Doi: 10.32677/IJCR.2015.v01.i02.004 **Case Report**

Giant Fronto-parietal Encephalocele: A rare case report

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Received: 15 February 2015 *Initial Review: 16 April 2015* *Accepted: 14 July 2015* *Published Online: 25 July 2015*

ABSTRACT

Although encephaloceles are common, but giant encephaloceles are rare with only few published short series and are mostly described in occipital location. Giant frontal encephaloceles are very rarely reported with only four published case reports in the world literature and giant encephalocele in fronto-parietal region has not been reported till date. Herein, we report a rare case of giant frontoparietal encephalocele in a six month old girl.

Keywords: *Encephalocele, Fronto-parietal, Giant, Neural Tube Defect*

Encephalocele is defined as protrusion of neural element through a defect in the skull or dura. Encephaloceles are common with a reported frequency of 1 per 5000 live births [1]. Giant encephalocele is one of the rarely reported central nervous system malformations. They present early in the life because of their huge size and are mostly described in the occipital region. Giant encephalocele in frontal location is quiet rare with only a few isolated case reports in the world literature [2-6]. Recently, we came across a case of giant fronto-parietal encephalocele in a six months old girl.

CASE REPORT

A 6 month female baby with huge fronto-parietal swelling presented to our tertiary hospital. This full term, 3.1 kg baby was delivered vaginally to a 28 year old third gravida mother at primary health centre. There was no history of radiation exposure or any drug intake including folic acid during antenatal period. The newborn was referred for further investigation and treatment to higher centre but the parents took no medical or surgical consultation for the baby during the initial six months. The parents reported that the swelling was present at the time of birth and enlarged progressively. There was history of poor weight gain, irritability and delayed developmental milestones. However, no seizure or cerebrospinal fluid (CSF) leaks were reported.

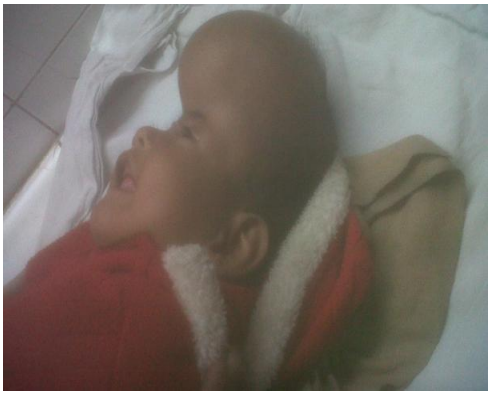

Figure 1: Giant encephalocele in fronto-parietal location

On local examination, huge swelling of size 15 cm × 12 cm was present in the left fronto-parietal region (Figure 1). Skull radiography revealed defect in the fronto-parietal region. On cranial ultrasonography, cystic mass was evident. Computed tomography showed large cystic mass communicating with lateral ventricle through a large skull

defect in the fronto-parietal region (Figure 2a, 2b). All the routine investigations were done and baby was taken for surgery in a staged manner.

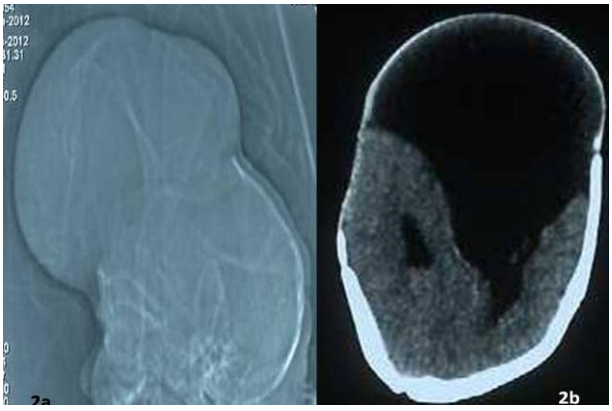

Figure 2: 2a - Computed tomography showing defect in bony calvaria in sagittal section. 2b - Encephalocele communicating with lateral ventricle

Ventriculo-peritoneal shunt surgery was done on left side as encephalocoele was communicating with the left lateral ventricle. The size of encephalocele decreased to 8 cm × 8 cm after 2 weeks, then excision of the encephalocele was carried out. The postoperative period was uneventful and baby was discharged after 15 days. There was no neurological deficit or CSF leak at the time of discharge. Baby did well in follow up period till six months after that she lost to follow up.

