Single stage repair & reconstruction of trigonocephaly & fronto orbito ethmoidal encephalocele

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Abstract
A nine-year-old Iranian girl presented with trigonocephaly and frontonasal deformity secondary to encephalocele of fronto orbito ethmoidal region. She had undergone an attempt to correct the deformity at the age of three. The case was managed by a single stage repair and reconstruction of both fronto orbito nasal deformity and trigonocephaly along with plication of the herniating encephalocele.

Fronto nasal deformity, interorbital hypertelorism, nasal deformity and associated trigonocephaly are the main aesthetic concerns in the management of craniofacial disfigurement associated with fronto ethmoidal encephaloceles. A single stage repair and reconstruction of these cases is always preferred. *(p94-97)*

***Key words**: Fronto ethmoidal encephalocele, fronto orbital, nasal deformity, encephalocele, trigonocephaly and hypertelorism*

Introduction

Encephaloceles are developmental malformations of the central nervous system due to the defective differentiation of the surface ectoderm and neuroectoderm. This results in herniation of intracranial contents through the osseous defect at different locations of the skull. Encephaloceles of the anterior cranial fossa are mainly divided into sinicipital and basal encephaloceles. These can be further classified depending on the location of the bony defect and the course of the encephalocele.[16] In south-east Asia encephaloceles show a relatively high incidence (one in 5000 population) when compared to western countries (one in 40,000).[2,13,14] Increased rate of consanguineous marriages is believed to be the cause of the high incidence of congenital deformities in certain regions.

A fronto naso ethmoidal encephalocele is formed by herniation through a congenital bone defect at the junction of the frontal and ethmoidal bones. Naso orbital and naso ethmoidal encephaloceles have sacs with long necks, adding to the difficulty of obtaining adequate closure at the internal orifice when approaching from below.[16] Moreover, the combination of interorbital hypertelorism, nasal deformity and trigonocephaly in such patients is to be addressed along with other facial deformities.[3] Here a case of encephalo meningeocele of fronto orbito ethmoidal area associated with a short nose deformity and trigonocephaly is presented, that was managed by a single stage repair of both fronto orbito nasal deformity and trigonocephaly along with plication of the encephalocele.

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Case Report

A nine-year-old girl, the first child of consanguineous Iranian parents, presented with a prominent bulge at the glabellar region involving the medial orbital wall and the roof of left orbit.

Parents gave a history of an extra cranial surgical attempt to correct the deformity of fronto nasal region during her early childhood. There is no available record of the previous surgery. As per the parents, the deformity was partially corrected by this procedure leaving a scar at the glabellar region. The residual swelling gradually increased in size eventually resulting in a short nose deformity. The child was more concerned about the short nose deformity than the frontal swelling which is presently rather static.

Physical examination revealed a bony hard protuberance at

the glabellar region of a trigonocephalic head extending more towards the medial wall and roof of the left orbit. Even though moderate interorbital hypertelorism and orbital dystopia were evident, the patient did not complain of double vision. There was no complaint of anosmia. Nasal deformity was marked with a short nose and loss of the naso frontal angle along with collapse of the nasal tip. An oblong hypertrophic scar resulting from the previous surgery was present at the left medial canthal region.

Figure 1 - Preoperative photo-showing orbito frontal swelling of trigonocephalic head

A wide bony defect at the orbital roof was palpable extending posteriorly. The left globe was displaced laterally. However there was full range of ocular mobility with no evidence of diplopia or exophthalmia. A soft fluctuant mass was evident on palpation deep to the bony defect protruding to the orbit. An interorbital hypertelorism of 35 mm and inter papillary distance of 64 mm were measured. The collapse of the nasal tip resulted due to lack of osseo-cartilaginous support. However, the nasal passages were not obstructed.

Computed tomography (CT) revealed a 27 mm bony defect involving the left orbital roof medially along with deformed, medially displaced ethmoid air cells. A soft tissue density was seen protruding through it. Deformity of the medial wall of the orbit flattened the nasal bridge resulting in inter orbital hypertelorism. The intracranial measurements of clino clival angles were not possible to prove trigonocephaly due to the deformity of the bone contour at the naso frontal region.[17] However, CT images confirmed the trigonocephaly.

