Original Article - Evaluative Study

Management of the Frontoethmoidal Encephalomeningocele

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Abstract

Introduction: The Frontoethmoidal encephalomeningocele (FEEM) is a congenital neural tube anomaly, with herniation of intracranial material such as the brain and leptomeninges through a defect of the dura and anterior skull base at the junction of the frontal and ethmoidal bones. It may result not only in neural defects, sensorimotor deficits, neurological morbidities, visual impairment, impaired nasal function, and a potential risk of intracranial infection, but also in significant craniofacial disfigurement with complex deformities in the frontal, orbital, and nasal regions. **Materials and Method:** The standard two-staged surgical protocol comprises of the first stage performed by a neurosurgeon, which aims at correcting the neural defect by a formal craniotomy; then the second stage performed by a craniomaxillofacial or plastic and reconstructive surgeon, to correct craniofacial hard and soft tissue deformities. The case discussed was managed using a modified intracranial-transcranial single stage approach, achieving both the desired objectives. **Results and Conclusion:** This protocol elucidates the importance and value of teamwork between the Neurosurgeon and Craniomaxillofacial surgeon, in comprehensively and efficiently managing small to moderately sized FEEMs, assuring their complete elimination, satisfactory defect closure, effective functional treatment as well as esthetic correction and reconstruction of the attendant craniofacial deformities by means of a single-stage definitive surgical approach.

Keywords: Encephalocele, frontoethmoidal encephalomeningocele, intercanthal distance, interorbital distance, meningocele, meningoencephalocele

Introduction

Meningoceles, sometimes known by the Latin name "Cranium bifidum," are neural tube defects[1] characterized by sac-like protrusions of the meninges, i.e., the membranes that cover the brain, herniating through openings in the skull, while encephalomeningoceles are protrusions of the brain tissue, cerebrospinal fluid (CSF), as well as the overlying leptomeninges through these cranial defects. Their pathogenesis may be explained by a disturbance in separation of surface ectoderm (epithelial layer) and neurectoderm (nervous tissue) in the midline just after closure of the neural folds.[2] This is regarded as a "late" neurulation defect taking place during the 4th gestational week, occurring due to a failure of fusion during embryogenesis of the cartilaginous neurocranium and the membranous neurocranium or viscerocranium.[3]

A fairly common classification still widely used is that first described by Suwanwela and Suwanwela in 1972, which divides the encephalomeningoceles according to the location as follows:[4,5]

A. Occipital encephalomeningocele (75% of cases)
B. Sincipital encephalomeningocele (10% of cases)
   i. Frontoethmoidal encephalomeningocele (FEEM)
      a. Nasofrontal (which exits the cranium between the frontal and nasal bones and appears at the root of the nose, above the level of the nasal bone)
      b. Naso-ethmoidal (which exits between the nasal bones and nasal cartilage and is located inferior to the nasal bones)
      c. Naso-orbital (which exits through a defect in the maxillary frontal process and causes proptosis and displacement of the globe).
   ii. Interfrontal encephalomeningocele
   iii. Associated with craniofacial clefts
C. Convexity encephalomeningocele (5% of cases)
   i. Parietal encephalomeningocele
D. Basal encephalomeningocele (10% of cases)
   i. Intrasphenoidal

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ii. Transsphenoidal (which herniates in the nasopharynx via a defect posterior to the cribriform plate)
iii. Transethmoidal (which exits through the cribriform plate into the superior meatus, extending medially to the middle turbinate)
iv. Sphenoethmoidal (which exits through the cribriform plate, between the posterior ethmoidal cells and sphenoid to present in the nasopharynx)
v. Frontosphenoidal or spheno-orbital (which enters the orbit via the superior orbital fissure and may produce exophthalmos).

FEEMs manifest as a clinically visible facial mass along the nose, the location and size of which vary depending on the variety. The intracranial root of most FEEMs lies at the foramen cecum, a small ostium located at the bottom of a small depression anterior to the crista galli and formed by the closure of the frontal and ethmoid bones.[6]

Case Report

A 6-year-old male child was brought by his parents with complaints of swelling between the forehead and nose, which had been present since birth but had progressively increased in size to be easily noticeable over the past 2 years. The child also suffered from recurrent attacks of seizures and headaches, vomiting, and urinary incontinence. Weakness of the limbs, uncoordinated muscle movements, vision impairment, and mental or growth retardation or delayed developmental milestones were not observed.