DISCUSSION

Encephalocele is a type of neural tube defect that occurs early in the fetal life resulting in neural tissue protruding through the defect in skull and/or dura. In developing countries, encephaloceles are not very uncommon. The reported incidence is one per 5000 live births in south-east Asian countries including India as compared to a reported incidence of 1:35000 to 1:40000 live birth in Western countries [1,2].

The most common site of encephalocele is occipital (75%) followed by fronto-ethmoidal (13-15%), parietal (10-12%) or sphenoidal [7]. Rarely, a combination of frontal and occipital locations or inter-parietal location have been reported [3,4]. The encephalocele of huge volume, the giant encephalocele is a rare entity with few published reports. The only series of giant encephalocele reported in the world literature describes 14 patients with giant encephaloceles over a period of 9 years from a referral institute of India [5]. The location of encephalocele was occipital in 93% of the cases with only one patient presenting with anterior encephalocele that too over anterior fontanelle. The index case presented with a rare giant encephalocele in fronto-parietal location.

Most of the patients typically present in early months of life as the huge swelling produces nursing and feeding problems. Eleven out of fourteen patients in the series described were under 6 months and only 2 patients were over 1 year of age [5]. The index patient presented to us for the first time at an age of 6 months due to lack of awareness and probably slowly increasing size of swelling over the time.

There are few more isolated case reports suggesting rare giant anterior encephaloceles. In all of them, patients presented either in the newborn period or during early infancy [2-6]. Neuroimaging including computed tomography (CT) and magnetic resonance imaging (MRI) is the modality of choice for diagnosis. CT has the distinct advantage of showing bone defects while MRI has got greater value is delineating brain tissue anomalies like microgyria, macrogyria or heterotopias [8,9]. MRI was not done in the index patient due to financial constraints.

The prognosis depends upon the amount of neural element herniating through the defect. The neural tissue is often dysplastic and gliotic and rarely functional. Other prognostic factors are microcephaly, associated brain anomalies, and CSF leak [8,9]. Hydrocephalus is rare in anterior encephaloceles with only 10-15% of the patients having hydrocephalus. But patients with giant encephaloceles have greater risk of hydrocephalus, associated brain tissue anomalies and CSF leak.

The surgery for giant encephaloceles involves great risk of hypothermia, blood loss, CSF leak, meningitis and significant mortality rate owing to the huge size of encephalocele [5]. The index patient did not have hydrocephalus or CSF leak and was operated for ventriculoperitoneal shunt and dura closure in a staged manner and sustained the surgery well.

Date et al described long term outcomes in surgically treated occipital encephalocoeles and concluded that the presence of gross brain tissue and large size of sac are unfavourable factors for long term prognosis [10].

Mahapatra et al described clinical features and outcomes of 133 patients with anterior encephalocoeles and reported that cosmetic outcome was good and over 50% had normal schooling; although, 85% patients were lost to follow up after 3 years [9]. The index patient had a satisfactory cosmetic outcome; although, she was lost to follow up rather quickly. Associated microcephaly and possible brain anomalies owing to large sac size may lead to a guarded prognosis.

CONCLUSION

Giant fronto-parietal encephalocele is a rare condition and pose therapeutic challenge. We report a rare case of giant frontoparietal encephalocele which was treated successfully and this, once again, emphasizes the importance of routine supplementation of folic acid to all reproductive age group females to prevent these dreaded complications.

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Funding: None; Conflict of Interest: None Stated

How to cite this article: Dalal P, Rattan KN, Gupta M, Rattan A. Giant Fronto-parietal Encephalocele: A rare case report. Indian J Case Reports. 2015; 1(2): 44-46