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Vertex encephalocele: Rare case report

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ABSTRACT

Introduction– Encephalocele is a mesodermal defect in the skull bones and duramater. Vertex encephalocele is a rare congenital anomaly of newborn with variable prognostic value. We report a case of a vertex encephalocele with no associated other system malformations..

Case description: A unique case of a 25 day neonate, with swelling over scalp was evaluated by the neurosurgical team and the patient underwent neurosurgery. Excision and repair of defect was done. Child stood surgery well and post operatively child could move all four limbs without any gross neurological deficit. Post-operative recovery was uneventful.

Conclusion: In planning the strategy for management of encephalocele, one needs to take into consideration the site, size, contents, patency of CSF pathway, neurological status and other associated anomalies. Inspite of such a big encephalocele in an atypical location, excision and repair gave excellent results.

Keywords: Congenital anomaly, interparietal encephalocele, Vertex encephalocele

INTRODUCTION

A vertex cephalocele, also known as a midline parietal cephalocele, is a congenital condition where the meningeal layers protrude through a skull defect in the vertex, or top, of the head. This anomaly involves the herniation of meningeal tissues, which typically include the membranes surrounding the brain, through a skull defect and is covered by skin.

Although vertex cephaloceles may seem innocuous externally, it is crucial to recognize that they can be associated with more intricate issues within the skull. These cephaloceles may also contain abnormal blood vessels (anomalous vessels) and components of the nervous system (neural elements).

A significant concern with vertex cephaloceles is the potential presence of underlying complications such as intracranial venous and/or brain malformations. These complexities may pose a greater risk than what is apparent from the external appearance of the cephalocele, suggesting that the visible part is just the "tip of the iceberg."

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Because of the potential for underlying issues, individuals with vertex cephaloceles often require thorough medical evaluation and management. Imaging studies, such as magnetic resonance imaging (MRI) or computed tomography (CT) scans, may be employed to assess the extent of the malformation and identify any associated intracranial abnormalities. Treatment strategies may involve surgical intervention to address the skull defect and manage any underlying issues within the skull, with the specific approach tailored to each case's unique characteristics and severity of malformations.

CASE DESCRIPTION

A neonate, 25-day-old female baby presented with congenital scalp swelling and was evaluated. Her mother belonged to a remote area where no prenatal ultrasonography was available; baby was delivered at a district hospital through LSCS. Scalp swelling progressively increased since birth.

Complete physical examination of the baby revealed a congenital scalp swelling with no other associated congenital anomalies such as spine defect, limb defect; vertebral anal anomalies, trachea oesophageal anomalies, cardiac anomalies and renal anomalies. There was no hypertelorism or proptosis and fundus examination had no early signs of hydrocephalous.

Two swelling of the size 3 x 3 cm adjacent to each other was located in midline, midway between frontal and occipital region. Size of the head as per age was normal with no hydrocephalous and other congenital anomaly. Detailed neurological examination and other systemic examination revealed no significant associated anomalies. Swelling was filled by aberrant brain tissue.

3D CT SCAN head showed a focal defect involving high parietal bone with herniation of neural tissue through this defect suggestive of encephalocele.

Figure 1:Patient with scalp swelling

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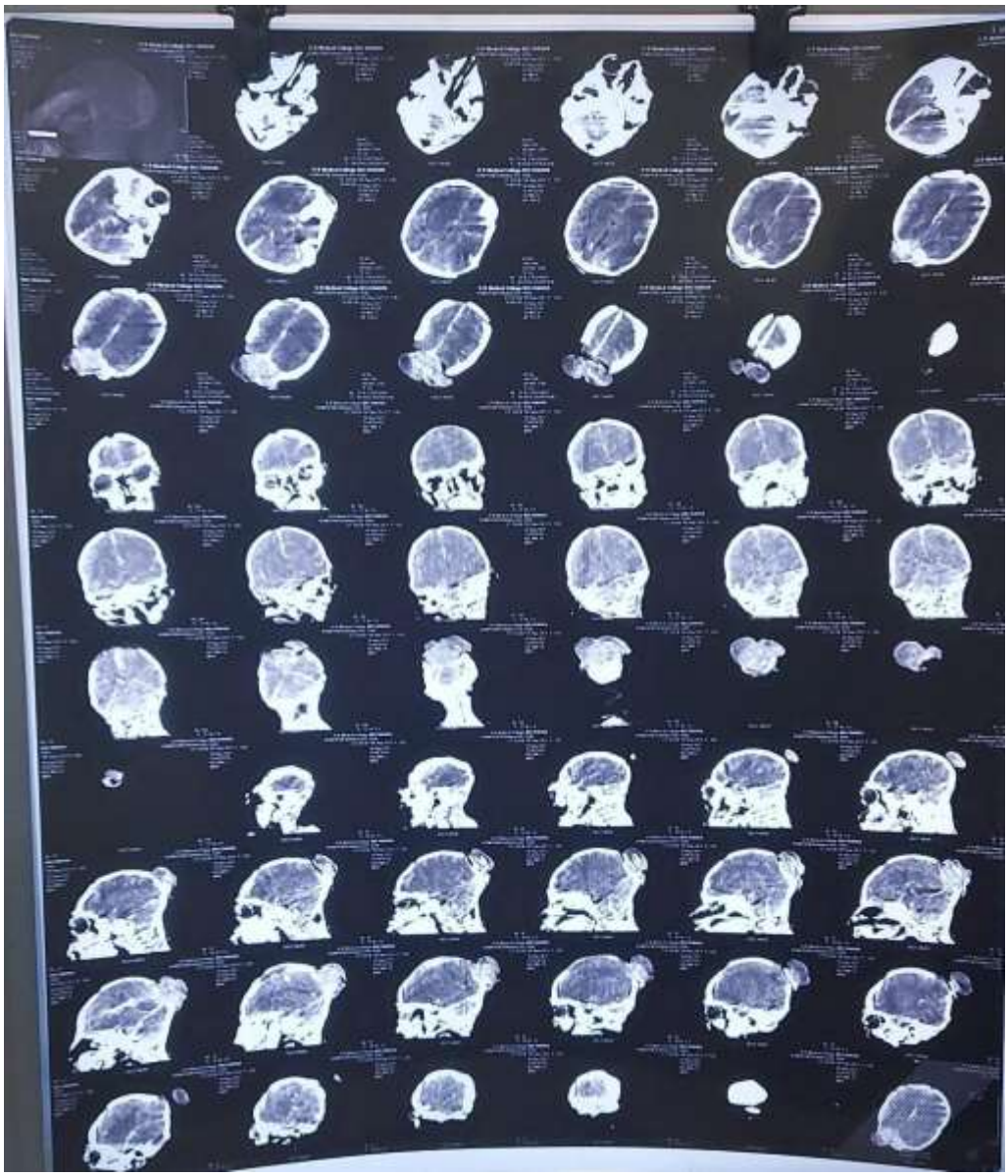
Figure 2: 3D CT RECONSTRUCTION OF BRAIN



