Case study

A Congenital Frontoethmoidal Encephalocele in a Female: A Case Report

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 a 39 years old patient with congenital Frontoethmoidal Encephalocele, a poorly followed pregnancy, 2 children in good health, lower socio-economic class, no family history of the same disease. The deformity caused by encephalocele led the patient to consult but late because of the unavailability of financial resources and the lack of information, making management even more complex if the diagnosis and management were early. The swelling (encephalocele) was evolving since birth time, increasing the volume gradually, causing facial deformities. Clinical examination revealed a 10 cm long mass in the Centro-facial region, painful on palpation with telecanthus, hypertelorism, orbital dystopia and epiphora, and deformities in the orbital regions. No obstruction of the nose and the visual acuity was conserved for the correction, trans-facial approach through a perpendicular incision along the long axis of the mass, the thickened discolorated skin was excised, nonfunctional brain frontal lobe resection. 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 dimension of the defect: 5 x 4 x 5 cm. The absence of recurrence marked the patient's follow-up. This case is presented for its rarity.

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, 4-9]. Swelling is the most dominant manifestation. The skin's appearance may be either 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 [3-5, 3, 4, 5].

2. CASE REPORT

We report the case of a 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 unpainful 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.
Fig. 1. A 39-year-old patient with fronto-naso-orbital encephalocele

Fig. 2. Perpendicular incision showing the frontal encephalocele
Fig. 3. MRI showing the naso-orbital encephalocele
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].

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

According to the literature, various environmental factors have been implicated in the development of encephalocele, primarily folate deficiency, although there is not much literature on the relationship between maternal folate levels and its incidence. Also, the role of teratogenic and fungal agents could be involved [10,11].

Many theories involved in the development of an anterior encephalocele:

- Primary osseous defect leading to failure of the ethmoidal plate to close around the olfactory nerve. Herniation of the brain then takes place at a later stage
- Increased ventricular pressure

"The skull derives from two portions," the endochondral cranial floor and the intramembranous cranial vault. The defect between the frontal and the ethmoidal bones in the embryonic life, almost in the three months old, could result in a herniation of the encephalocele.

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.

In the naso-orbital type, the defect is in the medial orbital walls situated in the maxilla's frontal process and the lacrimal bones. The frontoethmoidal encephalocele is associated with craniofacial deformity consisting of interorbital hypertelorism (rarely true orbital hypertelorism because the medial orbital walls are widened, but the lateral orbital walls are usually in the normal position). Hence, the term interorbital hypertelorism gives a better description of the skeletal deformity. Other deformities could be secondary trigonocephaly, orbital dystopia, elongation of the face, nasal deformity, and dental malocclusion. Some of the children have neurological complications or associated brain anomalies, although most are mentally normal.

The prognosis depends on site, size, encephalocele content, and any other associated congenital anomaly. The survival rate is higher, nearly 100% in anterior encephalocele than posterior encephalocele (55%), where the vital structure of brain parenchyma might have herniated to the skull defect.

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.


