Case Report

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A Case Report of Front Oethimoidal Encephalocele in a Neonate

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Abstract

Encephalocele are herniation of cranial content through defect in the skull. Unlike western countries, anterior encephaloceles are the commonest types of encephalocele in Southeast Asia, part of Russia, and central Africa. Although diagnosis is usually clinical, imaging plays an important role in the characterization of the lesion. This paper presents the clinical presentation and surgical management of an 8 days old neonate with fronto ethimoidal encephalocele.

Keywords: Encephalocele; Fronto-ethimoidal; Neonate

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Introduction

Encephalocele is defined as protrusion of cranial contents beyond the normal confines of the skull. They may contain meninges (meningeal), brain matter and meninges (meningoencephalocele) or they may communicate with the ventricles (meningoencephalocystocele) [1] Encephaloceles are subdivided into the following categories: occipital, basal, convexity, and sincipital (anterior) [2].

The incidence of encephalocele has been reported as 1 in 5,000 to 10,000 live births [2, 3, 15] the incidence of specific types of encephalocele varies by geographic location and race. Anterior encephaloceles occur with the greatest frequency in Southeast Asia, parts of Russia, and central Africa, where they are seen in up to 1 in 3,500 live births and are the most common type of encephalocele observed. In contrast, anterior encephalocele is seen in only 1 in 35,000 live births in North America, where occipital encephalocele comprises 85% of all encephaloceles seen [4].

Genetic and environmental factors, including maternal infections; medications taken; or insufficient vitamin B12, folic acid, or mineral supplementation during the first trimester, are considered to be risk factors that contribute to encephalocele development [17].

Computed tomography (CT) scan with three-dimensional reconstruction is preferred for visualization of internal and external bony defects. Magnetic resonance imaging (MRI) can visualize the herniated contents within the sac and help in detecting other brain anomalies [12] direct surgical repair is the mainstay of treatment [4, 5]. The goals of surgery are: Excision of non-functional herniated brain tissue and water-tight dural closure, early prevention of rupture of thin sac and associated complications like infections, bleeding and drainage of cerebrospinal fluid and crania-facial reconstruction to achieve acceptable cosmesis [6].

This case report present the clinical presentation and management of 8 days old female neonate presented with fronto-nasal encephalocele.

Case Presentation

This is an 8 days old female neonate presented with a front nasal swelling since birth. She is born to a 24 years old PIII mother at 8 month of amenorrhea. Mother lives at a country side where access to a health centre was difficult. She didn't have ANC follow up. She didn't take iron-foliate supplementation which is given to all pregnant women in Ethiopia during their antenatal follow up. Otherwise there is no maternal history of medical illness, drug usage, giving birth to a child with similar abnormality [7-9].

Delivery was at local health centre via spontaneous vaginal delivery to affect the delivery of 2200gm alive female neonate. Neonate cried immediately after birth. Neonate was referred to Zewditu Memorial hospital with the compliant of swelling over the forehead since birth. There is no history of fever, abnormal body movement, leakage from the swelling.

Upon examination, neonate is active with stable vital signs. Occipital frontal circumference is 35 cm. There was a 6 * 4 cm non-tender, soft, cystic lobulated swelling between the eye globes over the upper tip of the nasal bridge. Neonate is alert with complete moro, sustained sucking and strong grasp reflex. She moves all her extremities. Pupil is mid-sized and reactive bilaterally. There are no spinal deformities.

Complete blood count, serum electrolyte, liver enzymes, bilirubin level, creatinine, coagulation profile were all in normal range. Computed tomography showed anterior basal skull defect associated with bilateral medial frontal lobe encephalocele and associated cystic structures suggesting large front nasal meningoencephalocele (**Figure 1**).

Under general anaesthesia, patient in supine position, bi frontal skin flap raised and craniotomy done. Extradural dissection of the encephalocle done and the defect identified. Dura opened and dissection undertaken till the base of the defect is identified. The external swelling dissected circumferentially separating the dysplastic tissue from the skin. Dysplastic tissue cauterized and removed. Split calvarias bone graft used for defect construction. Dura closed with water tight closure. Bone replaced and fixed. Skin closed in 2 layers after leaving epidural drain. Plastic surgery team closed the external wound of the encephalocele with primary closure.

Antibiotics were given for 5 days. She had smooth post-operative recovery period. She was discharged with follow up to the neurosurgery clinic with acceptable cosmesis to the parents (**Figure 2**).

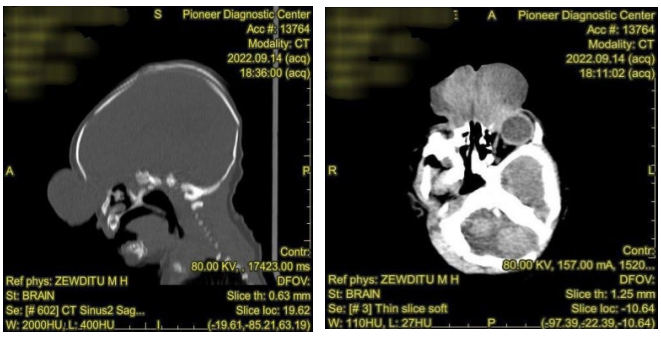

Figure 1 The lesion was isodense with the brain parenchyma with defect on the anterior basal skull on computed tomography.

