
Sincipitalencephalocele: A case report

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Abstract

Encephalocele is a congenital anomaly characterized by herniation of brain and meninges through a defect along the midline of the cranial vault or at the base of skull. It occurs in 1–4 cases per 10,000 live births. The commonest location is occipital (75%) whereas frontonasal location is rare. We report a case of four months old male baby who presented with gradually progressive swelling at the root of nose since birth. Initially, it used to increase in size on crying but since 1 month there was no change in size noticed. On examination a midline solitary swelling of 8×10cm size in the frontonasal region was noted. It was non tender, cystic, non-pulsatile, non-transilluminant, without cough impulse, non-reducible with stretched hyperpigmented overlying skin and normal surrounding skin. Underlying bone defect could not be made out. Intraoperatively, there was 5×3 cm bony defect in anterior half of cribriform plate up to frontonasal junction through which herniation of the frontal lobe covered by thinned out duramater was noted. Gliotic changes were seen at the tip of herniated brain matter. Bifrontal craniotomy followed by circumferential dissection and repositioning of herniated frontal lobe was done. Primary dural closure with excision of redundant skin sac was done. Postoperative period was uneventful.

Keywords: encephalocele, neuroschisis, neurulation defects, sincipital

1. Introduction

Encephalocele is a congenital anomaly characterized by herniation of brain and meninges through a defect along the mid line of the cranial vault or at the base of skull.[1] This is far less common than spinal dysraphism. It occurs in 1–4 cases per 10,000 live births.[2] Commonest site is occipital (75%), followed by frontoethmoidal (13% to 15%), parietal (10% to 12%) and sphenoidal(2%). Occipital encephalocele is common in western hemisphere where as sincipitalencephalocele is common in South East Asia.[3]

2. Case Report

A four months old male baby presented to us with swelling at the root of nose since birth. Initially, it was about 3 cm in size which gradually increased to attain a size of 8×10 cm. There is a history of increase in size of the swelling on crying which is not being noticed since 1 month. There is no history of seizures, vomiting, trauma or secondary

changes in the swelling. There is a history of 2nd degree consanguineous marriage between parents. Mother underwent regular antenatal surveillance and had full term normal delivery. Baby had a birth weight of 2.7 kilograms and he cried immediately after birth. Developmental milestones were normal. On examination, baby was stable clinically and neurologically. On local examination, a solitary midline swelling measuring 8×10cms was present at the root of the nose with a base measuring about 3 cms. It was non tender, cystic, non-pulsatile, non-transilluminant, without cough impulse, non-reducible with stretched hyperpigmented overlying skin and normal surrounding skin. Underlying bony defect could not be made out. Interpupillary distance was increased(55mm).

