Successful One Stage Operation for Frontonasal Encephalocele in very Young Children 2 Case Report

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Abstract:

Encephalocele is defined as protrusion of cranial contents (meninges and cerebral tissue) beyond the normal confines of the skull through a defect in the cranium. It is one form of a neural tube defect as are anencephaly and spina bifida . There are 2 main types of encephalomeningocele, frontoethmoidal and occipital, according to the location of the defect. The frontoethmoidal type defect, which is located in the area of the frontal and ethmoidal bones, The authors present 2 cases of frontonasal encephalocele in very young children.

Key words: *Frontonasal encephalocele Children One stage operation*

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Encephalocele is defined as protrusion of cranial contents (meninges and cerebral tissue) beyond the normal confines of the skull through a defect in the cranium. The population incidence of this congenital anomaly is estimated to vary from 1 per 300 to 1 per 10000 live births.[1-4] In respect to the incidence, cranial dysraphism, particularly encephaloceles, is far less common compared to its spinal counterpart, namely, myelomeningocele, accounting for only 8-19% of all dysraphism.[4-9]

It is one form of a neural tube defect as are anencephaly and spina bifida [10]. There are 2 main types of encephalomeningocele, frontoethmoidal and occipital, according to the location of the defect. The frontoethmoidal type defect, which is located in the area of the frontal and ethmoidal bones, is exclusively common in Southeast Asia [11]. There has been no consensus about the etiology, prognosis-related classification, or surgical strategy for encephalomeningoceles.

The aim of surgical treatment is to restore the functional brain tissue in the cranial cavity,perform dural repair,correct bone defect and restore esthetic facial appearance safely and successfully in a singe stage. We present 2 cases of successful one stage operation for frontonasal encephalocele in very young children.

Case report 1:

History,presentation and examination: An 11-month-old male child was admitted to our hospital,presented with a protruding soft swelling with a wide base between the eyes (Fig. 1). According to his mother's description, he had been born with the facial swelling. It had slowly enlarged as he grew up.There were no complaints with regard to vision. No congenital anomalies were mentioned in his siblings.

The physical examination revealed nonpulsatile swelling near the left side of the nasal bidge and left medial canthus with underlying bony defect,interorbital hypertelorism, long-nose deformity,and medial canthal dystopia. The neurological status revealed no abnormalities.

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Fig. -1: *Preoperative photograph of 11-month-old male child with frontonasal encephalocele*

Fig.-2: *Postoperative photograph (after 6 weeks of operation)*

Neuroimaging studies: He was investigated with CT scan of the head that showed a defect in the anterior cranial base involving the crista galli and medial wall of the left orbit. The defect directly communicated with the anterior cranial fossa through which herniated brain could be seen extending in to the soft tissue area of left medial canthus.

Fig. 3: *3D CT scan of head Showing external bone defect*

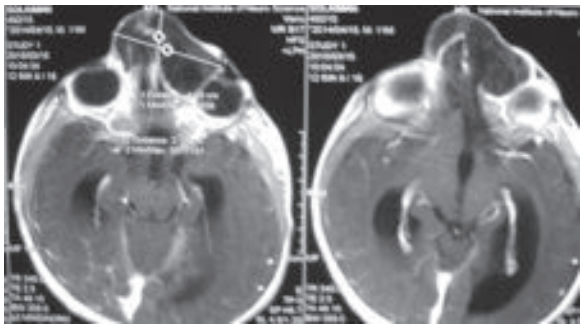

Fig. 4: *CT scan axial view showing anterior cranial base defect*

Operation and postoperative courses: : In this cases a combined approach was used and scheduled for single stage surgery for repair of dura. Craniotomy and repair from inside was done. This operation was performed in collaboration with the Maxillofacial surgeon.

