DOI: 10.21276/sjmcr.2016.4.11.5

Scholars Journal of Medical Case Reports **ISSN 2347-6559 (Online)**
Sch J Med Case Rep 2016; 4(11):822-824 **ISSN 2347-9507 (Print)**

(An International Publisher for Academic and Scientific Resources)

Frontal Encephalocele – A Rare Case Report with Review of Literature

Dinesh Kumar Barolia*, Sunil Mehra, Vinita Chaturvedi, Neeraj Tuteja, Soumyodhriti Ghosh, Dr. Ramjee Prasad

Department of Pediatric Surgery, J. K. Lon Hospital, S. M. S. Medical College, Jaipur, Rajasthan, India

***Corresponding author**
Dinesh Kumar Barolia
Email: dbaroliarnt@gmail.com

Abstract: Encephalocele is a congenital anomaly that represents the protrusion of brain component through the cranial defect. Frontal encephalocele is protrusion of brain content through defect in frontal bone. Frontal encephalocele is a rare entity. We report a case of congenital frontal encephalocele which is a rare neural tube defect.
Keywords: Encephalocele, Frontal, Frontal Encephalocele, Frontonasal Encephalocele.

INTRODUCTION

Encephalocele is a variant of neural tube defect. Incidence of encephalocele is not uncommon in developing countries. The incidence ranges from 1-3 per 1000 live births worldwide [1]. Anterior encephalocele is a rare entity among encephalocele. Incidence of anterior encephalocele in the western countries varies from 1/35,000 to 1/40,000 live births, but in Asian countries its incidence increases to 1/5000 live births [2]. Occipital region is the most common site for encephaloceles. Encephaloceles occurs commonly in the mid sagittal plane anywhere from frontonasal region to the occiput [3]. Occurrence of encephalocele in occipital region (75%), followed by frontoethmoidal (13% to 15%), parietal (10% to 12%) or sphenoidal. Occurrence of occipital encephalocele is common in western hemisphere where as anterior encephaloceles are found more often in south East Asia [4, 5].

CASE REPORT

We report a three month old male child presenting to the department of pediatric surgery in SMS Medical College with complaint of frontal swelling since birth. Swelling was progressively enlarged in size. The baby was born at full term vaginal delivery without any complications. The gestational period of the mother was uneventful except that she reports of not having taken folic acid tablets during this pregnancy. The baby also has a sibling who is asymptomatic. The swelling was soft, brilliantly transilluminant and pulsatile (figure 2). There was no neurological deficit. NCCT brain showed frontal bone defect through which meninges with CSF and part of right frontal lobe of brain protruded. MRI brain showed cranio-bifida in right frontal region with evidence of frontal encephalomeningocele with adjacent brain parenchyma showing cystic gliosis, communicating with right lateral ventricle (figure 3). Both lateral and third ventricles are dilated. He was admitted in surgical nursery at the age of two days. He was operated for insertion of ventriculo-peritoneal medium pressure shunt (VPMP) for hydrocephalous on right side. At the age of three months, his shunt became nonfunctioning due to blockade. The patient underwent a revision of shunt again. After insertion of the new shunt, the size of encephalocele was dramatically decreased (figure 1). Patient was discharged on post operative day three. The present status is uneventful.

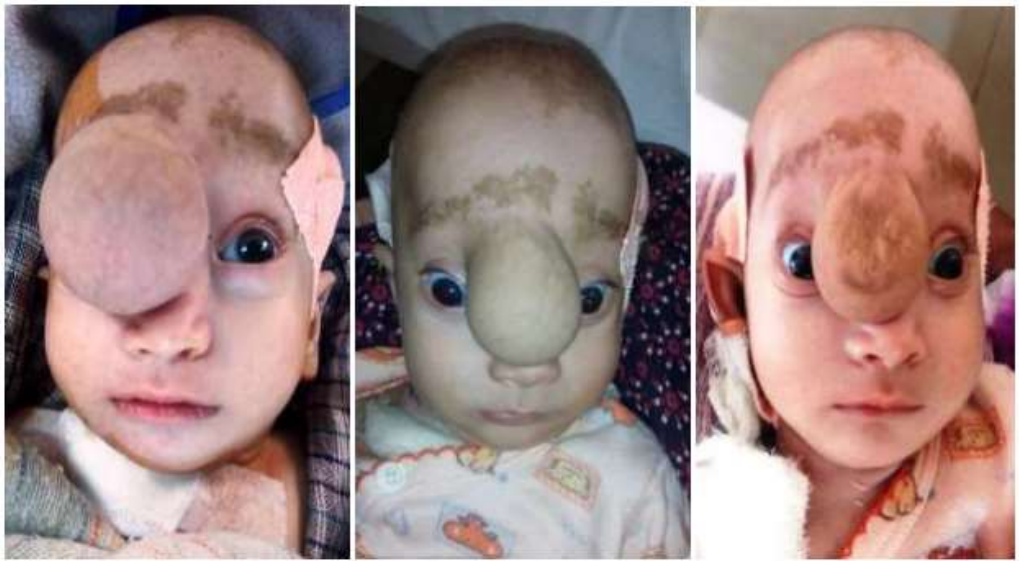

Fig:-1: Showing reduction of size of encephalocele on post operative day one, two and three