Excision and **r**epair of the defect was done under general anaesthesia with prone position. Elliptical incision encircling swelling was made and duramater was defined all around the sac. Excision and Dural defect was meticulously repaired, to get a watertight closure.

Child stood surgery well and post operatively child could move all four limbs without any gross neurological deficit. No ventilator support was needed post operatively. Post-operative recovery was uneventful.

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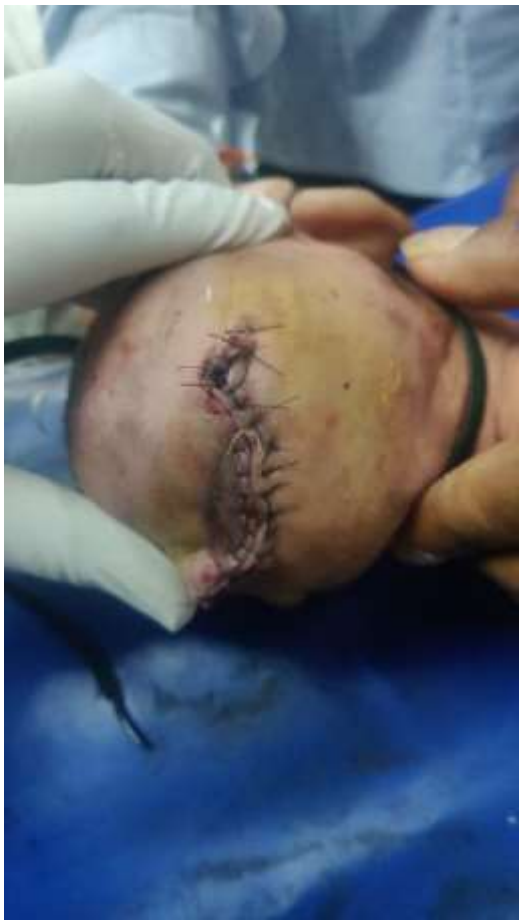
Figure 3: POST OPERATIVE



DISCUSSION

Encephaloceles typically manifest in the occipital and anterior regions, with occurrences at mid-interparietal locations being exceptionally rare [1]. Within vertex or midline parietal encephaloceles, the atretic form is more commonly observed compared to the occurrence of large-sized encephaloceles. [2]. The literature contains substantial case series focusing on both anterior encephaloceles and occipital encephaloceles, contributing to a comprehensive understanding of these specific manifestations. [3,4]. However, large encephaloceles located in midline interparietal region are limited [5-8]. The management of encephaloceles is contingent upon factors such as the type, size, and the presence of associated hydrocephalus. Surgical intervention typically involves excising the sac and repairing the dural defect, with a focus on reintegrating viable, healthy brain tissue into the cranial cavity. It is important to note that the ultimate outcome of encephalocele surgery is often influenced more by the underlying brain involvement and the presence or absence of other congenital defects. [9] Postoperative complications following encephalocele repair may include the development of hydrocephalus, which might not be evident before the surgery but can manifest after the encephalocele repair. Notably, giant occipital encephaloceles tend to be linked with a higher prevalence of underlying malformations and often carry a less favorable prognosis [10].

CONCLUSION

Interparietal encephaloceles are considered rare among cases of encephaloceles. To diagnose underlying brain malformations, a CT image morphology is essential.When planning a management strategy for encephaloceles, several factors must be considered. These include

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the site, size, contents, patency of the cerebrospinal fluid (CSF) pathway, neurological status, and any associated anomalies. Timely surgical intervention, as seen in our case, has shown favorable outcomes. Excision and repair resulted in the child exhibiting movement in all four limbs with intact reflexes. Postoperative follow-up is crucial and should occur at regular intervals to monitor for the development of hydrocephalus or any other neurological deficits. In the case discussed, there were no significant neurofocal findings postoperatively. This underscores the importance of ongoing surveillance to ensure the sustained well-being of the patient.

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