Case Report

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Bilateral Cerebello-Pontine Angle Arachnoid Cyst in a Newborn with Facial and Auditory Nerve Paresis

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Abstract

Background: Arachnoid cysts (AC) are benign, congenital, non-neoplastic, extra axial, intra-arachnoid lesions filled with cerebrospinal fluid (CSF). Only 10% of arachnoid cysts occur in Cerebello-Pontine Angle (CPA). Bilateral ACs are very rare. *Case Description:* In this case report we present a one month baby who was evaluated for failure to thrive and hydrocephalus. Patient examination revealed cranial nerve seven and eight paresis and imaging was suggestive of bilateral CPA ACs which was treated with surgery. *Literature Review:* Pubmed search showed only three cases of bilateral ACs -one in an adolescent and two in adult population. *Clinical Relevance:* This is probably first reported case of bilateral CPA AC in a newborn with facial paresis.

Keywords: Arachnoid cysts (AC); Cerebrospinal fluid (CSF); Cerebellopontine angle (CPA).

Introduction

Arachnoid cysts (AC) are benign, congenital, non-neoplastic, extra axial, intra-arachnoid lesions filled with cerebrospinal fluid (CSF)[1,2]. They are postulated to occur due to splitting of embryonic meninges and thus are filled with CSF [10].

These constitute about 1% of all nontraumatic intracranial space-occupying lesions. Only 10% of all AC occur in the Cerebello-Pontine Angle (CPA) and 60% to 80% of AC are diagnosed in children [5,8]. Bilateral CPA AC are very rare, and only three cases have been reported till date – one in an adolescent and 2 in adults [7]. In this article we report a case of newborn with bilateral CPA AC with unilateral facial paresis and hydrocephalus. This is probably the first reported case of bilateral CPA AC in a newborn with associated facial paresis.

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Case Report

Our patient, an one month old had an abnormally increasing head circumference while on a regular follow up with the department of paediatrics for failure to thrive and recurrent episodes of upper respiratory tract infections. The child had born prematurely at 33 weeks after gestation with a birth weight of 1.8kg. Subsequently he also had transient tachyapnoea of new born (TTN) and neonatal sepsis which required neonatal intensive care management. Examination revealed bulging and enlarged anterior fontanelle, sutural diastasis, left infranuclear facial paresis **(Figure 1)** and reduced click evoked acoustic responses. Further systemic evaluation showed presence of atrial septal defect (left to right shunt) and an inguinal hernia.

Magnetic Resonance Imaging (MRI) brain in its T2 sequence showed bilateral CPA extra axial hyperintense lesions which followed CSF densities on T2 **(Figure 2)** and fluid attenuated inversion recovery (FLAIR) imaging compressing the fourth ventricle and causing hydrocephalus **(Figure 3)**.

In view of significant hydrocephalus due to obstruction of the fourth ventricle and infranuclear facial nerve paresis, midline sub- occipital craniotomy and fenestration of bilateral AC was performed. The child showed improvement of the facial paresis and

feeding post operatively but anterior fontanelle remained full. Post operative imaging demonstrated adequate decompression of both CPA AC but dilation of ventricular system persisted **(Figure 5)**. Two weeks post surgery, child had features of sun setting with tense anterior fontanelle. Computerised tomography (CT) evaluation showed persistence of hydrocephalus despite resolution of compression over the fourth ventricle. Hence a ventriculoperitoneal (VP) shunt was performed. The subsequent post operative period was uneventful.

At follow up after one year, the child had adequate weight gain for his age and remained asymptomatic. The shunt was functioning; the facial paresis had improved and the developmental milestone were normal for the age.