On examination, a 5 cm × 4 cm swelling was seen in the region of the bridge of the nose, extending bilaterally in a bilobulated fashion, and appeared larger on the right side than on the left [Figure 1a]. The swelling was firm in consistency, and there was a palpable impulse on coughing. The soft-tissue mass was accompanied by an obvious hypertelorism, with an interorbital distance (IOD) of 27 mm and intercanthal distance (ICD) of 36 mm.

Computed tomography scans revealed herniation and extrusion of cranial contents including meninges and brain tissue through a defect in the dura and anterior cranial base at the region of the foramen cecum, confirming a frontonasoethmoid encephalomeningocele [Figure 1b and c]. The patient was planned for a single-stage surgical excision of the encephalomeningocele and correction of the craniofacial deformity by a team consisting of a neurosurgeon and a maxillofacial surgeon.

The cranial approach was employed using the standard bicoronal incision extending from the tragus of one side to that on the other. A full-thickness scalp-galeal flap was raised exposing the frontal bone, frontonasal sutures, supraorbital rims and nerves, eye capsules, and the bony orbital funnels bilaterally [Figure 2a-c]. It was extended to a transcranial approach by bringing the bicoronal flap well down over the nasal bones exposing the herniating mass. Pericranium was harvested carefully [Figure 2d] from the reflected flap for later use in bridging the dural defect in the region of the FEEM. A bilateral coronal craniotomy was performed, and the bicoronal calvarial bone flap was carefully elevated [Figure 2e-g], after detaching all adherent dura from its undersurface, exposing the underlying frontal lobes of the brain [Figure 2h and i]. The frontal lobes were carefully retracted epidurally, exposing the large defect in the anterior skull base contiguous with the foramen cecum [Figure 2j]. After identifying the bone defect, the herniated portion of abnormality was opened and noted to contain brain tissues The herniated mass was approached through the extradural route without injuring the dural protective layer, detached, extricated, and excised by the combined intracranial and transcranial approaches, and the dural defect closed by patch duraplasty using the harvested pericranial graft.

This procedure was followed by reconstruction of the craniofacial deformity and correction of hypertelorism by a T-shaped fronto-orbital craniotomy which was performed with the removal of central sections of bone [Figure 2 k-o] to reduce the IOD, bringing the medial walls of both orbits closer together so as to recreate a new appropriate medial IOD. Fixation of bone segments in their new locations was accomplished using titanium micro- and mini-bone plates and screws [Figure 2p]. The upper frontal bar was left after the bifrontal craniotomy and used for plating and screwing the bilateral orbital frames. Cranial bone defect at junction of frontal and ethmoid bones was closed using the excised cranial bone chips. No additional anterior facial incision was employed, and the same bicoronal approach was thus used for the deformity correction as well.

Antiepileptic medication and broad-spectrum antibiotics were prescribed prophylactically. Postoperative recovery of the patient was smooth, uneventful, and complication free, and the results were gratifying with successful management of the FEEM as well as esthetic correction of the hypertelorism with a postoperative IOD of 21 mm and ICD of 27 mm [Figure 3].

Discussion

The craniofacial deformity associated with FEEM cases may consist of hypertelorism, orbital dystopia, elongation of the face, and dental malocclusion. These reflect the distorting influence of the extruded intracranial contents on facial growth.[7] Early removal of encephalomeningocele by the craniofacial route is recommended to allow normal growth forces to be reestablished. In older patients with established deformities, translocation of the orbits may be necessary. IOD is defined as the distance between the orbits measured at their medial margins. ICD is defined as the distance between left and right medial canthi. Both parameters reflect the degree of hypertelorism in the patients.

