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A Congenital Fronto-ethmoidal Encephalocele in a Female: A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. Authors OK and AEHJ wrote the first draft of the manuscript and managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Encephalocele is defined as the protrusion of cranial contents beyond the normal confines of the skull. They may contain meninges (meningocele), brain tissue, and meninges (meningoencephalocele), or they may communicate with the ventricles (meningoencephalocystocele). We report the case of 39 years old patient with congenital frontoethmoidal encephalocele. Through trans-facial approach resection of nonfunctional brain tissue was performed then the dural defect was repaired. The nasal deformity was corrected using the cement to provide dorsal nasal support from the nasal bones, the orbit's inner walls, to the lateral nasal cartilage. The absence of recurrence marked the patient's follow-up. This case is presented for its rarity.

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Keywords: Cement reconstruction; congenital abnormalities; Encephalocele; frontal; trans-facial approach.

1. INTRODUCTION

Encephalocele is a protrusion of brain contents beyond the normal limits of the skull. It may have meninges, brain tissue, or communicate with the ventricles [1,2]. Swelling is the most dominant manifestation. The skin's appearance may be normal, thin, or thick, with skin expansion because of the mass's slow evolution. The mass's power can obstruct the lacrimal ducts, telecanthus, strabismus, and nasal obstruction and a decrease visual acuity [3,4,5].

2. CASE REPORT

We report the case of 39 years old patient with a poorly followed pregnancy, 2 children in good health, no family history of the same disease, and swelling (encephalocele) evolving since birth time, which is increasing the volume gradually causing facial deformities with preservation of the general condition. Clinical examination revealed a 10 cm long mass in the Centro-facial region not tender at the palpation with telecanthus, hypertelorism, orbital dystopia, and epiphora deformities in the orbital regions. No obstruction of the nose and the visual acuity was conserved. The thickened discolored skin was excised for the correction, trans-facial approach through a perpendicular incision along the mass's long axis. Nonfunctional brain frontal lobe resection was performed then the dural defect was repaired.

The nasal deformity was corrected using the cement to provide dorsal nasal support from the nasal bones, the orbit's inner walls, to the lateral nasal cartilage. The dimension of the defect: 5 x 4 x 5 cm.

A costochondral graft for nasal reconstruction is used for giving an esthetic nasal tip that is not overly rigid [6] in our case; we opted for the cement to provide a dorsal nasal pyramid. The follow-up of the patient was normal. The absence of recurrence marked the patient's follow-up. She has been advised to undergo procedures for further staged cosmetic correction.

3. DISCUSSION

Anterior encephaloceles are congenital anomalies characterized by the defect in the closure of the neural tube's anterior neuropore, resulting in herniation of brain tissue through the skull's bony defect and face [7]; some suggest that the etiology could be ethnic, genetic, environmental factors, and the father's age. In our case, the most preponderant etiology is a folic acid deficiency as the patient had a poorly followed pregnancy suggesting folic acid deficiency, which is consistent with the literature. These lesions affect the lower socio-economic class children, but their etiology remains poorly known [8].

