Case Report

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Surgical Repair of a Giant Encephalocele with Chiari Malformation Type III- A Therapeutic Challenge

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Abstract

An encephalocele is a congenital neural tube defect characterized by herniation of the intracranial contents through a defect in the cranium. It is caused by failure of closure of the cranial part of the developing neural tube during the first few weeks of foetal life. An encephalocele is called a giant encephalocele when its size is larger than the size of the head. Surgical procedures for giant occipital encephalocele are challenging for both the neurosurgeons and anaesthesiologists due to the large size, associated intracranial anomalies, intraoperative blood loss, intraoperative hypothermia, and prolonged anaesthesia. The index case was antenatally diagnosed to have an encephalocele. Postnatal examination revealed a large meningoencephalocele – 10* 10 cm size in the occipital region. MRI brain was suggestive of occipital meningoencephalocele with syrinx involving cervical spinal cord. Baby was operated on day twenty of life. Post operatively, wound site healed with primary intention and was discharged after ten days and child was neurologically normal at the time of discharge.

Keywords: Giant Encephalocele, Neural Tube Defect, Chiari Malformation Type III, Syringomyelia, Hydrocephalus

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Introduction

An encephalocele is a congenital neural tube defect (NTD) characterized by herniation of the intracranial contents through a defect in the cranium.[1] Encephalocele is caused by failure of closure of the cranial part of the developing neural tube during the first few weeks of foetal life. An encephalocele is called a giant encephalocele when size of the encephalocele is larger than the size of the head.[2] Giant encephaloceles are also known as massive encephaloceles and large encephaloceles.[3] Giant encephaloceles are most commonly observed in the occipital region.

Surgical procedures for giant occipital encephalocele are challenging for both the neurosurgeons and anaesthesiologists due to the large size, associated intracranial anomalies, intraoperative blood loss, intraoperative hypothermia, and prolonged anaesthesia.

Occipital encephaloceles are most commonly operated in the prone position. Partial aspiration of cerebrospinal fluid (CSF) is an option to reduce the volume of the giant occipital encephalocele to facilitate the process of endotracheal intubation.

Giant occipital encephalocele associated with Chiari malformation Type III is an extremely rare occurrence.[4,5] Chiari malformation Type III is characterized by the herniation of posterior fossa contents, including cerebellum, brain stem, and fourth ventricle, and sometimes, the upper cervical spinal cord through a low occipital or an upper cervical osseous defect. MRI brain is the preferred investigation.

Surgical procedures for giant occipital encephalocele are a challenging task, not only for neurosurgeons, but also for the anesthesiologists. These challenges are due to its site, large size, contents, associated intracranial anomalies, intraoperative blood loss, intraoperative hypothermia, and prolonged anesthesia.[3–5] The problems faced by anesthesiologists during anesthesia management of patients with giant occipital encephaloceles are difficult tracheal intubation, instability in hemodynamic, excess blood loss, intraoperative hypothermia, and cardiorespiratory complications. Occipital encephaloceles are best operated in the prone position. Partial aspiration of CSF is also a better option to reduce volume of giant occipital encephalocele to facilitate the endotracheal intubation.

Operative repair for giant occipital encephaloceles consists of excision of the excess meninges, partial or complete excision

of the protruded brain tissue, watertight closure of the dural defect and approximation of the overlying skin.[6]

Associated hydrocephalus is quite frequently present with giant occipital encephaloceles or develops later following surgical repair, and CSF diversion in the form of ventriculoperitoneal shunt is also an additional surgical procedure required.[3,5] Complications observed after repair of giant occipital encephaloceles are CSF leak, postoperative meningitis, wound infection, wound dehiscence, and hydrocephalus.

The prognosis of children treated for giant occipital encephaloceles depends on many factors such as the size, amount of brain tissue involved in the encephalocele, associated intracranial (microcephaly and hydrocephalus), extracranial anomalies, and the treatment offered for correction of the disease.[3] Associated hydrocephalus and microcephaly are associated with a significant morbidity in these children.[6]