Comprehensive management of FEEM encompasses the following:[8]

- Accurate diagnosis, delineation of anatomy, and surgical planning
- Single- or multiple-staged surgeries preferably with both a craniomaxillofacial surgeon and a neurosurgeon present

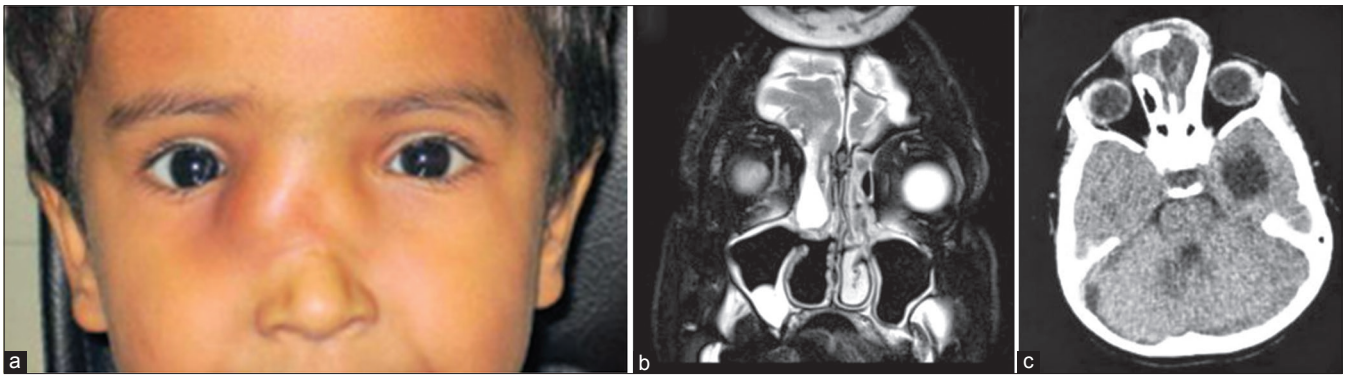

Figure 1: (a) A 6-year-old child presenting with a progressively enlarging swelling at the bridge of the nose which exhibited impulse on coughing and was accompanied by orbital hypertelorism. (b and c) Computed tomography scan revealed herniation and extrusion of cranial contents including meninges and brain tissue through a defect in the dura and anterior cranial base at the region of the foramen cecum, resulting in a frontoethmoidal encephalomeningocele

