

Fronto-nasal Encephalocele and Hypertelorism in a Newborn: Surgical Management and Case Report

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Abstract: Encephalocele is described as a protrusion of cranial contents (meninges and brain tissue) through a congenital or acquired defect of the skull. Congenital is due to a multifactorial neural tube defect. In the prenatal period, the diagnosis is made by ultrasound and in the postnatal period by tomography and / or magnetic resonance imaging. The goal of surgical treatment is to reduce brain herniation and obtain a sealed closure of the dura and a proper coverage of the skin. In this paper we describe a case of fronto-nasal encephalocele and hypertelorism and surgical management.

Keywords: Encephalocele, Neural tube defect, Hypertelorism, Surgical management.

INTRODUCTION

Encephalocele is described as a protrusion of cranial contents (meninges and brain tissue) through a congenital or acquired defect of the skull [1]. In our case we refer to the birth defect.

Anterior encephaloceles are rare conditions. Its incidence in the western hemisphere varies from 1/35,000 to 1/40,000 live births, but in Asian countries its incidence increases to 1/5000 live births [2]. As it is known, a defect of the neural tube is produced. The cause of the defect is the failure of early separation of the ectoderm surface in the embryo development.

There is evidence that supplementation of folic acid significantly reduces the occurrence of neural tube defects. Supplementation is recommended at least four weeks prior to conception, and continued until the end of the first trimester [3-5].

75% of encephaloceles are occipitally located and 25% are anteriorly located. The diagnosis of encephalocele is usually performed by prenatal ultrasound and tomography and / or magnetic resonance imaging in the post-natal period. Anterior encephaloceles are classified as frontal, sincipital and basal [6].

CASE REPORT

Newborn male patient product of the second pregnancy of a 30 year old patient whose previous birth

was 14 years ago. The gestational age was 39 weeks. Her pregnancy was controlled from the eighth week in primary care medical center. As the only significant maternal history there is a diet low in folate. The last control before delivery was performed in our institution of III level of complexity and it was decided to schedule cesarean. Physical examination of the newborn showed fronto-nasal protrusion of 2 cm in diameter (Fig.1A) and bilateral equinovarus foot. Magnetic resonance imaging was performed which diagnosed fronto-nasal encephalocele (Fig.1B and 1C). Surgery was decided together with maxillofacial surgery.

Patient supine and in neutral position, anesthetic induction and intubation is performed.

A bicoronal skin incision was marked to gain wide exposure (Fig. 2A). A scalp flap was raised and at the same time, a pericranial graft was also harvested to repair the dural defect. The scalp was reflected to expose frontal bone, both supraorbital rims, and the nasal bridge (Fig. 2B). The frontal sinus was not well developed and did not pose much of a problem in performing the craniotomy. A bifrontal craniotomy and bilateral supraorbital orbitotomy were performed (Fig. 2C and 3A). The frontal bone flap was removed and a midline piece of frontal bone was removed separately, leaving the lateral part of the nasal bones intact with supraorbital ridges on both sides. A thin rim of frontal bone was left attached on both sides to the cranium in order to facilitate the fixation of bone flaps. The basal

dura was separated all around the herniating gliotic brain, and the defect was defined through brain tissue that was herniating in the nasal cavity. Redundant brain tissue was excised and dural margins were freed all around. The dural defect was closed with free pericranial graft using prolene 5-0 in a water-tight manner. Heterologous patch of dura is positioned on autologous plasty as a measure of reinforcement to prevent cerebrospinal fluid (CSF) leak. A piece of mid frontal bone, which was removed separately, was used as a split graft to reconstruct the anterior cranial fossa

floor. Correction of hypertelorism was performed by shifting both the supraorbital ridges medially and fixing with miniplates (Fig. 3B). To provide a firm and immobile skeleton, the supraorbital ridges and frontal bone flap were fixed to the bar of the frontal bone which was left attached to the cranium. Redundant skin of the nasal region was resected. After achieving haemostasis, the incision was closed in layers (Fig. 4A and 4B). There were no intraoperative or postoperative complications. Prophylactic antibiotic was left for 3 days and the patient was discharged on the seventh day.

