



AN UNUSUAL CASE OF BILOBED ENCEPHALOCOELE – CASE REPORT

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ABSTRACT Encephalocoels refer to a group of congenital malformations where brain tissue herniates through a defect in the skull. Various mechanisms have been proposed for its development, but, it is agreed that it is a defect of the mesoderm, that fails to fuse, resulting in herniation of neural tissue, along with CSF or meninges. Here, we present a case of an encephalococele which was abnormally shaped, being bilobed, while the normal shape being globular.

KEYWORDS : encephalococele, congenital ,bilobed

Introduction

Encephalocoels are congenital malformations resulting from neural tissue herniating out of the skull . the incidence is around 1per 4500 births. Various subtypes have been mentioned in the literature, common being the occipital subtype.[1] Occipital encephalocoels vary in shapes and sizes , and few studies have mentioned that more than 15% lesions are greater than 20cm in size.[2]

Our case of occipital encephalocoele is unique due to its big size and because of its bilobed structure.

Case report

A 4-day female baby was brought to us by the parents , with presence of a massive swelling over the occipital region. The baby was prematurely delivered by caesarian section and was 2.6kg in weight. She was taking her feeds normally , but the mother noticed the swelling getting tensed while the baby cried. On measurement the bilobed swelling measured about 12 cm in diameter at its widest part and a cranial defect could easily be palpated. The fontanelles were non tense.[Figure 1]



Figure 1. A large occipital encephalocoele with a bilobed morphology

CT brain revealed a large mass protruding out from the occipital region containing brain parenchyma , surrounded by meninges and an intact overlying skin. [Figure2,3]

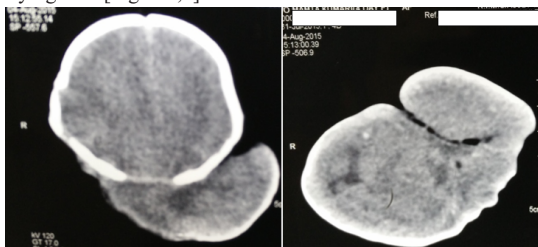


Figure 2 and 3 CT brain showing a defect in occipital skull bone with herniating brain with intact covering . the second picture shows the double lobed nature of the swelling.

The parents were counseled and advised surgery . the patient was operated with proper care , so as not to rupture the swelling . The patient was positioned prone with non cuffed ET tube placed. An elliptical incision was given near the base of the swelling with aim of separating the sac .once the sac was dissected , it was opened to remove the herniating content that consisted of redundant brain parenchyma. Haemostasis was secured and sac followed by skin closure was

performed. Postoperative phase was uneventful and the patient did not develop hydrocephalus ,over the next one year of follow up.

Discussion

Occipital encephalocoels are the most frequently encountered subtype of encephalococele, mostly found in females[2]. They are thought to arise due to a defect in the developing mesoderm, creating a defect in the skull, through which the neural tissue herniates. The overlying skin is found to be intact in most cases. With increasing size, increasing difficulty is encountered in operating upon these cases. The aim of the surgery remains removal of nonviable parenchyma and dural closure. Giant encephalocoels have been reported a few times.[3,4] But a large encephalocoele with such a unique shape is rarely encountered.

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