Case Report

Giant encephalocele – A rare case report with review of literature

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Background: A congenital neural tube defect known as an encephalocele is characterised by the herniation of cranial contents via a defect in the skull and is brought on by a failure of the cranial part of the developing neural tube to close. When an encephalocele is larger than the head, it is referred to as a "giant encephalocele". For a positive neurological outcome, they consider the size of the sac, the percentage of neural tissue inside, hydrocephalus, infection, and other related disorders.
Case Presentation: We describe a case of a four-month-old child who had a massive occipital encephalocele measuring 23× 19X 8cms in size. This case presented us with both surgical and anaesthetic challenges. Intubation was accomplished while lying on the side. The sac contained some of the parenchyma of the occipital and cerebellum. The surgical excision and repair we did had a positive overall result.
Conclusion: A large occipital encephalocele poses a difficulty for anesthesiologists and neurosurgeons alike in terms of perioperative treatment. Such a situation necessitates the search for further congenital anomalies, airway management competence, and appropriate perioperative care. For a satisfactory outcome, careful preparation and perioperative management are crucial.

KEY WORDS: Encephalocele, Occipital encephalocele, Giant encephalocele, Neural pore defect

BACKGROUND

Encephalocele is an uncommon neural tube condition that normally affects one in every 5,000 infants worldwide, with 70% of cases being occipital.[1] Multiple cranial contents herniating during the first few weeks of fetal development due to a defect in the cranium brought on by improper closure of the growing cranial section of the neural tube.[2] The term "giant encephalocele" is used when an encephalocele is larger than the size of the head and can range in size from a few centimeters to a huge swelling.[2]

Size of the sac, hydrocephalus, potential infections, other associated illnesses, the neurological condition of the patients before surgery, and the herniation of the cerebral contents into the sac continue to play a significant role in determining the long-term prognosis. Due to blood loss, the need for postoperative care, and apparent difficulty in successfully opening the airway when prone, the anesthetic management of occipital meningoencephalocele provides a challenge.[3]

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CASE PRESENTATION

A 3 month/F presented to OPD with the chief complaint of huge progressive occipital swelling present since birth. On examination, the child was active, midline pedunculated swelling of approximately 23 × 19 × 8 cms at the craniocervical junction, soft, transilluminant, and non-pulsatile in nature [Figures 1 and 2]. The overlying skin was intact, though few areas of skin discoloration were present. No apparent traumatic injury was observed. Anterior fontanelle was lax and open, measuring about 3.5 × 2.5 cm. The child was moving all four limbs and power and tone were normal.

There was no history of antenatal checkups and the baby was delivered through cesarean section at term in a local hospital.

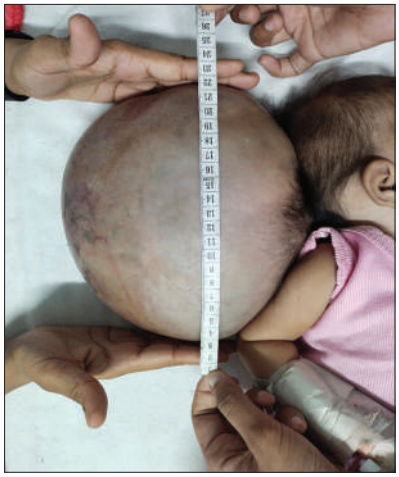

Figure 1: Vertical axis of swelling

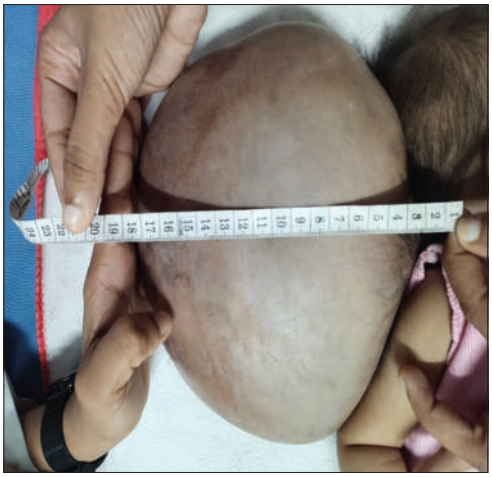

Figure 2: Horizontal axis of swelling

Routine investigations were normal.