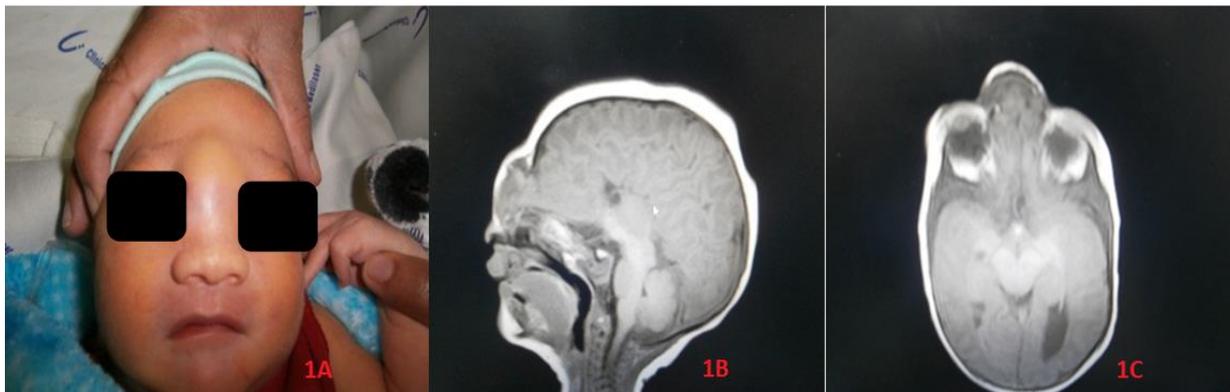


Fig. 1A: Anterior view showing newborn fronto-nasal encephalocele and hypertelorism; **1B and 1C:** MRI brain: Sagittal and axial view evidence fronto-nasal encephalocele.

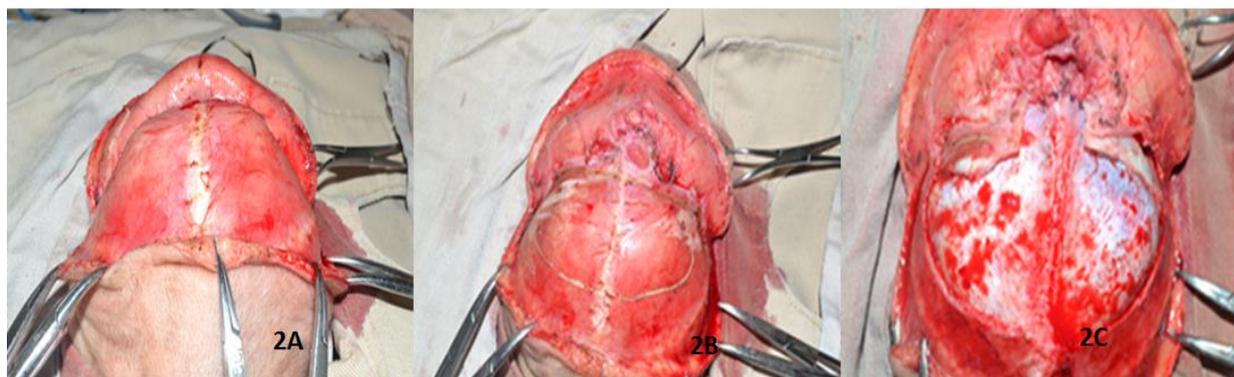


Fig. 2A: Bicoronal approach, flap exposure and midline demarcation; **2B:** Bifrontal craniotomy; **2C:** Osteotomies of the medial walls of the orbits and Post resection of fronto-nasal encephalocele and duraplasty.