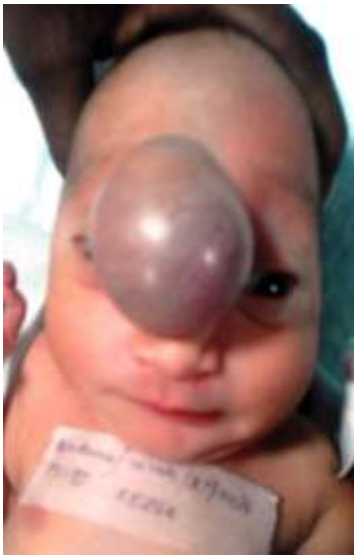

Fig-2: Showing encephalocele of same baby at the age of 2 days

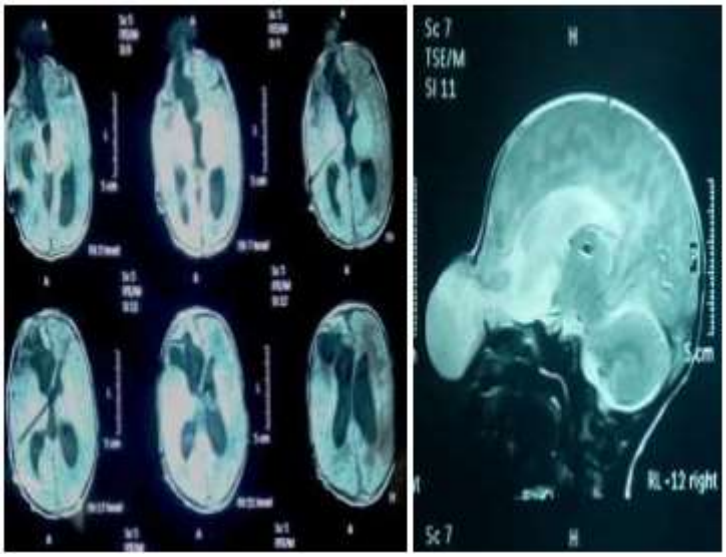

Fig-3: MRI shows communication between encephalocele and lateral ventricle

DISCUSSION

Frontal encephaloceles are rare entity. Incidence of frontal encephalocele in the western hemisphere is very less compare to Asian countries [2]. Ghosh Soumyodhriti et.al. did a retrospective study on neural tube defect. The incidence of encephalocele in this study was 14% but no mention was made of frontal defect probably because of absence of cases in his study [6]. Encephalocele is a type of neural tube defect which occurs in fetal life where the ectodermal cells that form the skull do not come together to close over the brain. This leads to protrusion of brain tissue and meninges through bony defect. Encephalocele may be present anywhere in head but most commonly located in midline occipital region. [7]. Etiology of encephalocele is multifactorial. It is documented that encephalocele is common in parents having history of neural tube defect. 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 [8]. Amarin ZO, Obeidat AZ reported that fortification of food with folic acid reduces the incidence of encephalocele [9]. Confirmative diagnosis of frontal encephalocele is made by CT head and MRI brain. CT scan helps to know about bony abnormalities in skull. Magnetic resonance imaging is better to know brain tissue details, including any intracranial connection [10]. Encephalocele is often associated with craniofacial abnormalities or other brain malformations. Symptoms may include neurologic problems, hydrocephalus, spastic quadriplegia, microcephaly, ataxia, developmental delay, vision problems, mental and growth retardation, and seizures [11]. Morina A. *et al*. found that the prevalence of encephalocele is higher among rural patients of our country with poor living conditions, malnutrition . They advised to avoid surgery in very young children until body weight is around 5-6 kilograms because of complication from blood loss and hypothermia. Hydrocephalus should be treated before corrective surgery for encephalocele, to avoid the risk of postoperative CSF leak [12].

CONCLUSION

Frontal encephalocele is a variant of neural tube defect. It is a rare entity. We recommend treating the hydrocephalus initially and observing the baby for reduction in size of the encephalocele. Definitely surgery should be deferred for an elective date.

REFERENCES

1. Rad IA, Farrokh-Islamlou HR, Khoshkalam M. Neural tube defects prevalence in a hospital-based study in Urmia. Iranian Journal of Child Neurology. 2008 Oct 18;2(3):19-23.
2. Barrow N, Simpson DA. Cranium bifidum:Investigation, prognosis and management. AustPaediat J. 1966;2:20-26.
3. Ramachandra CR, Phelps PD. Nasal encephalocoeles associated with unilateral absence of the cochlea. The Journal of Laryngology & Otology. 1977 Sep 1;91(09):813-7.
4. Mahapatra AK, Aqrawal D. Anterior Encephaloceles: a series of 103 cases over 32 years. J Clin Neurosci. 2006;13:536–9.
5. Hoving EW. Nasal Encephaloceles. Child's Nerv Syst. 2000;16:702–6.
6. Soumyodhriti G, mujaldevikram Singh S, Kumar G. Neural Tube Defects-Demography in a Tertiary Care Center. IOSR Journal of Dental and Medical Sciences (IOSR-JDMS);1(15):80-2.
7. Siffel C, Wong LY, Olney RS, Correa A. Survival of infants diagnosed with encephalocele in Atlanta, 1979–98. Paediatric and perinatal epidemiology. 2003 Jan 1;17(1):40-8.
8. MRC Vitamin Study Research Group; Prevention of neural tube defects: Results of the Medical Research Council Vitamin Study. Lancet. 1991;338(8760):131-137.
9. Amarin ZO, Obeidat AZ. Effect of folic acid fortification onthe incidence of neural tube defects. PaediatrPerinatEpidemiol. 2010;24:349-351.
10. Verney Y, Zanolla G, Teixeira R, Oliveira LC. Midline nasal mass in infancy:a nasal glioma case report. Eur J Pediatr Surg. 2001;11:324-7.
11. Encephalocele imaging (http:// emedicine. Medscape. Com/ article/ 403308- overview) at emedicine.
12. Morina A, Kelmendi F, Morina Q, Dragusha S, Ahmeti F, Morina D. Treatment of Anterior Encephaloceles Over 24 Years in Kosova . Med Arh. 2011;65(2):122-124.