan showing complete excision of the dermoid cyst via a midline suboccipital approach, and the cerebellopontine angle (CPA) arachnoid cyst, which has not yet been operated on.

Three months later, the child developed new symptoms with lateral deviation of the eyes to the right, related to brainstem compromise due to increased mass effect from the left cerebellopontine angle arachnoid cyst. She was readmitted to the operating room where a retrosigmoid craniectomy was performed with marsupialization of the arachnoid cyst: partial resection of the outer wall with communication to the cisterna magna, and fenestration of the inner wall with communication to the prepontine and prebulbar cisterns.
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Postoperative course was marked by disappearance of ocular deviation. Follow-up imaging at six months showed a reduction in the size of the arachnoid cyst (Figure 4)

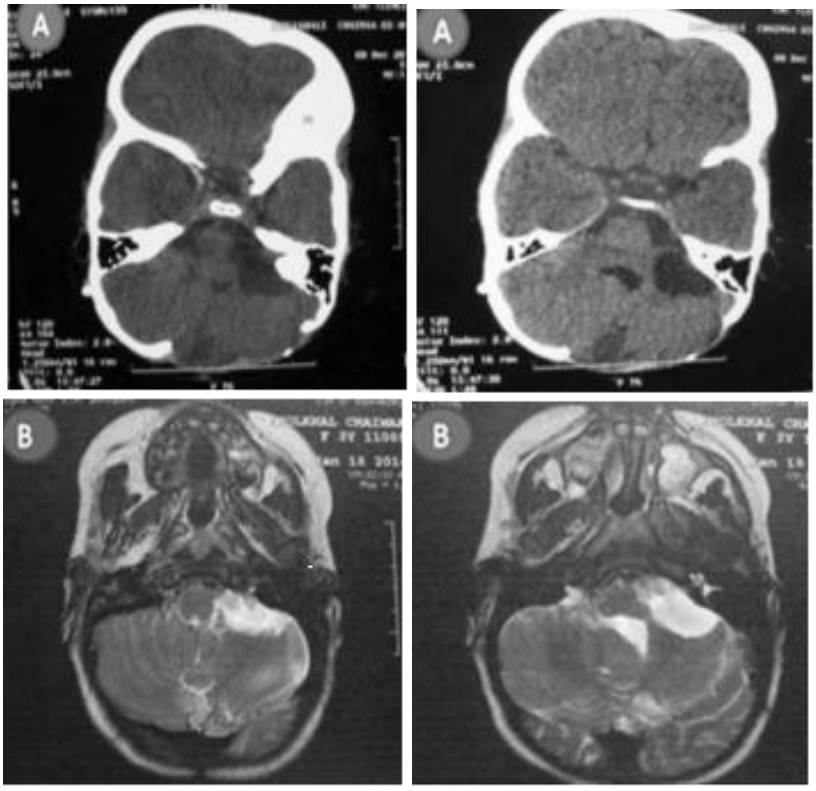

Figure 4: Postoperative brain imaging—brain CT scan (Fig. 4a) and brain MRI (Fig. 4b)—showing collapse of the cerebellopontine angle arachnoid cyst and complete excision of the vermian dermoid cyst.

DISCUSSION

PlSeveral theories have been proposed to explain the etiopathogenesis of arachnoid cysts, but the embryological theory is the most widely accepted. The subarachnoid space between the pia mater and arachnoid forms from the fourth month of intrauterine life, resulting from hydraulic dissection by CSF produced by the primitive ventricular system. [10, 11, 12] At this embryonic stage, the arachnoid is not well differentiated, and hydraulic dissection may occur between the two layers of the arachnoid, resulting in an arachnoid cyst. [10]

For dermoid cysts, an embryological cause is also accepted: they result from a developmental anomaly between the fifth and sixth week of gestation, due to inclusion of ectodermal elements within the neural tube following defective closure. [2, 3, 5, 6, 7, 8, 14, 16]

In our case, the association of two embryological developmental anomalies raises the question of whether a common etiological factor may underlie both. The case described by Chang WH in 1989 supports this hypothesis. [1]

Arachnoid cysts represent 1% of intracranial lesions; the sylvian location is the most frequent

(50% of cases). In 8% of cases they are located at the cerebellopontine angle. [9, 10, 11, 13]

Dermoid cysts represent 0.1% to 0.7% of intracranial lesions and tend to occur on or near the midline. [3, 5, 14, 16]

In the dermoid cyst–arachnoid cyst association described by Chang WH in 1989, as in our case, the two lesions were adjacent. In Chang's case, the arachnoid cyst was in the middle cranial fossa and the dermoid cyst was suprasellar. In our opinion, this proximity further supports the existence of a common etiological factor responsible for an embryogenesis disorder leading to both dermoid and arachnoid cysts. [1]

A symptomatic cerebellopontine angle arachnoid cyst generally presents with signs of intracranial hypertension, cerebellar symptoms, and cranial nerve involvement: V (trigeminal neuralgia), VI (diplopia), VII (hemifacial spasm or facial palsy), VIII (hearing loss, tinnitus, vertigo). Vermian dermoid cysts generally present with intracranial hypertension and cerebellar syndrome. Our patient presented with intracranial hypertension, unsteady gait, and rightward lateral deviation of the eyes. [9, 13, 3, 14, 16]

From a therapeutic standpoint, several options exist:

For arachnoid cysts, three surgical techniques are described:
a) Resection of the cyst wall requiring open surgery. [10]
b) Fenestration of the cyst wall, either endoscopic or microsurgical. [10, 11]
c) Cystoperitoneal or cystosubdural shunting. [10]

In cerebellopontine angle arachnoid cysts, the presence of intracystic septations, the proximity of delicate neural structures, and the high recurrence rate after fenestration support open microsurgical resection of the cyst wall. Bulent [10] noted that for infratentorial arachnoid cysts, size reduction is achieved only after wide excision of the wall under the operating microscope. In our patient, partial resection of the outer wall combined with fenestration of the inner wall was performed.

Dermoid cysts are composed of a thick cyst wall lined by stratified squamous epithelium, with contents including dermal elements, fat cells, hair, and sebaceous glands. Total microsurgical excision is preferred to avoid recurrence. [5, 6, 14, 15] However, when the cyst wall is firmly adherent to a sensitive neural structure, small capsular fragments may be left in place after careful coagulation; these fragments evolve slowly and do not require immediate additional treatment. [14, 16] Our patient had a vermian dermoid cyst with no relationship to the floor of the fourth ventricle, allowing total excision.

In this patient, the adopted therapeutic strategy consisted of an emergency ventriculoperitoneal shunt as a first step, followed by surgery for the dermoid cyst as a second step, and finally surgery for the arachnoid cyst. [1, 9, 10, 16] This chronology was based on clinical presentation: altered consciousness due to active hydrocephalus justified an

emergency ventriculoperitoneal shunt; static cerebellar syndrome due to the dermoid cyst justified second-stage surgery; and subsequent ocular deviation due to brainstem compression by the arachnoid cyst justified the third-stage operation.

CONCLUSION

Both arachnoid cysts and dermoid cysts are rare conditions; their association in the posterior cranial fossa is therefore exceptional. The simultaneous and adjacent presence of these two lesions raises the issue of a possible common factor responsible for an embryonic developmental disorder, affecting both the neural tube (leading to the dermoid cyst) and the subarachnoid spaces (leading to the arachnoid cyst).

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