We made a bicoronal scalp incision down to the anterior of the tragus. A paranasal skin incision was made in a reversed Y-shape because of the bilateral herniated masses in order to remove any redundant skin.The scalp was reflected to expose frontal bone,both supra orbital rims nasal bridge with bony defect. A typical bifrontal craniotomy and a Tshaped frontonasal osteotomy were done to obtain adequate exposure to the encephalomeningoceles. Supra orbital rim was removed. A satisfactory extradural exposure all around the herniating glioting brain was thus achieved.The margins were freed all around the bony defect.The skull defects over the inner wall of the orbit and ethmoidal sinus were located, and the herniated dura sac and the degenerative brain tissue it contained were resected. Watertight and durable closure of the dural defect was achieved by an autologous pericranial graft and artificial dura. The nasal bridge was

reconstructed with split frontal bone. All areas were fixed with 2-0 prolene. The medial canthus was repositioned with prolene 3-0 stay sutures.

Fig.-5: *Intraoperative photograph*

After achieving haemostasis,wounds were closed in layers.

Postoperatively,patient recovered uneventfully except there was subgaleal CSF collection in postoperative period (3rd POD) and managed by aspiration with pressure bandage.

A good cosmetic result had been achieved after operation (Fig. 2)

Case report 2:

History,presentation and examination: A 9-month-old female child was admitted to our hospital, presented with a protruding soft swelling with a wide base between the eyes (Fig. 6 and 7). According to his mother's description,she had been born with the facial swelling. It had slowly enlarged as she grew up.There were no complaints with regard to vision. No congenital anomalies were mentioned in his siblings.

The physical examination revealed nonpulsatile pedunculated swelling near the nasal bidge,between the medial canthus with underlying bony defect,interorbital hypertelorism, long-nose deformity,and medial canthal dystopia. The neurological status revealed no abnormalities.

Neuroimaging studies: She was investigated with CT scan of the head that showed a defect in the anterior

Fig. 6,7: Preoperative photograph of 9-month-old female child with Frontonasal encephalocele

Fig.8: *Postoperative photograph (after I month of operation)*

cranial base involving the crista galli and between frontal and nasal bone. The defect directly communicated with the anterior cranial fossa through which herniated brain could be seen extending through nasal bridge.

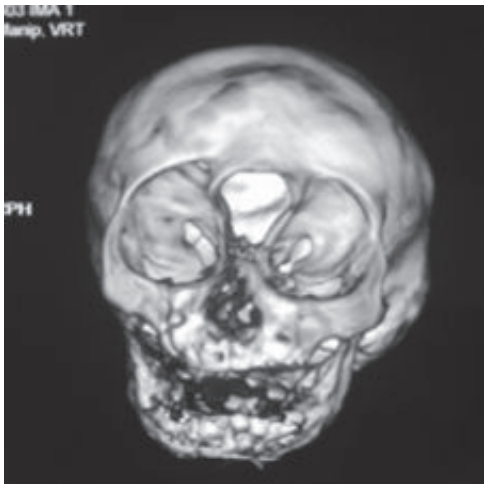

Fig.-9: *3 D CT scan head external bone defect*

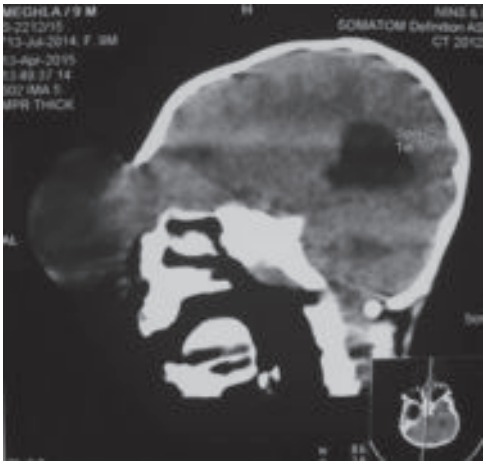

Fig.-10: *CT scan head showing bony defect*

Operation and postoperative courses: In this cases a combined approach was used and scheduled for single stage surgery for repair of dura. Craniotomy and repair from inside was done. This operation was performed in collaboration with the Maxillofacial surgeon.

