

Surgical Technique and Review of Management of Frontoethmoidal Encephalocoele - A Case Report

Hasna Abdourafiq, Farid Zahrou, Lamia Benantar, Khalid Aniba

Department of Neurosurgery, Ibn Tofail Hospital, University Hospital Mohammed 6, University Cadi Ayyad, Marrakesh, Morocco

Abstract

The Rationale: Encephalocoele is a herniation of the brain formed during embryonic development, because of the incomplete closure of Neural Tube. It is a rare skull defect and most cases are located in the occipital bone. Frontal encephalocoeles are very rare and they may involve the ethmoid bone, nasal bones and/or the orbits. Surgical repair is complex and usually requires a multidisciplinary approach. **Patient Concerns:** We present a case of a 6-month-old baby girl with a congenital frontoethmoidal encephalocoele. **Diagnosis:** The diagnosis was made by computed tomography scan that showed a defect on the frontoethmoidal zone. **Treatment:** Cranial approach was employed using standard bicoronal access. The malformation was removed and the defect was repaired by using an autologous parietal bone graft, without any complication in the follow-up. **Outcomes and Take-away Lessons:** The goal of the surgery is to reconstruct the normal anatomy, to achieve a good cosmetic repair, and to avoid a cerebrospinal fluid leak. A description of the case and the surgical technique is presented with a review of literature.

Keywords: Congenital malformation, *Encephalocoele*, frontoethmoidal encephalocoele

INTRODUCTION

Frontoethmoidal encephalomeningocoele (FEEM) is a congenital neural tube anomaly, with herniation of intracranial material through a defect in the dura and anterior skull base at the junction of frontal and ethmoidal bones. They may contain brain tissue and meninges (meningoencephalocoele) or only meninges (meningocoele).^[1] The causes of this malformation are still poorly understood. In addition, if the prognosis is often not engaged, social and esthetic prognosis can be terrible.

We describe a case of fronto-ethmoidal encephalocoele as well as the surgical technique performed and a review of the literature.

CASE REPORT

Clinical features

A 6-month-old baby girl was hospitalized for a medio-frontal large mass that has been present since birth. The lesion slowly increased in size, causing obvious facial disfigurement. The mass was bulging between the two orbits and occupying the lowest part of frontal area and superior part of the

nasal area [Figure 1]. The skin was thin but intact. She had no neurological deficit, the baby moved all four limbs spontaneously and her psychomotor development was normal for age.

In addition, the ENT examination was normal and no other congenital malformations were found.

At this clinical stage, the differential diagnosis could be made of traumatic encephalocoele, ethmoid-frontal sinus mucocoele, neurinoma or glioma.

Medical imaging

The computed tomography (CT) scan showed a bone defect in the zone of the frontal bone and anterior part of the ethmoid bone with extrusion of cranial contents including meninges and brain tissue through the defect.

Address for correspondence: Dr. Hasna Abdourafiq, Ibn Tofail Hospital, Abdelouahab Derrag Street, Marrakesh 40000, Morocco.
E-mail: hasnaabdourafiq@gmail.com

Received: 17-07-2020

Last Revised: 19-09-2020

Accepted: 11-02-2021

Published: 24-07-2021

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: WKHLRPMedknow_reprints@wolterskluwer.com

How to cite this article: Abdourafiq H, Zahrou F, Benantar L, Aniba K. Surgical technique and review of management of frontoethmoidal encephalocoele - A case report. *Ann Maxillofac Surg* 2021;11:132-5.

Access this article online

Quick Response Code:



Website:
www.amsjournal.com

DOI:
10.4103/ams.ams_274_20

The defect was about 13.5 mm in diameter [Figure 2]. The magnetic resonance imaging (MRI) was not performed for this patient.

The patient was planned for a single-stage surgical excision of the encephalomeningocele and correction of the craniofacial deformity by a multidisciplinary team including a neurosurgeon, maxillofacial surgeon, and paediatric anaesthesiologist.

Surgical technique

Cranial approach was employed using a standard bicoronal incision. Hair was shaved only in the area of incision. A full scalp-galeal flap was raised exposing frontal bone, frontonasal sutures, and supraorbital rims and nerves. After that a circumferential dissection of the encephalocoele sac was performed, freeing it from the surrounding tissue, until reaching the bone that delimits the malformation including the nasal bone [Figure 3].

