INTERNATIONAL JOURNAL OF HEALTH & MEDICAL RESEARCH

ISSN(print): 2833-213X, ISSN(online): 2833-2148

Volume 05 Issue 08 August 2026

DOI : 10.58806/ijhmr.2026.v5i8n01

Page No. 727-730

Congenital Frontoethmoidal Encephalocele: A Case Report

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ABSTRACT

Frontoethmoidal encephalocele is a rare neural tube defect in our community, but commonly seen in South-east Asia, less commonly in the western countries, where encephalocoele is more common in the occipital regions. Presentation of frontoethmoidal encephalocoele is swelling over the nasal bridge or between the inner canthi of the eyes since birth with a degree of hypertelorism. Diagnosis of frontoethmoidal encephalocele may pose a diagnostic dilemma due to multiple differential diagnoses which include fronto-ethmoidal sinus mucocele, haemangiomas, and gliomas. This paper emphasizes the importance of axial imaging, which includes computed tomography (CT) and magnetic resonance imaging (MRI), in the diagnosis of this condition, where early surgical intervention is needed to reduce morbidity and mortality associated with this congenital anomaly.

KEYWORDS: Encephalocele, Neural-tube defect, Hypertelorism, Computed tomography.

INTRODUCTION

Encephalocele are a group of disorders of the cranial defect which allows extracranial herniation of leptomeninges, brain, and cerebrospinal fluid (CSF)[1]. The defect can be classified as primary if they present at birth or secondary if acquired following trauma or surgery[2]. The occipital bone is the most common location, and frontoethmoidal encephalocele is less frequent. Frontoethmoidal encephalocele is a congenital defect involving the anterior skull base. It occurs as a result of persistent intracranial-extracranial communication, leading to herniation of cranial contents through a midline skull defect[3]. The cranial defect is classify based from the skull defect as occipital, parietal, basal, and frontoethmoidal. The frontoethmoidal encephalocele is also called sincipital encephalocele, and it is also classified according to the location of external skull defect as naso-frontal, nasoethmoidal, or naso-orbital, with some overlap or multiplicity[4]. The contents of frontoethmoidal encephalocele may vary from either the meninges (meningocele), meninges and brain (meningoencephalocoele), or part of a ventricle (hydromeningoencephalocele)[5]. The frontoethmoidal is the most common subtype of frontal encephalocele[6].

Clinical presentation of a patient with frontoethmoidal encephalocele is with a disfiguring swelling over the nose at the time of birth, with or without symptoms of nasal obstruction. The swelling may be ulcerative or not with CSF leak. Other presentation includes ataxia, seizure, delay milestone, hypertelorism, or quadriplegia, particularly in those with other associated anomalies like Dandy Walker malformation, agenesis of corpus callosum, or hydrocephalus.

Diagnosis of frontoethmoidal encephalocele can be readily made on cross-sectional images like computed tomography (CT), magnetic resonance imaging (MRI). Other imaging modalities include the conventional X-ray and ultrasound scan.

CASE PRESENTATION

ABY was a 12-month old female child from Gwagwalada-FCT Abuja, referred from primary healthcare centre old Kutunku in Gwagwalada to University of Abuja Teaching Hospital (UATH) Gwagwalada-Abuja, Nigeria. The child was said to have been born with a swelling around the nasal bone, the swelling was said to be increasing progressively with the child's age. Mother was said to have attended antenatal clinic at the same referring hospital and her antenatal visits were uneventful. Had an obstetric ultrasound scan during her antenatal visits and the result shows an increased amniotic fluid (polyhydramnious) otherwise other antenatal documentations were unremarkable.

Physical examination of the child revealed a healthy looking, not pale, acyanosed and anicteric child. Review of respiratory, central nervous (CNS) and cardiovascular systems were unremarkable. However, there was a fairly rounded swelling seen at the naso-ethmoidal region, the swelling was soft and fluctuant to touch measuring about 5 x 3cm in dimensions devoid of thrill or bruit at the region of the swelling. The skin covering the defect was intact with no leakage of CSF.

A computed tomography (CT) scan of the head was requested which revealed a rounded fronto-ethmoidal anterior base of the skull defect with protrusion of intracranial content through the defect (fig. 1-3).

In view of the aforementioned CT features a diagnosis of congenital frontoethmoidal encephalocele was made. Following this diagnosis patient was worked up for frontoencephalocle repair. Thus, a bicoronal incision was used to approach this malformation exposing the bony defect. The encephalocele was dissected, mobilized and excised. The dura was closed in a water-tight fashion using prolene 4/0. The cranial base defect was then repaired with bone graft from patient's calvarium and the wound closed in layers using vicryl 3/0.

Patient recovery was uneventful of complication and was discharged on the eight day post-op after removal of skin sutures. Had a single clinic follow-up visit but lost to follow-up after the first clinic visit.

