A Large Sincipital Frontonasal Meningo-Encephalocele: A Case Report

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ABSTRACT

Encephaloceles are herniations of intracranial structures through a defect in the skull. Meningoencephaloceles involve herniation of the meninges too. This condition is most commonly diagnosed antenatally. Postnatally, infants may present with CSF rhinorrhea and recurrent meningitis. The prognosis and treatment depend on the site, size and the contents of the encephalocele.

We present a case of a large frontonasal meningo-encephalocele.

Key words: fronto nasal, sincipital, encephalocele, inter-frontal.

INTRODUCTION

Encephaloceles are extracranial herniations of intracranial structures through defects in the skull and dura. Meningoencephaloceles are herniations of meninges alone and meningoencephaloceles are herniations of brain tissue and meninges. If part of a ventricle is also included, it is called hydroencephalomeningocele. In most cases, these are detected prenatally by obstetric ultrasound. [¹] Incidence ranges from 1-3 times in 10000 live births with female preponderance with genetic and geographical variation. [¹]

CASE REPORT

An 8 month old male child brought to the Radiology department with fronto-nasal cystic swelling. The swelling measured 9.1 cm × 8.5 cm × 8 cm overlapping both eyes and forehead. The swelling was present since birth and was increasing in size. There was no associated neurological deficit or any congenital anomaly.

The patient underwent an MRI BRAIN with Screening of the whole spine which revealed a large anterior meningoencephalocele measuring 6.8 x 4.0 x 6.5 cm through the defect in basi-frontal / nasal bridge region in mid line (inter-orbital region). Meningo-encephalocele is showing parts of bilateral frontal lobes, meninges and CSF within. No hydrocephalus. Visualized brainstem and cerebellum appear normal. No e/o of any intracranial SOL .No e/o corpus callosal agenesis. Visualized spine was within normal limits.

Hence it was labeled to be a sincipital meningoencephalocele which is a rare presentation seen more commonly in South East Asia than in India.
Figure 1 (A&B): T1 weighted sagittal images show defect in basi-frontal/nasal bridge region in mid line with herniation of brain parenchyma and meninges through it.

Figure 2 (A&B): T2 weighted axial images show defect in basi-frontal/nasal bridge region in mid line with herniation of brain parenchyma and meninges through it.

Figure 3 (A&B): T1 (A) and T2 (B) weighted sagittal images show no significant abnormality or congenital deformity.

The surgical planning was done by a team of neurosurgeons and plastic surgeons. CSF WAS drained and excess meninges trimmed, herniated bifrontal lobes removed with maximum preservation of parenchyma.
Z-plasty done on the nasal ridge for correcting skin defect without tension to avoid long nose deformity. Post operative period was uneventful. Patient shows normal sensory and motor functions.

Figure 4 (A&B): Preoperative (A) and postoperative (B).

**DISCUSSION**

Encephaloceles are seen due to failure of surface ectoderm to separate from the neuroectoderm. This leads to a bony defect in the skull table, which allows herniation of meninges or brain tissue.\(^2\)

Ingraham and Matson divided encephaloceles into three categories: \(^3\)

1. occipital
2. sincipital
3. Basal

Suwanwela subdivided the sincipital group further into.\(^4\)

1. Frontoethmoidal
2. Inter-frontal
3. Encephaloceles with craniofacial clefts

The frontoethmoidal group is subdivided further into

1. Nasofrontal
2. nasoethmoidal

Depending on the content of the sac herniation of cranial content can be meninges (meningocoele), with neural element (encephalocele), with ventricles (encephalocystocele).

Anterior encephalocele is common in South East Asia and Africa while posterior defects common in the western hemisphere which includes occipito-spinal defects.\(^1\)

Mostly the encephaloceles are diagnosed on antenatal ultrasound. Postnatally, the role of the radiologist is to delineate precisely the size of the encephalocele, mention the contents of the encephalocele and identify associated anomalies.

**MRI is the modality of choice**

Brain parenchyma is directly visualized. The relation of the encephalocele and its contents to the extra cranial soft tissues are clearly shown. Large arteries and veins are also well visualized.\(^5\) Any associated findings like corpus callosum agenesis are also visualized on MRI.

**CT can provide excellent extent of the bony defect.**

Survival rate is better in anterior defects than the posterior defects. Certain side effects like ocular problems and mental retardation are sometimes noted.
CONCLUSION
Encephaloceles are of 2 types: anterior and posterior. Anterior encephaloceles have a better prognosis than posterior encephaloceles. Encephaloceles are easily identified on antenatal ultrasonography. Early identification of the associated anomalies with MRI and its associated comorbidities and surgical intervention is very important for preventing complications which may lead to neurological deficits in the future.

REFERENCES