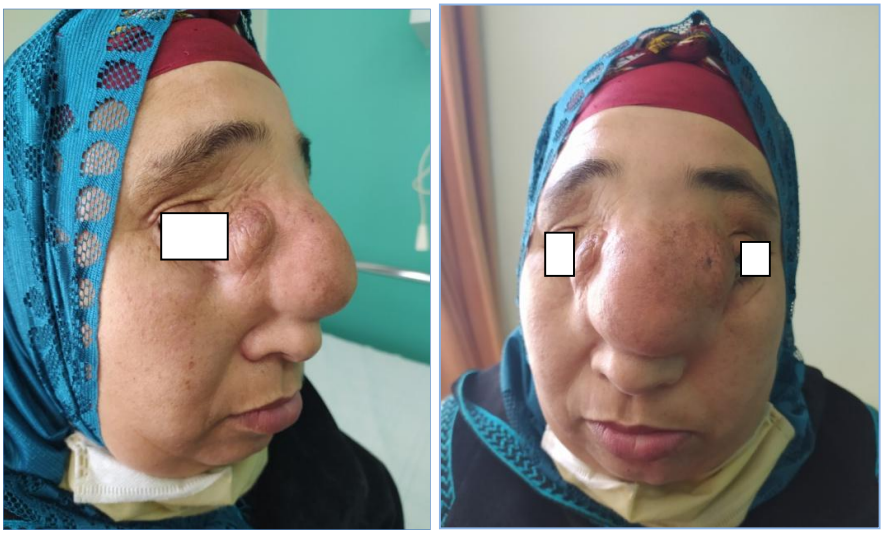

Fig. 1. A 39-year-old patient with fronto-naso-orbital encephalocele

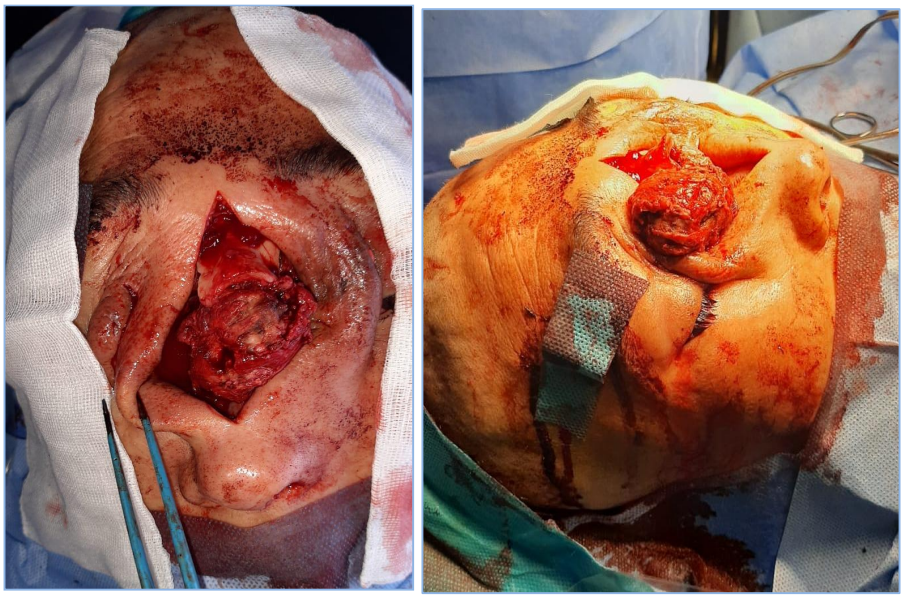

Fig. 2. Operative Image: Lower facial approach showing the frontal encephalocele

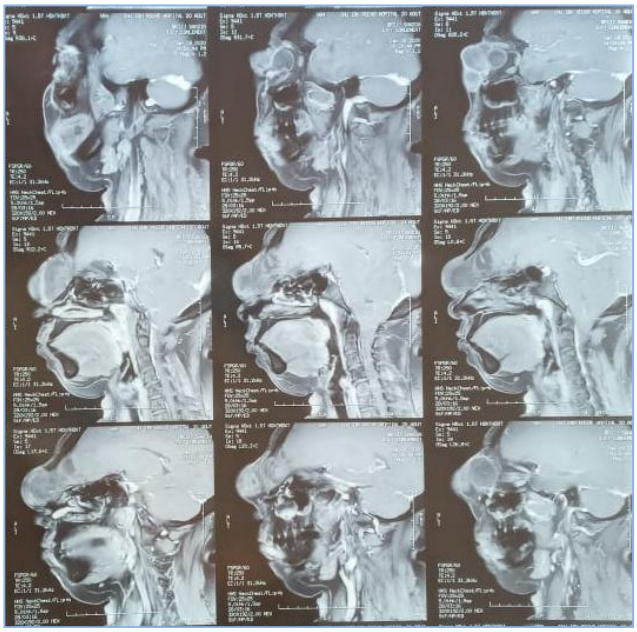

Fig. 3. MRI showing the naso-orbital encephalocele

The encephaloceles can be congenital or acquired as a result of a tumor, hydrocephalus, or other cause [9].

The fronto-ethmoidal encephalocele is a rare congenital disease affecting patients in Southeast Asian countries such as Thailand, Malaysia, and Indonesia, with a frequency of 1 in 5000 [10]. Very rare cases have been reported in Europe, North America, and the Middle East; and the reasonable explanation for this particular geographical distribution is unknown. [11]. The sincipital encephalomeningocele seems to be more frequent than occipital lesions in Bangkok, Manila, and Singapore, where a sincipital lesion was found without occipital malformation. The reverse is true for Europe, North and South America, and Australia [12].