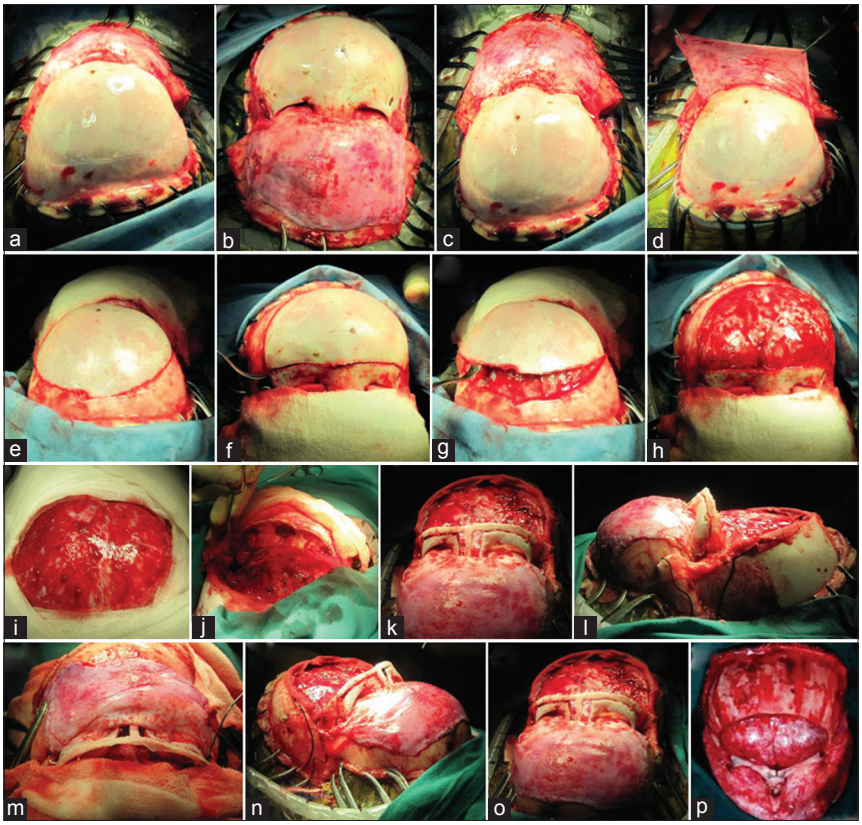

Figure 2: (a-c) Bicoronal incision employed and full-thickness galeal flap raised. (d) Pericranium harvested for bridging dural defect. (e-g) Bifrontal craniotomy. (h and i) Bifrontal bone flap lifted off exposing the underlying frontal lobes of the brain. (j) Frontal lobes retracted epidurally, exposing defect in the anterior skull base. Herniated mass excised and the dural defect closed. (k-o) T-shaped fronto-orbital craniotomy performed with removal of sections of bone to reduce interorbital distance. (p) Fixation using titanium and micro- and mini-bone plates and screws

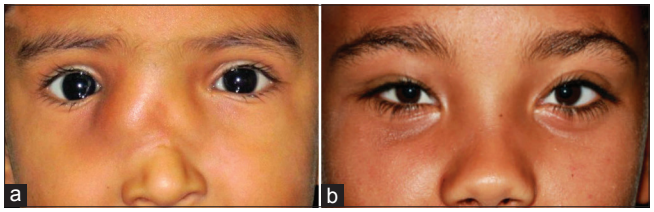

Figure 3: (a) Preoperative appearance of the patient. (b) 2 year Postoperative appearance of the patient showing efficacious surgical management of the frontoethmoidal encephalomeningocele with successful and esthetically gratifying correction of the hypertelorism, with nil facial scarring

- Osteotomies and bone movements that correct all deformities, including the interorbital hypertelorism
- Nasal reconstruction (if required) to address the long-nose hypertelorism deformity
- Skin closure that removes abnormal skin and places incisions in advantageous locations.

Steps generally advocated in the surgical management of FEEM:

- Intracranial/subcranial/transfacial/combined approach for exposure of the anomaly

- Craniotomy/trepanation
- Repositioning the bulging herniated tissue mass back into the cranium
- Surgical excision with removal of nonfunctional extracranial cerebral tissue
- Watertight closure of the dural defect
- Closure of the internal cranial bone defects
- Correction of the craniofacial deformities such as hypertelorism and relieving intracranial pressure that may delay normal brain development
- Nasal augmentation
- Medial canthoplexy
- Placing a shunt if needed.

Comprehensive and holistic management of FEEMs involves both removal of the herniated mass of the encephalomeningocele and reconstruction of the attendant craniofacial deformity caused by this entity and can be achieved by means of two main surgical techniques.[9]

A. Combined intra- and extra-cranial/anterior facial approach includes bicoronal incision, anterior nasal incision, bifrontal calvarial bone flap, nasofrontal bone flap, and facial reconstruction.[9] This is the combined intracranial and anterior (nasal) approach, also known as the nasal-coronal approach, employed for large meningoencephaloceles,[9] in which, in addition to the bifrontal osteotomy, a nasal approach is employed to remove the herniated mass and redundant skin over the mass and also to carry out medial canthopexy and nasal augmentation.[9] Nasal reconstruction may be performed simultaneously to address the long-nose deformity or later during the age of skeletal maturity for a definitive reconstruction. Cranial bone or costochondral grafts with or without fascia or dermis fat graft may be used for the same.

The classic approach of Tessier[10] involves a large bifrontal trepanation/craniotomy of the skull and also detachment, realignment, and re-fixation of the osseous orbits. As the complete rim of the orbit is mobilized to correct hypertelorism, this procedure presumes craniofacial and neurosurgical expertise.[10]

A modification to the Tessier's approach is the Chula technique,[11] in which osteotomy and reconstruction are restricted to the medial portion of the superior orbital rims, upper medial walls of orbits, and nasal bones.[12] To correct the hypertelorism, a central portion of the T-shape bone is removed, and then, the upper parts of the medial orbital walls are moved medially to recreate a new appropriate medial IOD. It is hence less extensive than the standard Tessier's approach, reducing the intraoperative time and blood loss considerably. However, access provided for larger meningoceles may be inadequate.[11]