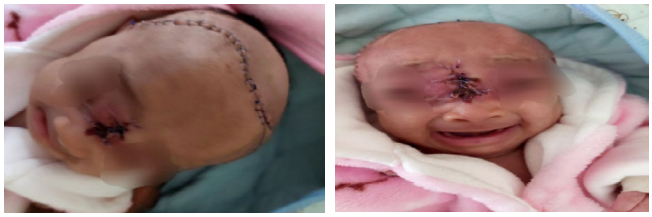

Figure 2 Post-operative day picture of the patient.

Discussion

An encephalocele is a herniation of the cranial contents out- side of the skull, which is described as a meningeal when the herniation sac contains leptomeninges and cerebrospinal fluid (CSF) [4] Encephaloceles are subdivided into the following categories: occipital, basal, convexity, and sincipital (anterior) [6] Suwanwela and Suwanwela have subdivided the sincipital group further into frontoethmoidal encephaloceles, interferential encephaloceles, and those encephaloceles associated with craniofacial clefts. Frontoethmoidal encephaloceles are herniations of the intracranial contents through a defect in the skull at the junction of the frontal and ethmoid bones. The frontoethmoidal group is subdivided further into nasofrontal, nasoethmoidal, and nano-orbital types [10-14].

The incidence of encephalocele has been reported as 1 in 5,000 to 10,000 live births [2, 3, 15]. A considerable disparity in the occurrence of encephalocele in different African countries was discovered in one systematic review and meta-analysis done in Africa. Nigeria 0.06%, Sudan 0.04%, Egypt 0.04%, Congo (DR) 0.02%, Ethiopia 0.02% and Tanzania 0.02% had a high birth prevalence of encephalocele [13] Anterior encephaloceles occur with the greatest frequency in Southeast Asia, parts of Russia, and central Africa, where they are seen in up to 1 in 3,500 live births and are the most common type of encephalocele observed [7]

Many theories have been postulated for the development of an anterior encephalocele

- Primary osseous defect leading to failure of the ethmoid plate to close around the olfactory nerve. Herniation of the brain then takes place at a later stage
- Increased ventricular pressure in the embryo could force the developing brain through the incompletely developed osseous structures
- The theory proposed by Jeffrey-Saint Hillarie (most acceptable): “The skull derives from two portions,” the endochondral cranial floor and the intramembranous cranial vault. At birth the frontal and ethmoidal bones are joined, but when the embryo is 3 months old, they are apart. Therefore, a weak point exists between the frontal and ethmoid bones and that a congenital defect could result in an encephalocele herniation
- A persistent craniopharyngeal canal could explain the rare encephaloceles through the sphenoid bone, but an early protrusion of cranial contents through this canal could lead to its persistence
- Developmental failure of the ossification centers in the sphenoid bone could also be considered as a possible cause of encephaloceles in this region [16].
- Genetic and environmental factors, including maternal in-fections; medications taken; or insufficient vitamin B12, folic acid, or mineral supplementation during the first trimester, are considered to be risk factors that contribute to encephalo- cele development [17].

The diagnosis is usually clinical. Usually the diagnosis is obvious, especially if there is clinical or palpable cerebral pulsation. Plain skull and facial radiographs can help delineate the size and position of the cranial defect through which the herniation passes. Encephaloceles can often be associated with other central nervous system abnormalities, in particular hydrocephalus. Ultrasound may be helpful in determining ventricular size, but ultimately computed tomographic (CT) scans with appropriate

brain windows and three-dimensional reconstruction give the best assessment of the diagnosis and pathological anatomy [5].

Management of these anomalies requires a multidisciplinary team of neurosurgeon, neuroanesthetist, pediatric, maxillofacial, and plastic surgeon working in congruity [16]. Treatment can be performed electively if the encephalocele is not associated with a CSF leak. However, a significant treatment consideration is the potential effect of the encephalocele mass on the development of the craniofacial skeleton that may prompt earlier intervention. Direct surgical repair is the mainstay of treatment [4].

Traditionally, two-stage correction by preliminary intracranial disconnection and subsequent extra cranial correction of the facial deformity was done [8]. A one-stage operation has been developed. In this a combined nasal-coronal approach with a frontal craniotomy is done. With this method, the frontal bone flap can also be remodelled to eliminate the trigon cephalic bulge, repair any external skull defects, and restore an aesthetic appearance such as with nasal augmentation [9].

The goals of surgery are: Excision of non-functional herniated brain tissue and water-tight Dural closure, early prevention of rupture of thin sac and associated complications like infections, bleeding and drainage of cerebrospinal fluid and cranio-facial reconstruction to achieve acceptable cosmesis [11]. The prognosis depends on site, size, content of encephalocele and any other associated congenital anomaly. Survival rate is higher, nearly 100% in anterior encephalocele compared to posterior encephalocele (55%), where vital structure of brain parenchyma might have herniated to the skull defect [10].

Learning Points

- Encephalocele is congenital or acquired lesion characterized by herniation of brain tissue through defect in the skull.
- Encephalocele prevalence at birth is high in Africa. Provision of folate supplementation during pregnancy aids in the prevention of neural tube defects.
- Management of patients with encephalocele with multi-disciplinary team involvement is necessary.

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