

Giant Occipital Encephalocele

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Abstract

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Encephalocele is a broad term representing herniation of cranial contents through a congenital defect in the cranium. If only cerebrospinal fluid (CSF) and meninges herniate, it is termed as a meningocele. A meningoencephalocele is herniation of both neural elements and meninges. The incidence of encephalocele is 1 per 5000 live births. Anterior encephaloceles are more common in males while 70% of posterior or occipital encephaloceles occur in females. We present one case of a giant occipital encephalocele to highlight the problems encountered in its management.

Keywords: Microcephaly; Occipital Encephalocele; Neurological Development.

Introduction

Encephalocele is a broad term representing herniation of cranial contents through a congenital defect in the cranium. If only cerebrospinal fluid (CSF) and meninges herniate, it is termed as a meningocele. A meningoencephalocele is herniation of both neural elements and meninges. The incidence of encephalocele is 1 per 5000 live births [1]. Anterior encephaloceles are more common in males while 70% of posterior or occipital encephaloceles occur in females. Occipital encephaloceles represent approximately 85 percent of lesions seen in the western hemisphere. The occipital encephalocele may occur through a bony defect in the occipital bone or extend into the foramen magnum and involve the posterior arch of atlas. The size of occipital encephaloceles may vary from small to large masses. 15 to 20 percent of children have additional congenital anomalies, including neural tube defects [1-3]. 60 to 70% of patients with posterior encephaloceles will develop hydrocephalous requiring ventriculo-peritoneal shunting [4].

Case Report

A 3 day-old female neonate born at term in a

peripheral hospital following caesarian section was transferred to a tertiary care teaching hospital. Baby with maternal history of three spontaneous abortions and no live siblings. At birth APGAR score was 10 and weight 4.8 kg. There was a large tense cystic swelling measuring 17 x 10 cm arising from posterior part of the head. The inferior part of the swelling ended at the neck. There was no abnormality on physical examination except for a large cystic mass in the occipital region. It was larger than the size of the head [Figure 1]. The skin over the swelling was stretched but well formed. The anterior fontanelle was closed. Baby was able to track objects and light and pupils were reactive. Routine hematological and biochemical investigations were reported as normal.



Fig. 1: Child with Giant Occipital Encephalocele

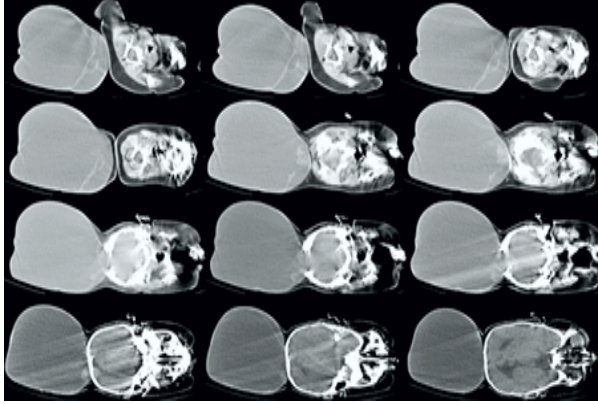


Fig. 2: CT scan brain showing encephalocele sac with protrusion of the contents

A computerized tomography scan (CT) demonstrated the encephalocele with evidence of herniation of very thin looking redundant brain tissue into the sac. CT images also revealed a significant defect of the occipital bone with well formed parietal bones [Figure 2].

Conclusions

The ultimate prognosis depends on the extent and nature of herniated contents and associated anomalies and many large encephaloceles have an excellent prognosis despite their size.

References

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