Case History

Baby was born to a 30 year old G2P1 mother at 40 weeks gestation by LSCS i/v/o fetal distress. Baby cried immediately after birth. There was no history of any medical or surgical illnesses in the mother. Antenatal sonography was suggestive of encephalocele in occipital region with herniation of cerebellum. Postnatal examination revealed large meningoencephalocele of size ten cm by ten cm size in the occipital region. [Figure 1] Occipital defect of one cm by two cm was felt. The swelling was small at the time of birth, but gradually increased in size. The overlying skin was tense and without any CSF leak. Swelling was cystic and its size increased on crying. Fluctuation could be elicited and transillumination test was also positive. [Figure 2] There was no bruit or murmur over the swelling. Anterior fontanelle and posterior fontanelle were both open. Baby was active and had no focal neurological deficit. Systemic examination was unremarkable. Activity and tone in all four limbs was good. There was no abnormality on physical examination except for a large cystic mass in the occipital region. It was larger than the size of the head. Baby was able to track objects and light and pupils were reactive. Routine hematological and biochemical investigations were reported as normal. MRI Brain was suggestive of a well-defined T2 hyperintense and T1 hypointense outpouching with septations measuring 9.6 * 8.5 * 11 cm containing herniated bilateral occipital lobe, superior cerebellum and areas of hemosiderin staining were seen arising out of the defect of size 0.9 * 1.8 cm in occipital bone. Flow void of posterior superior sagittal sinus and confluence was seen within the outpouching. Stretching and flattening of corpus callosum and effacement of fourth ventricle was seen. Syrinx was present in cervical spinal cord. Multiple T2 hypo intense sub ependymal nodules were noted along both lateral ventricles. MRI brain was suggestive of occipital meningoencephalocele with sub ependymal nodular heterotopia with syrinx involving cervical spinal cord. [Figure 3] Baby was operated on day twenty of life. Occipital encephalocele repair was done under general anaesthesia in prone position. Gliotic brain tissue and multilobulated fluid cyst was found on examination. Post operatively, wound site healed with primary intention and baby was discharged after ten days. [Figure 4] Sutures were removed after fifteen days post operatively. Baby was neurologically normal at the time of discharge.

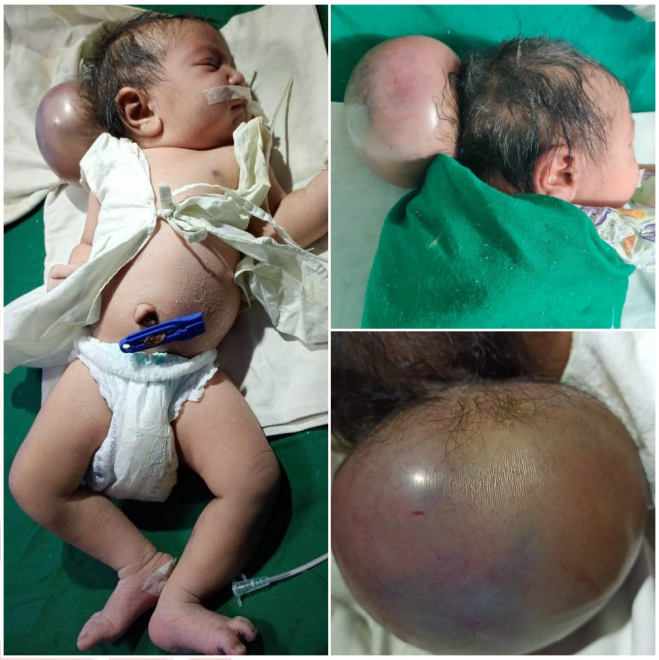

Figure 1: A giant encephalocele sac in the occipital region with intact overlying skin.

Discussion

Repair of a giant encephalocele is a therapeutic challenge. Agarwal and colleagues described a case of a four month old female baby who presented with progressively increasing swelling over the occipital region. This swelling was present since birth. During surgery, the redundant sac was excised and reduced to accommodate the healthy looking brain tissue. The infant did well postoperatively. Patient was positioned in lateral position. In our study, patient was positioned intraoperatively in the supine position. Baby was operated on day 20 of life.[7]

Bulut et al described a giant cystic lesion in the cervical-occipital and upper thoracic regions of the fetus. Brain MRI showed a defect at the lower occipital/upper cervical region and a large encephalocele sac herniated from this