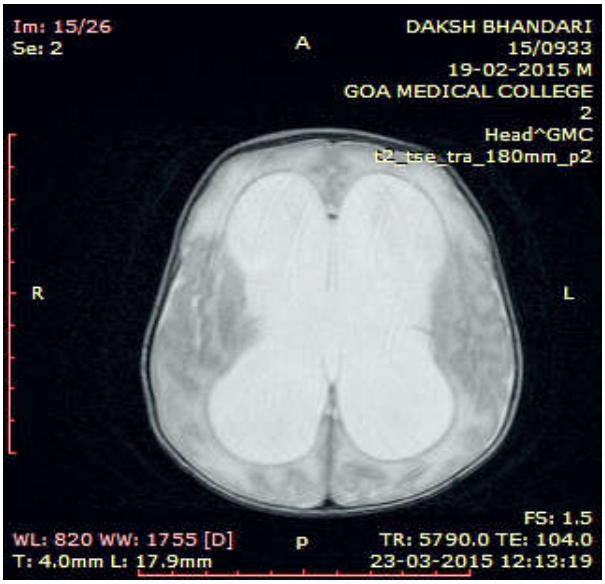

Fig. 3: MRI axial T2 showing hydrocephalus

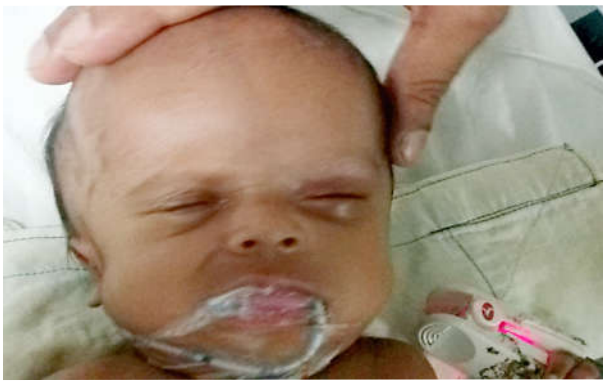

Fig. 1: Reduced wrinkling of fore head , eye and facial muscles on the left side

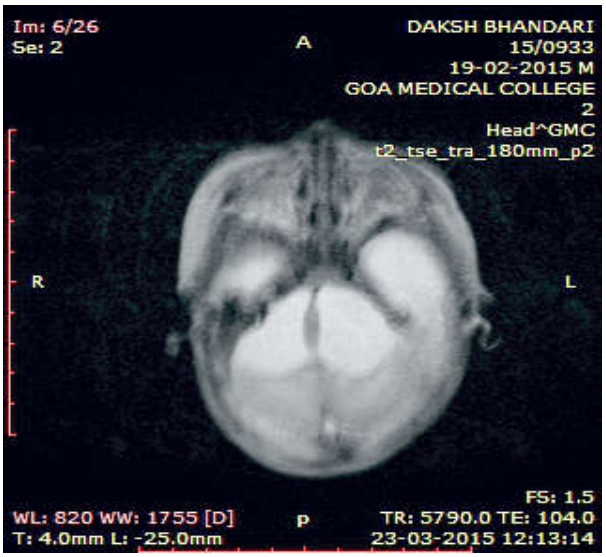

Fig. 2: MRI brain T2 axial showing bilateral CPA AC compressing the brainstem

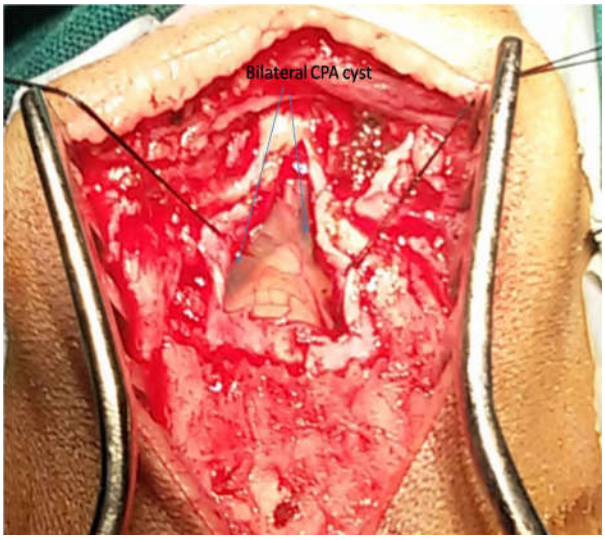

Fig. 4: Bilateral CPA cyst

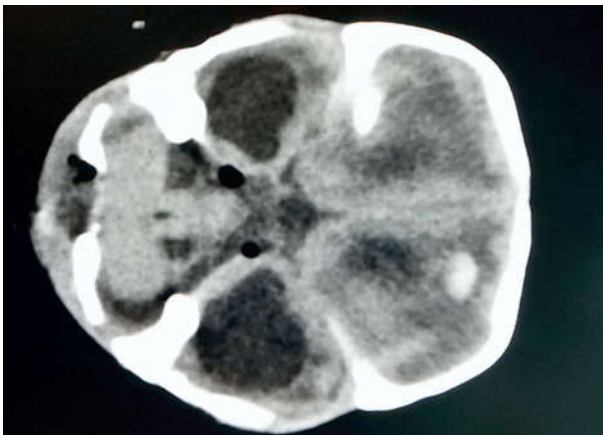

Fig. 5: Computerised tomography of brain showing decompressed aracgnoid cyst bilaterally with opening up of fourth ventricle

Discussion

Primary AC is a congenital cyst lined by a single layer of flattened, arachnoid cells in a vascular collagenous membrane that lies entirely within the arachnoid layer. The cysts may be loculated, compartmentalized, or freely communicating with the surrounding CSF cisterns. Secondary AC develops as a result of post inflammatory accumulation of CSF in subarachnoid space in patients with head injury, haemorrhage or infection. The distinguishing features of the arachnoid cyst wall versus a normal arachnoid membrane include the split of the arachnoid layer at the margin of the cyst, the increased thickness of the collagen layer, and the absence of the cobweb-like trabeculations of normal arachnoid. The cyst fluid is clear and devoid of cellular or proteinaceous material. Several mechanisms have been accounted for enlargement of cysts like secretion by the cells forming the cyst wall and the presence of an unidirectional valve effect for fluid movements secondary to pulsations of veins [3].

They are mostly diagnosed incidentally on radiological imaging and are frequently asymptomatic. They cause symptoms when they become sufficiently large to compress the adjacent brain structures like cranial nerves and CSF pathways like in our patient.