B. A pure extracranial approach/anterior facial approach[13] is performed in cases of relatively minor craniofacial malformation and the lack of availability of neurosurgical expertise. This technique promises feasibility in institutions with limited resources where it is difficult to perform craniotomy or any neurosurgical procedure.[14] However, this technique proves to have some difficulties and risks. Technical difficulties are caused primarily by the restricted exposure of the neck of the hernia sac, which is limited by the size of the external bony defect, making dural closure much more difficult than when using a frontal osteotomy to directly expose the herniation sac and the surrounding brain. This approach is associated with a higher incidence of CSF leakage than the intracranial technique. This is due to the fact that tearing of the dura is almost unavoidable with a purely extracranial approach in medium or large meningoceles. Furthermore, dural repair is not as sufficient as if an intracranial duraplasty had been performed.[15] Another issue with this technique is the donor-site morbidity associated with closure of the external defect and/or nasal reconstruction.

Some institutions perform a two-staged surgery; the first stage was performed by a neurosurgeon which aims at correcting neural defect by a formal craniotomy; then the second stage was performed by a craniomaxillofacial or plastic and reconstructive surgeon, which aims at correcting craniofacial hard- and soft-tissue deformities, including increased IOD and ICD and nasal deformities. With this approach, there are some limitations including the difficulty to achieve a good esthetic outcome because usually the skin overlying the mass has shown degenerative changes, hyperpigmentation, hyperkeratosis, and significant scarring from the previous surgery.

The case discussed was managed using a modified intracranial–transcranial single-stage approach with no anterior facial surgical exposure. The advantage of this protocol was that performing the procedure with both the neurosurgeon and craniomaxillofacial surgeon present, in one single stage, assured an adequate elimination of the anomaly, complete dural closure, as well as correction of the craniofacial deformity. In this case, it was possible to completely eliminate the use of a facial incision as the size of the herniation was not large and the simple protrusion of brain and meninges allowed the skin to retract after the mass was excised from beneath, giving a better cosmetic result. On the other hand, large lobulated masses that are filled with both brain and CSF unfortunately require a facial incision, leaving the patient with a rather prominent scar along or across the bridge of the nose.

A watertight and durable closure of the dural defect could be achieved using the autologous pericranial graft harvested while reflecting the scalp flap, thus preventing and safeguarding against postoperative complications, such as meningitis, epidural abscess, CSF leak, and brain herniation. This was further reinforced by reconstructing the defect in the anterior cranial fossa base with an autologous bone graft. We used calvarial bone chips that had been sectioned from the frontal bone, as a graft to bridge the cranial defect at the herniation site in the anterior cranial fossa, as the design of the T-osteotomy (performed to correct the hypertelorism), provided adequate graft material needed for the closure without entailing

need for any separate donor site. Furthermore, the craniofacial deformity could be satisfactorily addressed definitively in the same surgery, without the need for a secondary or late corrective surgery. Moreover, the incisions placed (being restricted to the extended bicoronal incision) were camouflaged within the hairline and the preauricular crease and were thus esthetically pleasing, avoiding any scarring/disfiguration of the prominent forehead and nose regions.

Conclusion

Successful treatment of the FEEM depends on a thorough understanding of its pathological anatomy (site of herniation, such as the junction of the frontal and ethmoid bones; coexisting craniofacial deformities such as interorbital hypertelorism; and possible presence of secondary trigonocephaly); careful planning of bone movements to correct these deformities; attention to detail while planning incisions for ideal placement of scars; ideal positioning of the medial canthi; and an esthetic nasal reconstruction. The ability to eliminate the use of a facial incision, and the resultant scarring, is more dependent on the type and size of the pathology, rather than the surgeon's experience or expertise, as large lobulated masses filled with both brain and CSF unfortunately necessitate a facial incision for successful and complete excision.

In small to moderately sized FEEMs, we recommend a comprehensive and definitive one-stage repair via a combined intra- and trans-cranial surgical approach with craniofacial reconstruction to remove the herniated mass, to repair dural, and bone defects, to reconstruct the naso-orbital area, to correct the associated craniofacial deformity such as hypertelorism, and to restore aesthetic facial appearance, by a teamwork approach comprising of a neurosurgeon and a craniomaxillofacial surgeon. Nevertheless, this study is still a preliminary one, thus needing a long-term evaluation of more number of patients managed for different varieties and sizes of FEEMs.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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