Magnetic resonance imaging (MRI) showed herniation of meninges through the orbital roof defect with no other intracranial pathology or hydrocephalus.

Surgery

The case was managed by a single stage repair and reconstruction of the orbito fronto nasal defects, correction of trigonocephaly and septorrhinoplasty along with plication of the encephalocele sac. A coronal scalp flap access gained to elevate bifrontal bone flaps. Further access to the base of the encephalocele sac was attained by fronto naso orbital bandeau osteotomy. Hernial sac amputation and dural repair was done followed by orbital roof reconstruction using calvarial bone grafting. A water tight closure of the dura and the cranium was gained by BioGlue (bovine serum albumin and glutaraldehyde) application. The fronto orbital bandeau remodelling to correct the trigonocephaly was done by inner lamina coring and digital reshaping. The nasofrontal angle was created by remodelling the glabellar area and the minimal hypertelorism was corrected by lateral nasal osteotomy. A medial canthopexy after excising the hypertrophic scar of the previous surgery corrected the canthal positions. A split thickness calvarial bone graft was used to regain nasal form as well as to support the collapsed nasal tip. The augmentation septorrhinoplasty was performed via two lateral nasal incisions. The nose was splinted by nasal splints and an anterior nasal pack using bismuth iodine paraffin pack. The pack was kept for four days and the nasal splint removed after ten days. Postoperatively, patient was nursed in our Pediatric Intensive Care Unit for two days and discharged home after one week of an uneventful postoperative period.

Discussion

Although the cause of encephaloceles remains unclear, it appears to involve a variety of genetic as well as environmental factors.[4] Relatively high incidences of fronto ethmoidal encephaloceles are reported in south-east Asia with possible aetiologic factors like increased consumption of aflatoxins and or possibly folic acid deficiency.[1,8,12] Congenital deformities are increasingly reported in consanguineous parents and the high rate of consanguineous marriages is reported in some tribes of Iran. However, a negative history of consuming folic acid had the most significant statistical correlation with malformations associated with neural tube defects.[10]

As we do not have the details of the previous surgery performed for this patient the exact mode of initial presentation is not clear. The bony defect mainly involved the orbital roof extending towards the medial wall. Most frontonasal encephaloceles present between nasal and frontal bones and its sub division naso orbital group is usually seen in the medial wall of the orbit. However, many variations of these subdivisions are also possible.[15]

The association of encephaloceles in craniosynostosis is not widely reported, however the obvious trigonocephaly is

noted in most patients in a series of 35 patients by Holmes, et al.[5] The metopic ridging in Chiari I malformation is reported explaining the increase in the intracranial pressure in sub clinical metopic craniosynostosis with or without signs of trigonocephaly.[5,6] As the details of this patient's neonatal care was missing the possibility of a clinically established craniosynostosis cannot be ruled out but intraoperative findings of metopic ridging and the trigonocephaly may support the hypothesis stated. Holmes states that the trigonocephaly may be due to metopic synostosis induced secondarily during development.[6,9]

There is enough evidence to prove the "long nose" deformity associated with early correction of the interorbital hypertelorism associated with fronto ethmoidal encephaloceles especially in a patient with long midface.[5] Ortiz-Monasterio and Fuente del Campo first coined the term long nose and short nose deformities.[11] A short collapsed nose was the primary complaint in this patient. The nasal deformity, possibly secondary to the lack of septal development, is pointing to the evidence of nasal deformities being associated with neural tube defects. The loss of frontonasal angle due to the prominent frontonasal bulge worsened the deformity. This, in turn, is the reason for the interorbital hypertelorism seen in most cases where a true orbital hypertelorism is not appreciated.

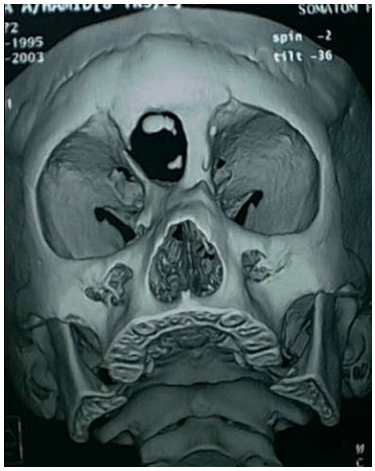

Figure 2 - 3D CT showing the orbito frontal bony defect

Primary treatment of encephaloceles is surgical mainly to prevent the damage of the herniated brain mass and to prevent the secondary aesthetic and functional deformity. The risk of intracranial spread of infection is higher in open encephaloceles when compared with closed ones.[7] Most researchers agree that the best time for the correction of the encephalocele is early infancy.

