

Giant high occipital encephalocele

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Abstract: Encephaloceles are rare embryological mesenchymal developmental anomalies resulting from inappropriate ossification in skull through with herniation of intracranial contents of the sac. Encephaloceles are classified based on location of the osseous defect and contents of sac. Convexity encephalocele with osseous defect in occipital bone is called occipital encephalocele. Giant occipital encephaloceles can be sometimes larger than the size of baby skull itself and they pose a great surgical challenge. Occipital encephaloceles (OE) are further classified as high OE when defect is only in occipital bone above the foramen magnum, low OE when involving occipital bone and foramen magnum and occipito-cervical when there involvement of occipital bone, foramen magnum and posterior upper neural arches. Chiari III malformation can be associated with high or low occipital encephaloceles. Pre-operatively, it is essential to know the size of the sac, contents of the sac, relation to the adjacent structures, presence or absence of venous sinuses/vascular structures and osseous defect size. Sometimes it becomes imperative to perform both CT and MRI for the necessary information. Volume rendered CT images can depict the relation of osseous defect to foramen magnum and provide information about upper neural arches which is necessary in classifying these lesions.

Key words: Giant encephalocele, high occipital encephalocele, occipital encephalocele, meningocele, cephalocele, posterior encephalocele

Introduction

Encephaloceles are rare embryological mesenchymal developmental anomalies resulting from inappropriate ossification defect in skull through with herniation of intracranial contents. (1-4) Nomenclature is mainly based on location and the contents of

the herniated sac. When meninges alone are the content they are meningoceles, when brain tissue along with meninges is the content of herniation it is called meningoencephalocele or encephalocele and occasionally they may include a ventricle then they are called as hydroencephaloceles. (3, 4) Dural sinuses as

contents of encephalocele are also seen infrequently. (3, 4) Atretic encephaloceles contain fibrous tissue within the sac and sometimes intracranial communication may not be present. (5) Encephaloceles cause severe morbidity and mortality if untreated. Occipital encephalocele (OE) is posterior encephalocele which is a result of herniation of intracranial contents through occipital bone defect with or without associated foramen magnum and upper cervical vertebral defects. (1, 2, 6-8) Giant occipital encephaloceles (GOE) because of their size they are a neurosurgical challenge. (1) Herein we report a 40 day old female child who presented to us with giant high occipital encephalocele and imaging played a decisive role in providing preoperative information.

Case report

A 40 days old female child was brought to pediatric outpatient department with complaints of swelling over back of head since birth. The mother due to her poor socio-economic status did not have regular antenatal checkups and antenatal ultrasound. Baby was delivered through caesarian section, as there was delay in head fixation after onset of labor pains. Baby cried immediately after birth. Large occipital swelling was noticed at birth and APGAR score was 10. There was no h/o similar births in family. On examination, there was a single large gray colored round shaped 8 x 8 cm swelling at occipital region of head. Skin over the swelling was normal. All the scalp veins were dilated. The swelling was compressible & baby cried on compression of the swelling. Transillumination test was positive with dark areas showing reduced

transillumination. Child did not have any other associated abnormalities. Routine investigations were normal except for persistent elevation of serum potassium levels (Serum potassium- 6.2 meq/L, Na-136 meq/L, Cl: 94 meq/L, Urea 14.8 mg/dl and creatinine: 0.56 mg/dl.). CT scan was advised which showed high occipital bony defect with herniation of meninges and posterior fossa contents (Figures 1, 2). Surface rendered and volume rendered images showed detail of giant occipital encephalocele, size of osseous defect, and relation to foramen magnum. (Figure 3). MRI showed details of total herniation of cerebellum, occipital horn of lateral ventricle and part of mid brain (Figures 4, 5). There was no associated chiari malformation. Diagnosis of high occipital encephalocele was made. The child underwent complete surgical excision and repair of the meningocele and doing well at follow up.

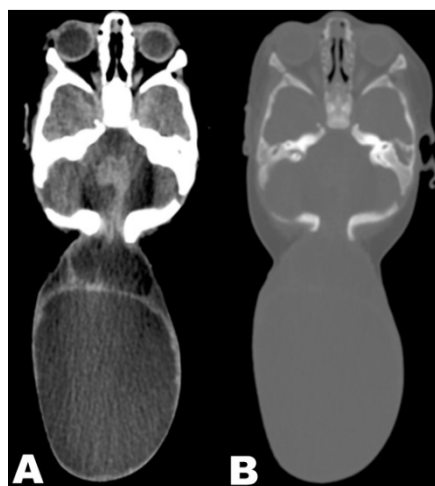


Figure 1 - Axial CT scan images brain window (A) and soft tissue windowing (B) showing larger posterior fossa defect, with herniation of brain tissue and meninges

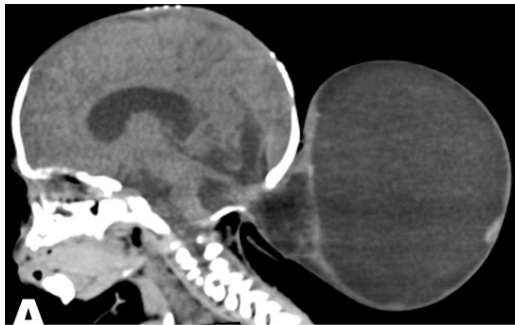


Figure 2 - Saggital reconstructed CT scan images showing herniation of posterior fossa structures through the defect