We made a bicoronal scalp incision down to the anterior of the tragus. A paranasal skin incision was made in a reversed Y-shape because of the bilateral herniated masses in order to remove any redundant skin.The scalp was reflected to expose frontal bone,both supra orbital rims nasal bridge with bony defect. A typical bifrontal craniotomy and a Tshaped frontonasal osteotomy were done to obtain adequate exposure to the encephalomeningoceles. Supra orbital rim was removed. A satisfactory extradural exposure all around the herniating glioting brain was thus achieved.The margins were freed all around the bony defect.The skull defects over the inner wall of the orbit and ethmoidal sinus were located, and the herniated dura sac and the degenerative brain tissue it contained were resected. Watertight and durable closure of the dural defect was achieved by an autologous pericranial graft and fibrin glue. The nasal bridge was reconstructed with split frontal bone. All areas were fixed with 2-0 prolene. The medial canthus was repositioned with prolene 3-0 stay sutures.

After achieving haemostasis,wounds were closed in layers.

Fig.-11: *Intraoperative photograph*

Postoperatively,patient recovered uneventfully except there was CSF leakage at nasal bridge through single stitch point in postoperative period (12 th POD) and managed with use of acetazolamide and single secondary suture.

A good cosmetic result had been achieved after operation (Fig. 8)

Discussion:

From the anatomic aspect, the most common sites for encephaloceles are occipital and frontonasal regions. In Asia and Africa, there is a predominance of the frontonasal group while 80-90% are found in the occipital region in the Western Hemisphere. Approximately 70% of occipital encephaloceles occur in females, but there is no sex predominance noted in the frontobasal type. The incidence of hydrocephalus in patients with encephaloceles is reported to be about 50%. In planning the strategy of management of encephalocele, one needs to take into consideration the site, size, contents, state of CSF pathway, neurological status, associated anomalies and overall general condition of the patient.

In both our cases,there was a fronto orbitonasal defect extending posteriorly up to crista galli with hypertelorism and cosmetic deformity.

The principle of repair is analogous to the management of hernias in general surgery, which includes dissection of the sac, isolation of the neck, adequate closure at the neck and reinforcement. The herniated part of the brain is usually gliosed and non-viable and can usually be safely amputated. Dural defect should be closed in a watertight fashion, using graft if necessary. In our cases, watertight and durable closure of the dural defect was achieved by an autologous pericranial graft and fibrin glue. Ideally, reinforcement of bony defect with bone graft (split cranium, split rib, or acrylic) will prevent reprotrusion through the defect. Reconstruction of bony abnormalities may be necessary at times for better cosmetic results. Associated hydrocephalus should be treated by shunting before managing the encephalocele. As mentioned before, surgical approaches for encephaloceles, based on its location and type, can be direct, indirect or both. In 2 cases,our operative approach involved combined approach (bifrontal craniotomy and direct repair) and were performed in collaboration with the Maxillofacial surgeon. We performed a typical bifrontal craniotomy with a T-shaped osteotomy for 1-stage reconstruction and obtained adequate exposure in order to perform dura repair and the encephalomeningocele resection.

The end result of encephalocele surgery is usually not determined by the neurosurgical procedure per se, but by the underlying brain involvement and presence or absence of other congenital defects. In long-term follow up, cases with anterior defect have better prognosis and more than half have normal intelligence quotient (IQ).[12]

Instead of the traditional 2-stage correction by preliminary disconnection and subsequent extracranial correction of the facial deformity [13], a 1- stage operation has become the standard treatment [14, 15]. Most Neurosurgeons and craniofacial surgeons prefer the combined nasal-coronal approach with a frontal craniotomy because of the wide exposure [14]. On the other hand, the frontal bone flap can also be remodeled to eliminate the trigonocephalic bulge [16], repair any external skull defects, and restore an esthetic appearance such as with nasal augmentation.

Conclusions:

Encephalocele is a relatively uncommon neurosurgical entity largely seen in the pediatric population. Treatment of this condition can be rewarding if properly managed early. Occipital type may be approached without opening the cranium, while sincipital and basal encephaloceles usually require craniotomy. In this paper we present our experience in the operative management of encephaloceles with good outcome and also share our recommendation in technical consideration for surgical approaches.

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