Surgery primarily aims at watertight closure of the defect of the dura after the plication of the herniated brain followed by aesthetic and functional reconstruction of the primary and secondary deformities. Naso orbital and naso ethmoidal encephaloceles have sacs with long necks, adding to the difficulty of obtaining adequate closure at the internal orifice when approaching from below.[16] This most often necessitates a transcranial approach especially in orbital and ethmoidal encephaloceles. A sub cranial approach or a combined one is preferred in only a few cases.

Most authors prefer to manage the aesthetic and the functional deformities as a single stage procedure. Primary advantage is that it reduces the cost and effort of surgeries avoiding stress due to multiple procedures. Single stage procedure requires a team approach from neurosurgery, cranio facial surgery and anaesthesia. This allows surgical team to plan the reconstruction by analyzing spatial relation of the facial skeletal to anterior skull base. Moreover, no additional surgery is required for harvesting calvarial cantilever graft for nasal reconstruction and the coronal incision provides enough access for the frontonasal area reconstruction.

The orbito frontal bandeau osteotomy after the bifrontal bone flap helped the surgeon to gain ample access to the base of herniating sac. This also assisted in a watertight repair of the dura after effective reduction and plication of the herniated mass. In addition to this, application of BioGlue (bovine serum albumin and glutaraldehyde) over the repaired dura added in preventing a CSF leak. A split thickness inner lamina calvarial graft was preferred over other modalities in reconstructing the orbital roof, this graft being of similar thickness and morphology.
Anatomical reconstruction of the fronto orbital bandeau

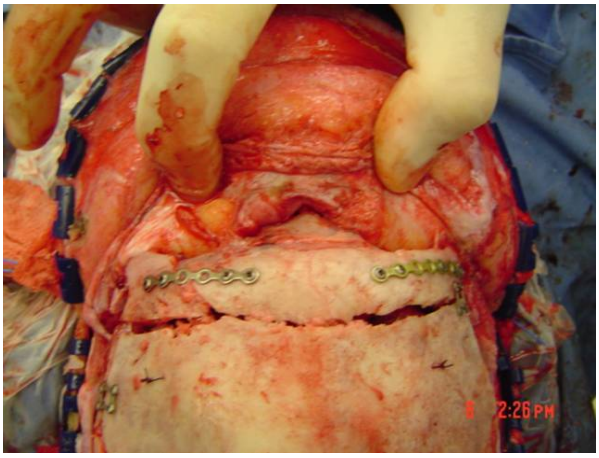

Figure 3 - Remodelled fronto orbital bandeau with plate osteosynthesis

after digital remodelling following inner laminar coring is an effective method to correct the frontal deformity of trigonocephaly. Correction of the hypertelorism was gained by a simple technique, as it was not marked after excision of the herniated mass. The frontonasal angle was created by remodelling the glabellar region and the placement of a cantilever nasal bone graft using split thickness calvarial bone. This helped to have a nasal tip projection and support to the nasal cartilages keeping the nasal valves patent.

Figure 4 - Postoperative right lateral view

The excision of scar tissues of the glabellar region and the scar revision at the medial canthal region added to maintain good frontonasal form and function. The bilateral lateral nasal incisions were unavoidable to osteotomise the rudimentary nasal bones. Strict asepsis was maintained throughout the procedure to prevent intracranial infection from the nasal cavity and each compartment was addressed separately providing separate set of instruments for nasal and cranial procedures.

Conclusion

The association of trigonocephaly, interorbital hypertelorism, nasal deformity and facial disfigurement were the main aesthetic concerns in the management of craniofacial deformity associated with encephalocele in this case, which was presented as a sequel following an inadequate treatment. The orbito frontal bandeau taken for correction of trigonocephaly gained more access to the neck of the hernial sac rendered secure dural repair assuring CSF seal. This helped to attain successful outcome in a single stage surgery addressing all the problems.

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