A bone flap was then made after making a median trephine hole, about 2 cm above the junction between nasal and frontal bone to give more exposure of the malformation that contained nonfunctional brain tissue.

Once the hernial sac was dissected, it was excluded by ligation of its base. Once ligated, the herniated tissue was resected with electrocautery. A pericranial graft was then placed and fixed by the biological glue to ensure a better closure of the dura.

This cranial procedure was followed by reconstruction of craniofacial deformity. For this, a bone graft was taken from parietal bone, according to the size and shape of the defect. The graft was fixed by absorbable suture taking support on the crista galli and the frontal bone shell [Figure 4]. The parietal defect was allowed to consolidate with growth. The bicoronal incision was then closed using continuous subcuticular sutures.

RESULTS

No intraoperative complications were observed, the surgical site was clean. There was no cerebrospinal fluid (CSF) leak or wound infection on postoperative outcomes.



Figure 1: Preoperative images of the frontoethmoidal encephalocoele

The histological examination of the herniated tissue did not reveal any abnormalities.

The duration of follow-up was about 1-1/2 years. During the follow-up, the patient's evolution was satisfactory both functionally and aesthetically [Figure 5 and 6].

DISCUSSION

Anterior encephalocoeles are rare congenital abnormalities characterized by herniation of the intracranial components through the cranial and facial bones due to a defect of the closure of the anterior neuropore of the neural tube.^[1]

Since 1976 Suwanwela and Suwanwela^[2] classified the malformation into three types depending on the site of bone defects [Table 1].

A FEEM is a herniation of the brain and meninges through a congenital bone defect in the skull base located at the junction of the frontal and ethmoidal bones.

Frontoethmoidal meningoencephalocoeles are common in Southeast Asian countries, with an incidence of 1 in 5000 live births. This malformation is absent in European and

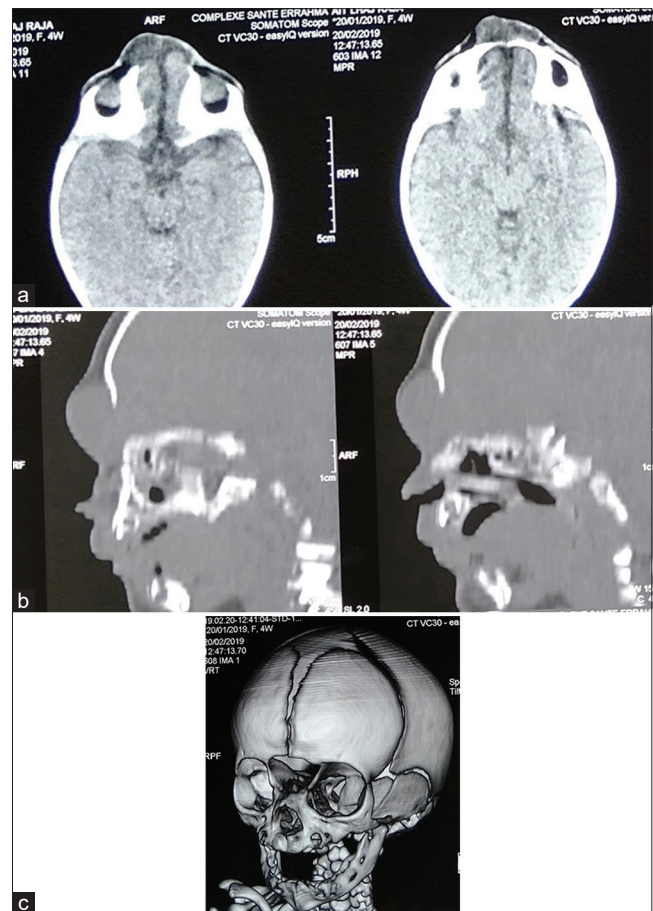


Figure 2: (a) Axial computed tomography scan image that shows a fronto-ethmoidal encephalocoele with herniation of brain tissue. (b) Sagittal image that shows a bone defect of 13.5 mm in diameter in the frontal bone. (c) Three-dimensional computed tomography scan