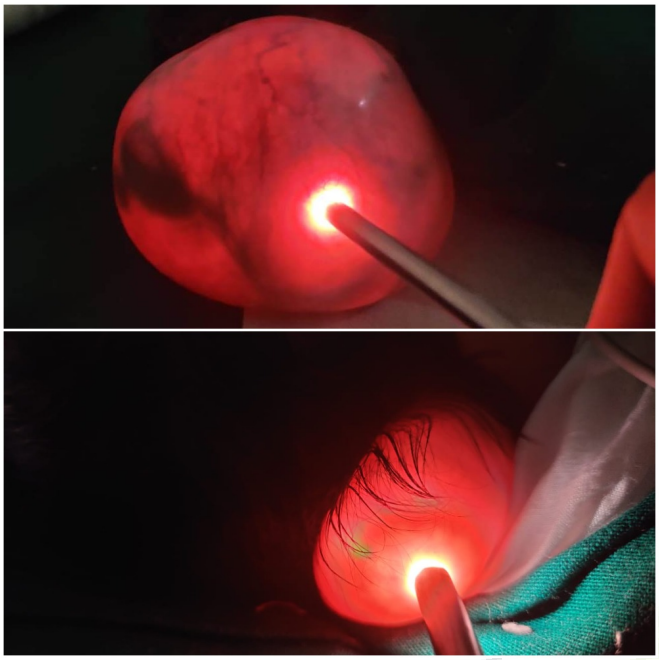

Figure 2: Transillumination test on the swelling was positive.

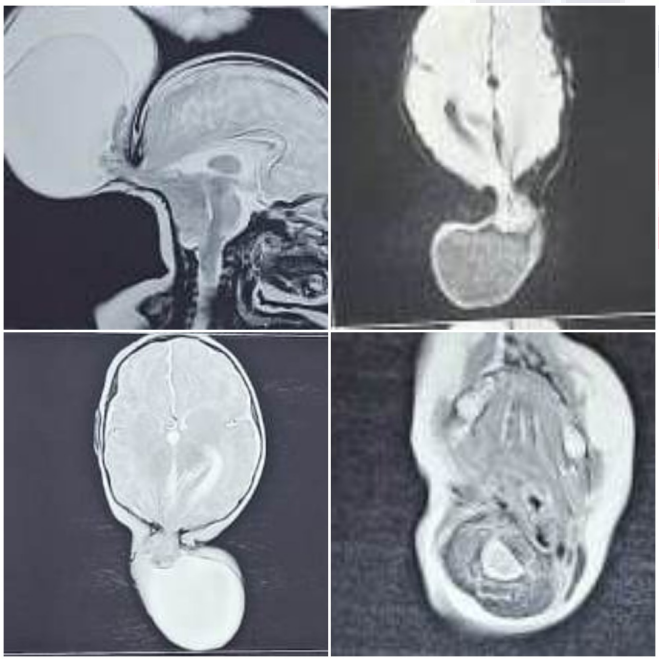

Figure 3: MRI Brain was suggestive of a well-defined T2 hyper intense T1 hypo intense outpouching with septations containing herniated bilateral occipital lobe, superior cerebellum.

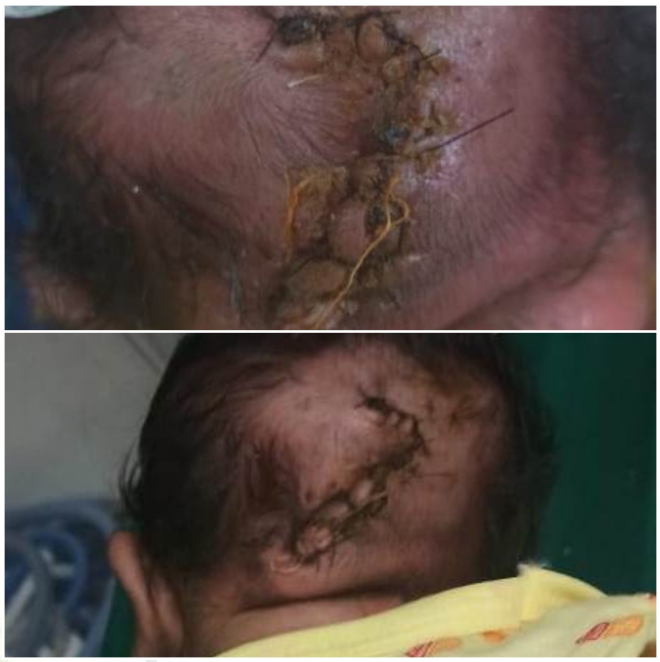

Figure 4: Post operatively, the wound site healed with primary intention.

defect, which contained the right cerebral occipital lobe, the bilateral cerebellar hemispheres and the fourth ventricle. A small posterior fossa gaping foramen magnum with downward herniation of the cerebellar vermis and brain stem into the foramen magnum, hypoplastic tentorium, dysgenesis of the corpus callosum and hydrosyringomyelia was seen. Six days after birth, the infant underwent surgery wherein dysplastic cerebellar tissue with a herniated sac was excised, and the primary closure of the defect was performed. But infant died one day after the surgery. This is similar to our case where there was herniation of cerebellum and occipital lobes and syringomyelia. But in our case, the patient survived after closure of the defect.[4]

