Frontoethmoidal Encephalocele: A Case Report and Review of the Literature

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Received: September 25, 2020; Published: February 27, 2021

Abstract

Encephalocele is a protrusion of cranial contents through the natural orifices of the skull.

We report the case of 39 years old patient, with no particular pathological history, 2 children in good health, an encephalocele 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 centrofacial region unpainful at the 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, transfacial approach through a perpendicular incision along the long axis of the mass, the thickened discolored skin was excised. nonfunctional brain frontal lobe resection was performed then the dural defect repaired.

Keywords: Frontoethmoidal Encephalocele; Telecanthus; Hypertelorism; Orbital Dystopia

Introduction

Encephalocele is a protrusion of cranial contents through the natural orifices of the skull [1]. The swellings are the most dominant manifestations. The skin over the mass may be normal in appearance, thin and shiny or thick and wrinkled associated with hyperpigmentation. Visual acuity may be decreased. Strabismus and lacrimal obstructions, resulting in epiphora and/or dacryocystitis can be observed [3-5].

Case Report

A 39-year-old patient, with no particular pathological history, 2 children in good health, an encephalocele since birth time which is increasing the volume gradually causing facial deformities with preservation of the general condition. On clinical examination revealed an asymmetric face with a fronto-nasal swelling 10 cm long, in the centrofacial region unpainful at the palpation with telecanthus, hypertelorism, orbital dystopia and epiphora and deformities at the orbital regions. No obstruction of the nose and the visual acuity was conserved. For the correction, transfacial approach through perpendicular incision along the long axis of the mass, the thickened discolored skin was excised. nonfunctional brain frontal lobe resection was performed then the dural defect repaired. The nasal deformity was corrected using the bone graft or the cement to provide dorsal nasal support.

The use of costochondral graft for nasal reconstruction is used for giving an esthetic nasal tip that is not overly rigid [6]. The follow-up of the patient was normal. She has been advised to undergo procedures for further staged cosmetic correction.

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

Figure 2: Perpendicular incision showing the frontal encephalocele.

Figure 3: MRI showing the naso-orbital encephalocele.

Figure 4: Reconstruction of the defect with bone cement.
Discussion

Anterior encephaloceles are rare congenital abnormalities which are characterized by herniation of intracranial components through the cranial and facial bones due to a failure to close the anterior neuropore of the neural tube [7].

Frontoethmoidal meningoencephaloceles are common in children from the low socioeconomic class, but its etiology still poorly understood [8]. Encephaloceles can be congenital or acquired secondary [9].

Among environmental factors, fungal and teratogenic agents could be involved [10]. Many authors assert the role of folic acid deficiency, but there is not enough literature on the subject [11].

The defect in the naso-orbital type is located in the medial orbital walls located in the frontal process of the maxilla and lacrimal bones [12]. Fronto-ethmoidal encephalocele is associated with a craniofacial deformity involving interorbital hypertelorism [6].

Other abnormalities may include facial lengthening, secondary trigonocephaly, orbital dystopia, nasal deformity. There are some patients with neurological complications or associated brain abnormalities, although most are mentally normal.

The site, size, content of encephalocele are the prognostic factors.

In the anterior encephalocele generally the survival rate is higher compared to the posterior encephalocele [13,14].

Conclusion

Early diagnosis and treatment is important, in order to avoid major deformities and to prevent infection in the event of a ruptured encephalocele.

The resection of pathological tissue is the rule associated with meningo-plasty, repair of the bone defect and reconstruction of facial deformities (bones and soft tissues).

**Bibliography**


Volume 12 Issue 12 December 2020
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