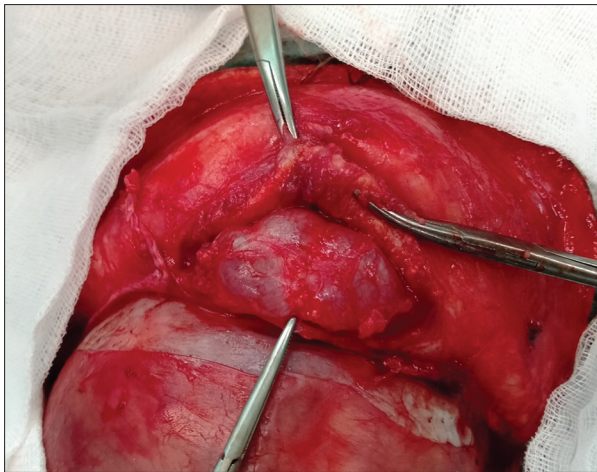


Figure 3: Dissection of the hernial sac from the surrounding structures in the naso-orbital area after a bicoronal approach

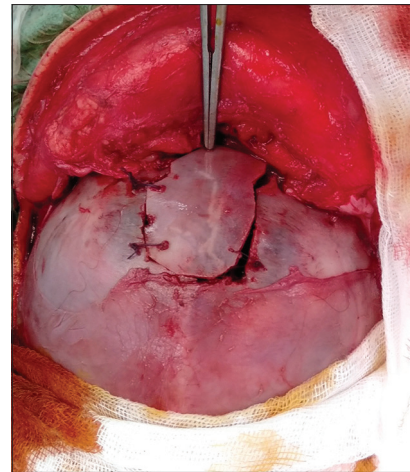


Figure 4: After placing a pericranial graft to close the dura, an autologous graft from the parietal bone was used to reconstruct the bone defect. The graft is fixed by absorbable suture



Figure 5: (a and b) Postoperative after 5 months of follow-up



Figure 6: Postoperative after 18 months of follow-up

North American population. These lesions affect children from the lower socioeconomic class, but its etiology remains poorly understood.^[3] Many have defined it as a multifactorial condition, with a combination of genetic and environmental factors.^[4]

The majority of cases are present at birth as a medio-frontal mass, which is usually covered by skin. In some cases, encephalocoeles are hidden and may present with symptoms as snoring, nasal obstruction, CSF leak or recurrent meningitis.^[5]

MRI and CT scan are necessary to plan the surgery. MRI is essential to understand brain herniation. AngioMRI can be used to evaluate the position of the anterior cerebral arteries. CT scan with three-dimensional reconstruction helps to define bone defect and to guide the surgery.^[6]

Differential diagnosis of this could be traumatic encephalocoele, ethmoid-frontal sinus mucocoele, neurinoma, hemangioma (the diagnosis becomes obvious if there is a clinical or palpable cerebral pulsation), and glioma.^[7]

Surgery is the only treatment. Surgical Planning and timing of operation are important in the management of anterior encephalocoeles.^[6] Surgery is usually long, thus, blood loss and hypothermia are frequent, which remain the two most important intra-operative complications. If there is no contraindication, surgery is performed and some authors recommend surgery at 8–10 months of age. On the other hand, frontoethmoidal encephalocoeles should be treated at an early age to avoid further facial disfigurement during growth.^[6,7]

A multidisciplinary approach is strongly recommended including Neurosurgeons and Maxillofacial surgeons. Most authors recommend the “combined procedure” which combines a bicoronal and a nasofrontal flap approach with facial reconstruction.^[2] This approach provides three main advantages to surgeons and patients: A more successful closure of the meningoencephalocoele, with lower risk of CSF leak, the possibility of telecanthus correction if necessary, and the reduction of facial scars if necessary. In some cases, only a bicoronal approach can be used to remove the sac and correct the defect.