Rehman and colleagues described fifty patients of occipital encephalocele of which 17 were males and 33 were females. The average age at presentation was 2.4 months. 16 (32%) patients had hydrocephalus, two (4%) had Dandy–Walker cyst, three (6%) developed developmental delay, and eight (15%) had a seizure disorder. One patient (2%) had a cerebrospinal fluid leak postoperatively while four (8%) developed hydrocephalus after repair of the sac and hydrocephalus was subsequently treated with placement of a ventriculoperitoneal shunt. One (2%) patient could not recover from anesthesia and expired subsequently. In our case, the baby was diagnosed antenatally and did not have any post-operative complications. Baby did not develop seizures or hydrocephalus.[8]

Mardzuki and colleagues reported three patients who underwent a two-stage management of mega (sac larger than 20cm) occipital encephalocele where fluid was drained via an external ventricular catheter over a period of two weeks prior to definitive surgery. Serum electrolytes were monitored and maintained in the normal range. The operation for closure of the occipital encephalocele was done according to the modified technique of Gallo in the prone position. Efficient CSF management will prevent the complications of sudden electrolyte loss and displaced brain structures during surgery.[9]

Mahapatra reported 14 patients with giant encephaloceles who underwent repair. Four patients had suturectomy of coronal suture in view of secondary craniostenosis. There were two postoperative deaths due to hypothermia. Among the 12 patients who survived, nine had a good outcome and three had poor mental development. There was overall good outcome in nine out of 14 (66%) patients.[10]

Kumar and colleagues described a two month old female with a giant encephalocele. The circumference of the occipital swelling was 63 cm. This patient had a good post-operative outcome.[11]

The management of occipital encephaloceles requires an individualized approach. In a giant occipital meningoencephalocele problems encountered are the large size, positioning in operation theater, intubation, and blood loss during resection of the large amount of redundant skin. The presence of gross brain tissue in sac, associated hydrocephalus, or congenital anomalies are unfavorable prognostic factors. It is observed that microcephalic neonates with sac containing cerebrum, cerebellum, and brain stem structures have poor prognosis.

References

1. Silva SLD, Jeelani Y, Dang H, Krieger MD, McComb JG. Risk factors for hydrocephalus and neurological deficit in children born with an encephalocele. J Neurosurg. 2015;15(4):392–398. Available from: https://dx.doi.org/10.3171/2014.10.peds14192.
2. Borkar S, Mahapatra A. Encephaloceles: A Study of 110 Patients Operated on at AIIMS. J Neurosurg . 2012;73(S 02):520. Available from: https://dx.doi.org/10.1055/s-0032-1314418.
3. Mahapatra AK. Giant encephalocele: A study of 14 patients. Pediatr Neurosurg. 2011;47(6):406–411. Available from: https://doi.org/10.1159/000338895.
4. Bulut MD, Yavuz A, Bora A, Gülşen İ, Özkaçmaz S, Sösüncü E. Chiari III Malformation with a Giant Encephalocele Sac: Case Report and a Review of the Literature. Pediatr Neurosurg. 2013;49(5):316–319. Available from: https://dx.doi.org/10.1159/000365661.
5. Kanesen D, Rosman AK, Kandasamy R. Giant Occipital Encephalocele with Chiari Malformation Type 3. J Neurosci Rural Pract. 2018;09(04):619–621. Available from: https://dx.doi.org/10.4103/jnrp.jnrp_103_18.
6. Ghritlaharey RK. A Brief Review of Giant Occipital Encephalocele. J Neurosci Rural Pract. 2018;09(04):455–456. Available from: https://dx.doi.org/10.4103/jnrp.jnrp_189_18.
7. Agarwal A, Chandak AV, Kakani A, Reddy S. A giant occipital encephalocele. APSP J Case Rep. 2010;1(2):16.
8. Farooq G, Rehman L, Bukhari I. Neurosurgical interventions for occipital encephalocele. Asian J Neurosurg. 2018;13(2):233. Available from: https://dx.doi.org/10.4103/1793-5482.228549.
9. Mardzuki AI, Abdullah J, Ghazaime G, Ariff AR, Ghazali M. Two-stage management of mega occipito-encephalocele. Med J Malaysia. 2003;58(1):115–124.
10. Mahapatra AK. Giant encephalocele: a study of 14 patients. Pediatr Neurosurg. 2011;47(6):406–417. Available from: https://doi.org/10.1159/000338895.
11. Kumar V, Kulwant S, Saurabh S, Richa S. Giant occipital meningoencephalocele in a neonate: A therapeutic challenge. J Pediatr Neurosci. 2017;12(1):46. Available from: https://dx.doi.org/10.4103/1817-1745.205655.

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