Fig 1.1: Lateral view of frontonasalencephalocele



Fig 1.2: Anterior view of frontonasalencephalocele



Fig 2.1: CT scan sagittal view showing the frontonasal defect

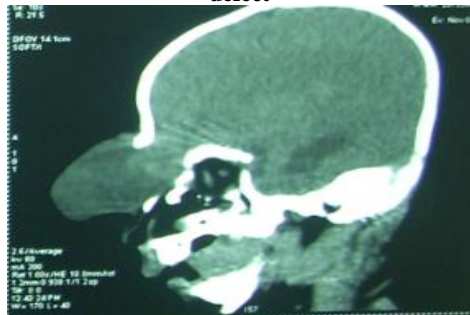


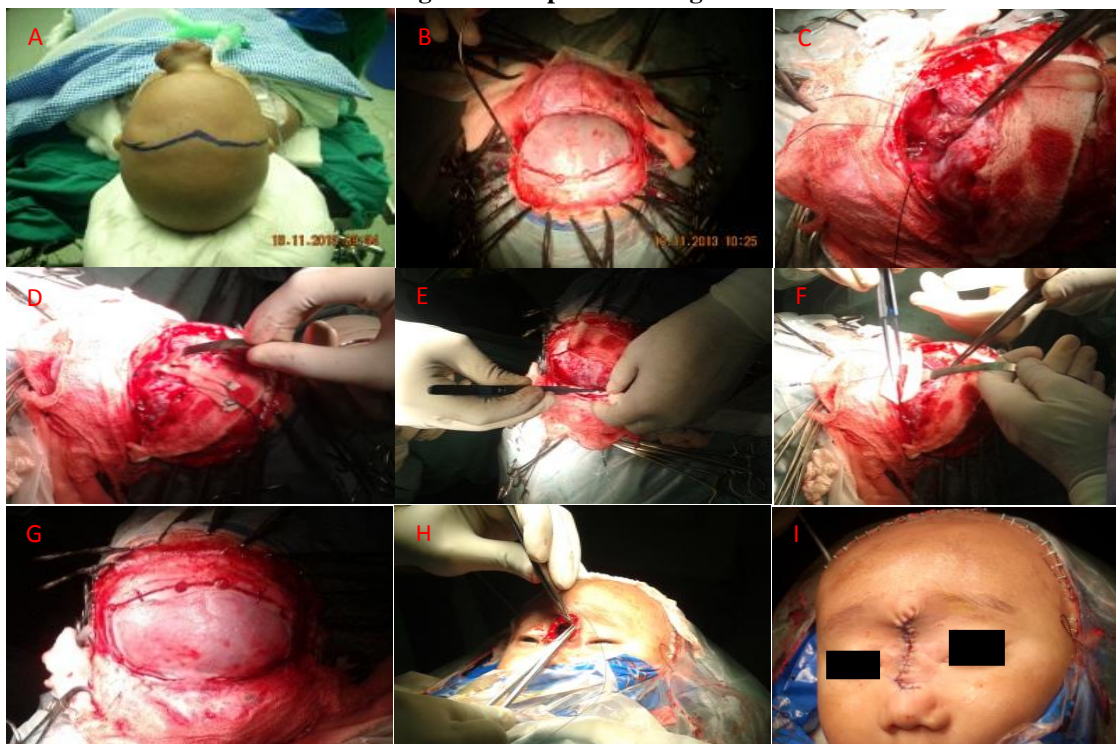
Fig 2.2: T2 weighted MRI scan-axial view showing the frontonasal defect



Intraoperatively, herniated frontal lobe covered by thinned out duramater and CSF around was noted through a 5×3 cm bony defect in the anterior half of the cribriform plate extending up to the frontonasal junction. Gliotic changes were seen at the tip of herniated brain matter.

Bifrontal craniotomy was done with a bicoronal flap. Circumferential dissection of the herniated frontal lobe was done releasing adhesions from the overlying dura. Gliotic brain tissue was excised and sent for HPE. The dissected frontal lobe was repositioned back into the anterior cranial fossa and watertight primary dural closure was done. Redundant skin over the sac was excised. Cosmetic skin closure was done by plastic surgeon. Post-operative period was uneventful. In view of small size of the bony defect and very young age of the patient bony defect was not repaired.

Fig 3: Intra operative images



Steps of Surgery (Fig 3)

- A) Skin incision marked
- B) Bicoronal flap elevated
- C) Demonstration of herniated lobe
- D) Dissection of adhesions
- E) Excision of gliotic tissue
- F) Primary dural repair
- G) Bone flap placed back
- H) Cosmetic repair by plastic surgeon
- I) Sutured wound

3. Discussion

Encephalocele does not appear to occur through defective neurulation, but due to a neuroschisis (fissure) in the neural tube after primary neurulation. This leads to scarring and subsequent adhesion between the skin and the neuroectoderm preventing interposition of the mesoderm causing bony defect.

Prenatal detection has an impact on the prevalence of encephalocele among live births which is done by antenatal ultrasound. The risk of mortality for infants born with encephalocele is highest during the first day of life and is influenced by several clinical and demographic characteristic including the site of defect, contents of sac and low birth weight. The same factors determine future physical and intellectual disability. The absence of viable brain tissue within the sac is the single most favourable prognostic factor for survival.[4] CT Scan is preferred for visualization of internal and external bony defects. MRI can visualize the herniated contents within the sac and help in determining associated anomalies. Nasofrontal encephaloceles appear at the root of the nose above the level of nasal

bone. Children with anterior encephaloceles have a better prognosis. Treatment aim is the closure of the defect. Management of these patients should involve multidisciplinary team approach with involvement of neurosurgeons, plastic surgeons, facial surgeons and paediatricians.

4. Conclusion

Developmental anomaly like sincipitalencephalocele is rare and needs early intervention to prevent complications and future physical and intellectual disability. CT scan or MRI of the brain must be done for proper evaluation of encephalocele before an invasive procedure to avoid complications in the course of management. Early surgical excision and a cosmetic repair is the treatment of choice. Prenatal counselling and screening with perinatal monitoring play an important role in holistic management of encephalocele. Multidisciplinary team approach is ideal in management of such cases.

References

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