Table 1: Classification of encephaloceles

Frontoethmoidal	Nasofrontal
	Nasoethmoidal
	Naso-orbital
Basal	Transethmoidal
	Transsphenoidal
	Sphenoethmoidal
	Sphenomaxillary
	Frontosphenoidal/spheno-orbital
Encephalocele of the cranial vault	Interfrontal
	Anterior fontanelle
	Interparietal
	Posterior fontanelle
	Temporal
Occipital	Involves vascular structures
	Posterior fossa
	With cerebellar tissue and ventricular component

Cerebral tissue or meningeal herniations are excluded by electrocautery and the dura is closed and repaired.^[8]

Some authors prefer to limit the repair of the encephalocele through the transfacial approach, without the need of a coronal approach and bifrontal craniotomy, making the procedure less aggressive, however, this can be used only for small malformations.^[9]

Frontoethmoidal encephaloceles often coincide with interorbital hypertelorism. In this case, the medial orbital walls should be surgically replaced, and the reconstruction of both the skull defect and the interorbital hypertelorism can be performed.^[10]

Mahatumarat *et al.* have proposed another procedure, the Chula technique, which has substituted a T-shaped osteotomy of the fronto-naso-orbital bone, in the place of frontal craniotomy. Compared with previous techniques, this one provides a lower risk of cerebrospinal fluid leakage (2.8%) and meningitis (2.8%). In addition, the resected middle portion of the T-shaped bone can be used to augment the nose and decrease the distance between the medial orbital walls.^[10]

Postoperative follow-up is essential because of the risk of CSF rhinorrhea and infection. CSF leaks are the most common postoperative complication.^[11]

The prognosis depends on site, size, content of encephalocele, and any other associated congenital anomaly. The survival rate is higher, nearly 100% in anterior encephalocele compared to posterior encephalocele (55%), where vital structure of brain parenchyma might have herniated through the skull defect.^[11]

CONCLUSION

Encephaloceles are a rare type of neural tube defect caused by herniation of brain tissue through areas of arrested bone development in the skull. Frontoethmoidal encephaloceles are rare and repairing of the defect is mandatory. Approach and timing of surgery have to be carefully planned and cases should be individualized.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- Mahajan C, Rath GP, Dash HH, Bithal PK. Perioperative management of children with encephalocele: An institutional experience. *J Neurosurg Anesthesiol* 2011;23:352-6.
- Suwanwela C, Suwanwela N. A morphological classification of sincipital encephalomeningoceles. *J Neurosurg* 1972;36:201-11.
- David DJ. Cephaloceles: Classification, pathology, and management – A review. *J Craniofac Surg* 1993;4:192-202.
- Tirumandas M, Sharma A, Gbenimacho I, Shoja MM, Tubbs RS, Oakes WJ, *et al.* Nasal encephaloceles: A review of etiology, pathophysiology, clinical presentations, diagnosis, treatment, and complications. *Childs Nerv Syst* 2013;29:739-44.
- Horcajadas A, Palma A, Khalon BM. Frontoethmoidal encephalocele. Report of a case. *Neurocirugia (Astur)* 2019;30:94-9.
- Zabsonre DS, Kabre A, Haro Y. Frontoethmoidal cephalocele: Our experience of eleven cases managed surgically. *Pediatr Neurosurg* 2015;50:7-11.
- Geddard LM, Mahmoud MA, Pan BS, Stevenson CB, Kandil AI. Frontoethmoidal encephalocele: A pediatric airway challenge. *Can J Anaesth* 2018;65:208-9.
- Cohen LM, Jiménez Pérez JC, Holbrook EH, Curry WT, Yoon MK. Meningoencephalocele and cerebrospinal fluid leak complicating orbital decompression. *Ophthalmic Plast Reconstr Surg* 2018;34:e79-81.
- Holm C, Thu M, Hans A, Martina M, Gögeler-Schröder S, Moritz S, *et al.* Extracranial correction of frontoethmoidal meningoencephalocele: Feasibility and outcome in 52 consecutive cases. *J Craniofacial Surg* 2008;121:386-95.
- Mahatumarat C, Rojvachiranonda N, Taecholarn C. Frontoethmoidal encephalomeningocele: Surgical correction by the Chula technique. *Plast Reconstr Surg* 2003;111:556-65.
- Arifin M, Suryaningtyas W, Bajamal AH. Frontoethmoidal encephalocele: Clinical presentation, diagnosis, treatment, and complications in 400 cases. *Childs Nerv Syst* 2018;34:1161-8.