

# Transthmoidal encephalocele: an unusual cause of pediatric nasal obstruction

Mohamed Dhaha, Abd El Hafidh Slimane, Nadhir Karmani, Asma Bouhoula, Jalel Kalel

National Institute of Neurology Mongi Hmida, Tunis, Tunisia

## Abstract

An encephalocele is a congenital malformation characterized by the herniation of brain content beyond the normal confines of the skull. Transthmoidal encephaloceles represent a rare subtype of anterior encephaloceles and consists in a protrusion of the brain content in the nasal cavity through a defect in the horizontal plate of the ethmoid bone. Clinically it usually manifests by upper airway obstruction. We report the case of an 8-month-old infant with a transthmoidal encephalocele revealed by a chronic nasal obstruction and snoring. The patient underwent left frontal craniotomy allowing repositioning of the herniation. Ethmoidal defect was repaired by a fascia lata graft. Post operative recovery was uneventful apart from a CSF rhinorrhea which spontaneously dried up in 3 days.

## Keywords

Transthmoidal encephalocele, nasal obstruction, frontal craniotomy.

## Corresponding author

Mohamed Dhaha, National Institute of Neurology Mongi Hmida, Tunis, Tunisia; email: dhaha.mohamed87@gmail.com.

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## Introduction

An encephalocele is a rare congenital condition defined by the herniation of brain content beyond the normal confines of the skull. Anterior encephaloceles are classically divided into frontoethmoidal and basal encephaloceles depending on the protrusion of the cranial content in relation to the cribriform ethmoidal plate. Basal encephaloceles herniates posterior to the cribriform plate and are present in nasal cavity as opposite to external masses [1]. Therefore, these malformations are clinically not visible at birth and are usually diagnosed when exploring an upper airway obstruction [2]. Transethmoidal encephalocele is a subtype of basal encephaloceles and contributes only 8% of all anterior encephaloceles [3]. Surgery is the only treatment. We report the rare case of an 8-month-old child presenting a congenital transethmoidal encephalocele revealed by a nasal obstruction treated with frontal approach.

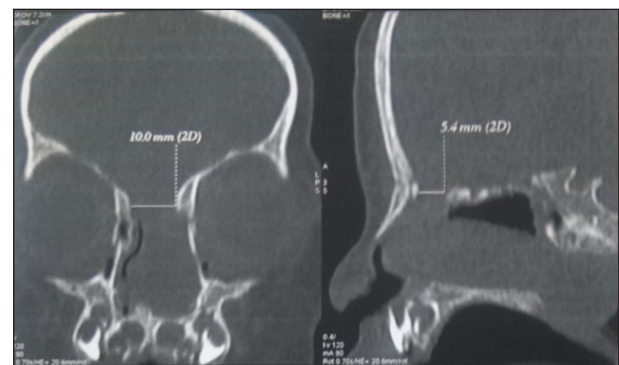
## Case report

An 8-month-old infant has been addressed by his pediatrician to the Department of Neurosurgery at the National Institute of Neurology Mongi Hmida, Tunis, Tunisia. Parents reported nasal obstruction, snoring and breastfeeding difficulty since birth. The baby was born at term via Caesarian section. The pregnancy was normal, and no particular medical history was noted. Physical examination showed normal size and weight which were in accordance with the age. No facial malformation such as hypertelorism and exophthalmos was observed. Rhinoscopy showed a grayish mass filling the left nasal cavity. Cerebral and facial massif CT showed a defect in the ethmoidal horizontal plate measuring 10 x 5 mm associated to a transethmoidal meningo encephalocele (**Fig. 1** and **Fig. 2**). The infant underwent left frontal craniotomy allowing repositioning of the herniated content using an intracranial approach. The defect was closed using a fascia lata graft. The patient presented a clear anterior rhinorrhea the first 3 days of the post operative period that spontaneously dried up. A noticeable clinical amelioration concerning nasal obstruction and snoring was reported since the 2<sup>nd</sup> month after surgery.

## Discussion

Encephaloceles represent a type of open neural tube defect that causes herniation of brain

content [4]. The pathogenesis of this malformation is not well elucidated and many theories have been proposed [5, 6]. A combination of genetic and environmental factors is the most admitted one. Based on location and type of skull defect, encephaloceles are classified as occipital encephaloceles, encephaloceles of the cranial vault and anterior encephaloceles [7]. Frontoethmoidal and basal encephaloceles represent the two subtypes of anterior encephaloceles and are both rare conditions with a relatively high incidence (1:5,000 live birth) in Southeast Asia [8]. This observation illustrates a rare case of basal encephalocele. The herniation of the brain content was through the cribriform ethmoidal plate into the nasal cavity and so called transethmoidal encephalocele. As



**Figure 1.** CT scan of the face in coronal and sagittal section showing a transethmoidal encephalocele through a bony defect in the horizontal plate of the ethmoid.



**Figure 2.** CT scan of the face in axial section showing a left transethmoidal encephalocele.

opposite to facial masses seen in frontoethmoidal encephaloceles, transethmoidal encephaloceles are clinically not visible. As reported in literature, it manifests with upper airway obstruction [9]. In fact, nasal obstruction and snoring were the 2 chief complaints in our case.

Brain and facial CT scan was the only imaging exam performed. In fact, CT is preferred in visualization of bony defect [10]. MRI was not realized due to lack of technical resources. This imaging technique is recommended as it allows a clearer vision of herniation content [11]. Digital angiography was indicated by some authors to evaluate the presence of vascular structure in the encephalocele [12]. Nasal biopsy of the herniated mass is contraindicated and should not be realized in any case [13].

Surgery is the only treatment of encephaloceles. These malformations are always protected by a normal skin or an epidermal layer [14]. Hence, treatment should not be considered as an emergency at birth, but intervention should be performed early in life to allow for a more complete repair of the dural defect [13, 15]. As seen in this case, transcranial route involving craniotomy remains the standard method as it offers an optimal exposition and allows a better repositioning of the herniated content. In order to repair the skull defect, we opted for a fascia lata flap rather than a full thickness scalp-galeal flap. In fact, fascia lata flap is more rigid and efficient in repairing a relatively large defect like this one. As reported by Abdel-Aziz et al., this flap is reliable in closing the cranial bony defect [13]. The endoscopic approach is more often used with older children [16, 17]. CSF rhinorrhea is frequently observed during the post operative period and usually persists for few days [18]. Prognosis is roughly good and is determined by the associated congenital anomalies [11, 14]. Mortality rate is low, with 3% surgery related mortality [19]. Evaluation of the neurocognitive development was not possible in this case because of the short term follow up, but it has been reported to be favorable especially with anterior encephaloceles [20].

## Conclusion

Transethmoidal encephaloceles are rare condition. However, pediatricians and otolaryngologists should evoke the diagnosis in front of chronic nasal obstruction in infants as the patient should be

prepared to a surgical management. Prognosis is good for isolated forms.

## Declaration of interest

The Authors declare that there is no conflict of interest.

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