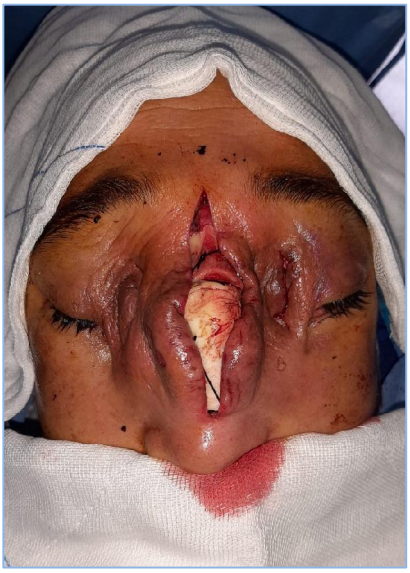

Fig. 4. reconstruction of the defect with the bone cement

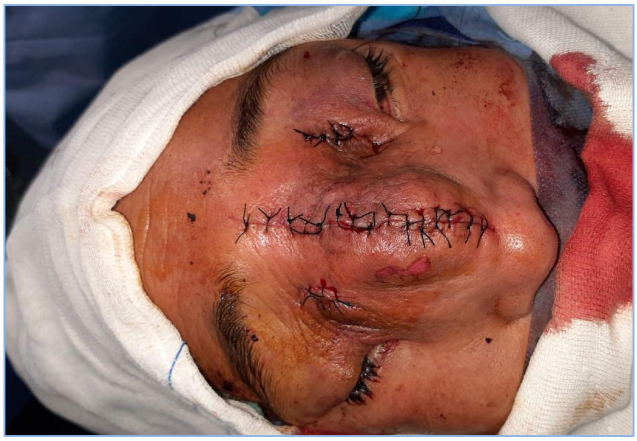

Fig. 5. Post-operative result

According to the literature, various environmental factors have been implicated in the development of encephalocele, primarily folate deficiency. However, there is not much literature on the relationship between maternal folate levels and their incidence. Also, the role of teratogenic and fungal agents could be involved [13,14].

The encephalomeningoceles can be classified according to their location, as described by Suwanwela in 1972 [12].

1. Occipital encephalomeningocele (75% of cases)
2. Sincipital encephalomeningocele (10% of cases)
   a. Frontoethmoidal encephalomeningocele (FEEM)

   i. Nasofrontal
   ii. Naso-ethmoidal
   iii. Naso-orbital

   b. Interfrontal encephalomeningocele
   c. Associated with craniofacial clefts
   Convexity encephalomeningocele (5% of cases)
   - Parietal encephalomeningocele
   Basal encephalomeningocele (10% of cases)
   - Intrasphenoidal
   a. Transsphenoidal
   b. Transethmoidal

c. Sphenoethmoidal
d. Frontosphenoidal or spheno-orbital

Several theories are implicated in the development of an anterior encephalocele, including increased ventricular pressure and primary bone defect around the olfactory nerve. The defect between the frontal and the ethmoidal bones in the embryonic life, almost three months old, could result in a herniation of the encephalocele. Protrusion through the craniopharyngeal canal is rare, but it leads to its persistence when it is early.

In the naso-orbital type, the defect is in the medial orbital walls situated in the maxilla's frontal process and the lacrimal bones [15]. The lateral orbital walls are respected while the medial walls are widened and repressed by the mass effect of the fronto-ethmoidal encephalocele causing craniofacial deformation. The skeletal deformity can manifest itself as interorbital hypertelorism, secondary trigonocephaly, orbital dystopia, facial elongation, nasal deformity, and dental malocclusion [6].

In our case, we opted for the lower facial approach; several approaches are used:

A. The combined intra- and extra-cranial/anterior facial approach includes a bicoronal incision with facial reconstruction [9].
B. A pure extracranial/lower facial approach; is performed in cases of relatively minor craniofacial malformation and promises to be feasible in resource-limited institutions where it is difficult to perform a craniotomy or other neurosurgical procedure [16].

Most children are mentally normal, and others have neurological complications or associated brain abnormalities.

The prognosis depends on site, size, encephalocele content, and any other associated congenital anomaly. The survival rate is higher in anterior encephalocele than posterior encephalocele, where the vital structure of brain parenchyma might have herniated to the skull defect [17,18].

4. CONCLUSION

Fronto-ethmoidal encephalocele is a rare congenital anomaly. Early surgical intervention can avoid medical and social problems.

In our case, surgical treatment was proposed at an advanced age. In our case, we opted for a trans-facial and lesional approach as the complications are less invasive.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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