The occurrence of cysts in neonates and siblings, the anatomic relationship of cysts to the cisterns, and the association of AC with other developmental anomalies all lend credence to the supposition that these cysts are usually developmental in origin and not acquired from other pathologic conditions. Associated anomalies are rare, the most common being hydrocephalus. Arachnoid cysts may coexist with other genetic disturbances in collagen formation like Marfan syndrome, Down's syndrome and neurofibromatosis type I [2].

There is a male preponderance and increased incidence on the left side with no plausible explanation for the same at present.

There have been reports in literature of unilateral CPA-AC with ipsilateral sixth, seventh or eighth cranial nerve paresis [4,6].

In our case report, the one month old male child had left facial and auditory nerve paresis, failure to thrive and an enlarged head circumference. MRI imaging showed hydrocephalus with bilateral CPA-AC which is probably the first reported case in literature in a newborn. The clinical condition improved after bilateral marsupialization of the cysts followed by a ventriculo-peritoneal shunt. Communicating hydrocephalus secondary to neonatal sepsis can be the possible explanation for persistence of hydrocephalus even after adequate surgical treatment.

The management of arachnoid cysts of the cerebellopontine angle remains controversial. Asymptomatic arachnoid cysts do not require treatment, and such patients should be monitored clinically and radiologically with serial MRIs. Surgical treatment for symptomatic patients consists of resection/ fenestration of cyst into subarachnoid spaces or cystoperitoneal shunting. Recently endoscopic cyst decompression has also been shown to be safe and effective [9].

Conclusion

ACs are being increasingly diagnosed in infancy with ready availability of modern investigative modalities. Their management often poses a dilemma and surgical intervention should be considered in selected patients in the presence of mas effect. The occurrence of cranial nerve palsy in the vicinity of the AC and/ or hydrocephalus is an indication for surgical intervention for a CP-AC.

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