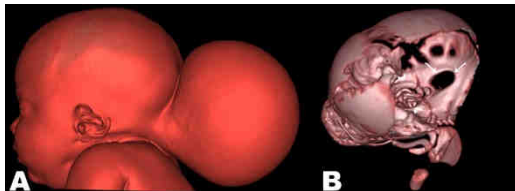


Figure 3 - Surface rendered images (A) showing the cyst size, volume rendered images of skull (B) showing the size of osseous defect (white arrows) in relation to foramen magnum (arrow heads)

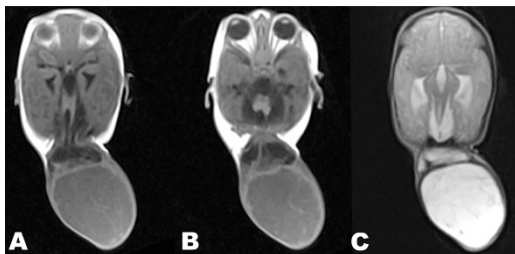


Figure 4 - T1 (A, B) and T2 weighted axial images (C) of MRI showing dysmorphic cerebellar tissue, fourth ventricular contents and meninges herniated through the defect, also note stretching of tectum



Figure 5 - T1 (A) and T2 (B, C) weighted axial images showing the herniation of posterior fossa structures and beaking of the tectum

Discussion

Encephaloceles are identified and classified based on the location of the osseous skull defect. When defect is located in frontoethmoidal, nasofrontal, nasoethmoidal, naso-orbital, interfrontal regions it is called as sincipital encephalocele. (3) Convexity encephaloceles have osseous defect located in parietal, occipital, occipital-cervical regions and finally basal encephaloceles have defect in the skull base. (3) Incidence of encephaloceles is 0.8 - 3.0 per 10,000 live births and varies based on geological location and race. (3, 6) However with improvement of antenatal detection of these lesions, incidence has decreased significantly in recent times. (4) Associated anomalies (microcephaly, chiari III malformation, craniosynostosis, and syringomelia and neural tube defects) are seen in approximately 20% of children. (3, 4) Anterior encephaloceles are more common than posterior ones in Asian continent. (3) OE are further classified as high OE when defect is only in occipital bone above the foramen magnum, low OE when involving occipital bone and foramen magnum and occipito-cervical when there involvement of occipital bone, foramen magnum and posterior upper neural arches. Chiari III malformation can be associated with high or low occipital encephaloceles. (9) The size of occipital OE may vary from small to giant masses. (1, 6) Giant encephaloceles can be some times larger than size of baby skull and because of their enormous size pose a great surgical challenge. (1) OE are normally covered by healthy skin or abnormal skin, sometimes only a thin

meningeal membrane may be covering the OE. (10) Contents within the herniated sac are variable. (1, 6) Ultrasound and doppler are cheap readily available tools for knowing contents of sac; however MRI may be required to know contents of sac in detail. CT is mainly used to assess the size of the osseous defect. (3, 4) Conservative approach for encephalocele is not advised as many complications such as infection, trauma, and hydrocephalus can worsen the clinical scenario. (3, 6, 8) Amount of viable brain tissue, size of the OE, skin covering and associated congenital anomalies should be considered in planning surgery. Repair of OE without active CSF leak or overlying skin necrosis should be an elective procedure and be done without delay. Anaesthetic support is extremely important while operating OE, endotracheal intubation is real challenge in these patients and it should be done in right or left lateral position based on individual anaesthetist expertise. (11, 12) In general dysplastic brain tissue and meninges should be excised preserving the vascular structures. Attempts should be made to preserve normal brain parenchyma and ventricles by expansion cranioplasty and ventricular reduction. Reconstructive surgeries like skin flap rotation should be performed when there is inadequate skin for primary closure. (1-3, 6) Great importance should be given for two-layered watertight closure. (6) Osteogenic properties of dura can reduce the bony defect obviating the need for cranioplasty. Titanium mesh closure of defect can also be tried. (3) Surgical outcome is generally good independent of size depending upon amount of neural tissue. Even giant OE

with less neural tissue have excellent prognosis. (1, 4, 6, 8) However in giant OE due to the removal of abundant CSF volume, electrolyte imbalances can occur which should be corrected peri-operatively. Amount of normal viable brain tissue after surgery also is important in assessing the prognosis.

Conclusion

Giant high occipital encephaloceles are rare congenital lesions. Diagnosis can be made soon after birth by clinical examination and because of their enormous size they pose a great surgical challenge. Elective surgical repair can be performed as early as possible. Pre-operatively it is essential to know the size of the sac, contents of the sac, relation to the adjacent structures, presence or absence of venous sinuses/vascular structures and osseous defect size. Sometimes it becomes imperative to perform both CT and MRI for the necessary information. Volume rendered CT images can depict relation of osseous defect to foramen magnum and information about upper neural arches which is necessary in classifying these lesions.

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