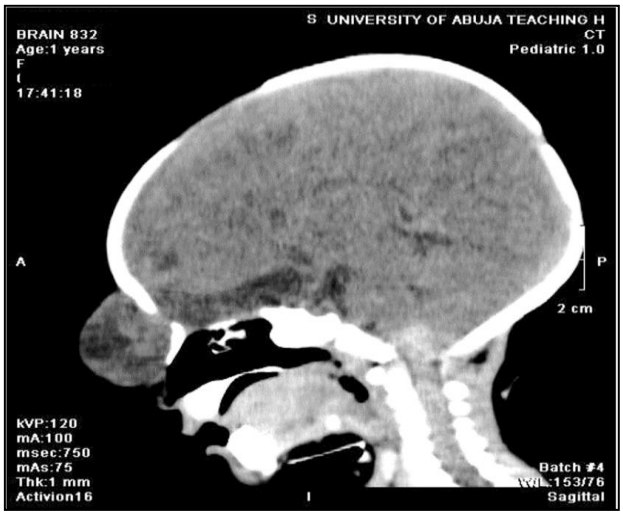

Figure 1: mid-sagittal computed tomography of the brain shows a well-defined frontoethmoidal osseous defect with protrusion of cranial contents

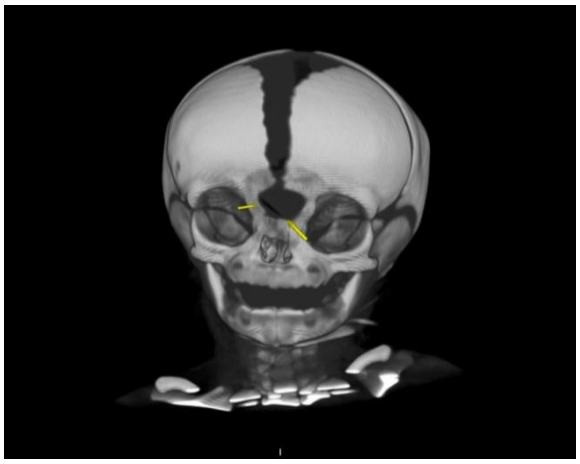

Figure 2: 3-D reformatted computed tomographic image revealed a rounded nasoethoimoidal osseous defect

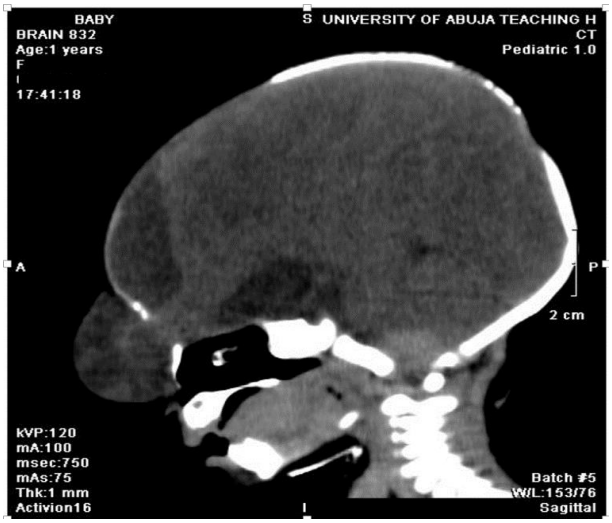

Figure 3: mid-sagittal computed tomography of the brain (soft tissue window) shows protrusion of frontal lobe of the brain through the cranial defect.

DISCUSSION

Frontoethimoidal encephalocele is a rare congenital anomaly characterized by herniation of intracranial contents through the cranial and facial bones due to a defect of closure of the anterior neuropore of the neural tube[7]. The lesion usually affects children of the low socioeconomic class with aetiology poorly understood.[8] However, some authors have suggested ethnic, genetic, environmental factor and paternal age as aetiologic factors in development of encephalocele. Fungal, teratogenic agents such as aflatoxin and ochratoxin found in moldy rice during wet season could be implicated in the aetiology of encephalocele.[9] Several theories have been postulated for the development of anterior encephalocele: primary osseous defect leading to failure of the ethmoidal plate to close around the olfactory nerve. Thus, herniation of brain take place at a later stage. Failure of ossification in the sphenoidal bone, persistent craniopharyngeal canal, increased intraventricular pressure in the embryo forcing the brain to herniate through the incompletely developed osseous defect.[10]

Suwanwela and Suwanwela et al[4] classified frontoethmoidal encephalocele based on the work of Mesterton[11] and Von Meyer.[12] Classifying the defect as nasofrontal, nasoethmoidal and naso-orbital. However, a modified classification of anterior encephalocele was proposed by Bhagwati and Mahapatra.[10] Where the naso-orbital type is found at the medial orbital wall walls situated in the frontal process of the maxilla and the lacrimal bones. The frontoethmoidal encephalocoele is associated with craniofacial deformity consisting of inter orbital hypertelorism (rarely true orbital hypertelorism/telorbitism because, the medial orbital walls are widened but the lateral orbital walls are usually in the normal position).[13]

Recurrent meningoencephalitis can be cause by encephalocele due to direct communication of the central nervous system with the external environment aiding the entry of microorganisms. Most common pathologic organism associated with such patients are streptococcus pneumonia, staphylococcus aureus and Neisseria meningitides.[14]

Diagnosis of frontoethmoidal encephalocele can be challenging in resource limited centres without axial imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI); because of the vast differential diagnosis of this anomaly which include traumatic encephalocele, frontoethmoidal sinus mucocele, neurinoma, haemangioma and glioma.

CONCLUSION

Frontoethmoidal encephaloceole is a rare congenital anomaly that may present as a nasal mass at birth. Early diagnosis using cross sectional imaging particularly CT and MRI is essential for accurate characterization of the skull defect and surgical planning. Prompt surgical intervention is necessary to reduce the risk of complications such as infection and neurological morbidity.

Sources of support: Authors bore the cost of this case report
Conflict of interest: Authors declare no conflicts of interest

Consent statement

Approval to report this case was obtained from the parents of the child and a written informed consent to publish the findings of this case to any journal was also obtained from the parents prior to writing this case report.

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