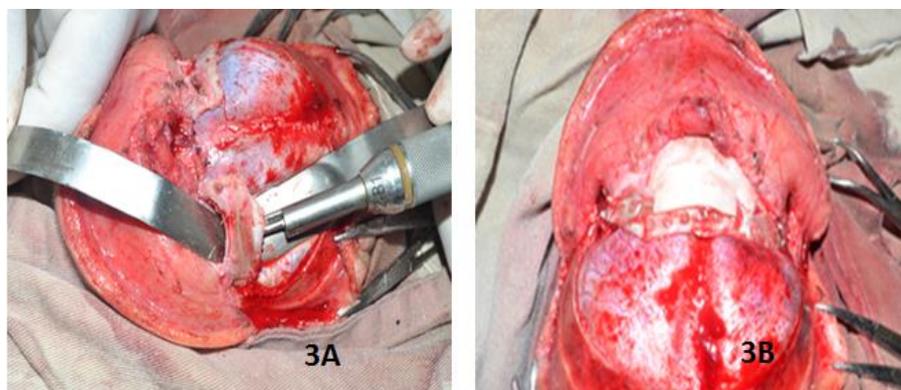


Fig. 3A: Orbital osteotomies, brain spatulas protecting the eyeball and the dura; **3B:** Hypertelorism correction with miniplates and miniscrews. Synthetic patch of dura placement on prior autologous plasty to prevent CSF leak.

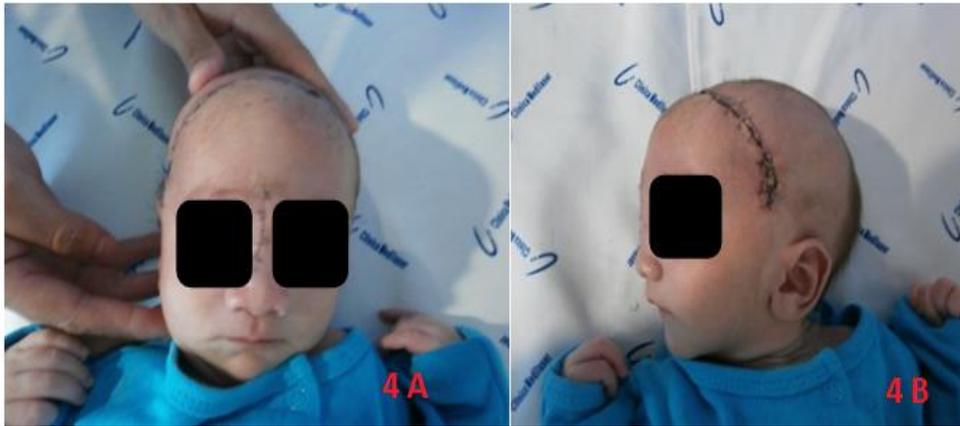


Fig- 4A and 4B: Anterior view and Lateral View of surgical wound

DISCUSSION

The incidence of anterior encephaloceles is very rare in our country. It is estimated in 1/35.000 to 1/40.000 live births in the western hemisphere. They are associated with hypertelorism, agenesis of the callosum corpus and heterotopias [7, 8]. It has been demonstrated that administration of folic acid in the preconception period and during pregnancy reduces the risk of this type of defect of the neural tube closure. The diagnosis is made by ultrasound in the pre-natal period and tomography and/or magnetic resonance imaging in the post-natal stage. If we know the diagnosis in the pre-natal period, we prefer that the delivery is cesarean to avoid infection and trauma resulting from vaginal delivery.

The risk of difficult intubation is higher in children with congenital malformation [9]. The location of the encephalocele precluded routine facemask ventilation. 75% of encephaloceles arise from the occiput and can cause restriction of head movement; may be associated with challenging direct laryngoscopy, whereas frontonasal and frontoethmoidal encephaloceles may be problematic to mask ventilate in up to 19% of the patients [10].

The inherent implications of pediatric anesthesia and difficult airway make surgical correction challenging for anesthesiologists. Difficult airway is not the only concern in children with encephalocele, but found to be associated congenital malformations, hydrocephalus, large size of sac, and hemodynamic disturbances that require careful considerations [11].

We used sevoflurane for induction and maintenance of general anesthesia because of getting rapid induction and not being irritable.

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. 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 [12].

The treatment of this condition is based on the surgical correction of the defect, the primary goal is the reduction of cerebral herniation, the seal of the dura to prevent Cerebrospinal fluid (CSF) leak and resection of redundant skin defect [13]. In this clinical case, correction of hypertelorism was performed too.

CONCLUSION

In this report we have described a case of fronto-nasal encephalocele and hypertelorism and surgical management.

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