MRI brain was suggestive of occipital encephalocele [Figure 3].

Parents were counseled in detail about the diagnosis and after obtaining informed written consent, surgical excision and repair were planned.

Other associated congenital anomalies were ruled out by pediatrics, cardiology, and anesthesia team. After optimization of all the parameters, the patient was taken up for surgery.

It was a significant challenge for the anesthetist too to intubate this child. Intubation was achieved in the lateral position. The patient was placed in the prone position while holding the swelling above the head throughout the surgery [Figure 4].

After painting and draping, incision of approximately 3 cms was given followed by blunt and sharp dissections in a circumferential manner. Collected CSF was aspirated, and sac and gliosed brain tissue were excised [Figure 5] and dural layer was precisely identified, and brain tissue was reduced [Figure 6]. Meninges were closed in a watertight overlapping manner using vicryl 4-0 suture [Figure 7]. Subcutaneous layer was closed with vicryl 3–0 suture.

Skin closure was done with prolene 3–0 suture [Figure 8].

The patient was shifted to NICU and kept on ventilator support. On POD0, the patient developed a single episode of seizure for which dual antiepileptics were started. On POD1, the patient had anterior fontanelle bulge, and NCCT head was done which was suggestive of hydrocephalus. Anterior fontanelle tap was done. On POD3, the patient got extubated and feeds were started through OG tube initially and later EBM was given. The patient was discharged in good condition.

DISCUSSION

A normal, dysplastic, or thin membrane is used to cover the enormous encephalocele's meningeal membrane. The distinctive swelling of gigantic occipital encephaloceles makes their clinical appearance clear.[2] Meninges and occipital lobes make up the majority of encephalocele in the occipital region. The brain stem, cerebellum, and ventricles may also be included. The location, size, and degree of brain herniation into the sac are the main parameters influencing the prognosis of individuals with occipital encephalocele. The result of the case is also influenced by the presence of the brainstem or occipital lobe, with or without the dural sinuses in the sac.[4]

The huge swellings might theoretically have microcephaly, a stunning brain herniation, or any of these conditions. The clinical examination includes a look at its size, extent, prominence, and position in addition to the size of the bone defect.[2] For the clinical suspicion of microcephaly, hydrocephalus, and extracranial abnormalities, the size of the head is significant.[2] MRI brain and three-dimensional computed tomography remain the standard investigations of choice and aid in assessing the abnormality further.[5]

Giant encephaloceles are uncommon, and removing them surgically is difficult for both anesthesiologists and neurosurgeons. The difficulties are mostly brought on by the location's complexity, size, and concomitant bulging contents, which might cause intracranial abnormalities, intraoperative blood loss, and prolonged anesthesia.[6] The anesthesiologist's primary goal is to stop the encephalocele from prematurely rupturing during surgery. The occipital placement of the encephalocele makes intubation difficult because the limited range of motion in the neck makes it difficult to open the airway. This further prevents one from being in the best position for tracheal intubation.[3]

The surgical approach also addresses the possibility of significant CSF loss, which would result in an electrolyte imbalance. Because the autonomic nervous system below the defect is dysfunctional, infants with encephalocele are susceptible to abrupt hypothermia.[3] However, issues related to hypothermia, blood loss, and these conditions must be urgently taken into account and managed. To avoid potentially fatal consequences such as infections of the central nervous system, respiratory distress, aspiration pneumonia, irreversible vagus nerve damage, and hypothermia, the surgery is advised to be performed as soon as feasible.[5]

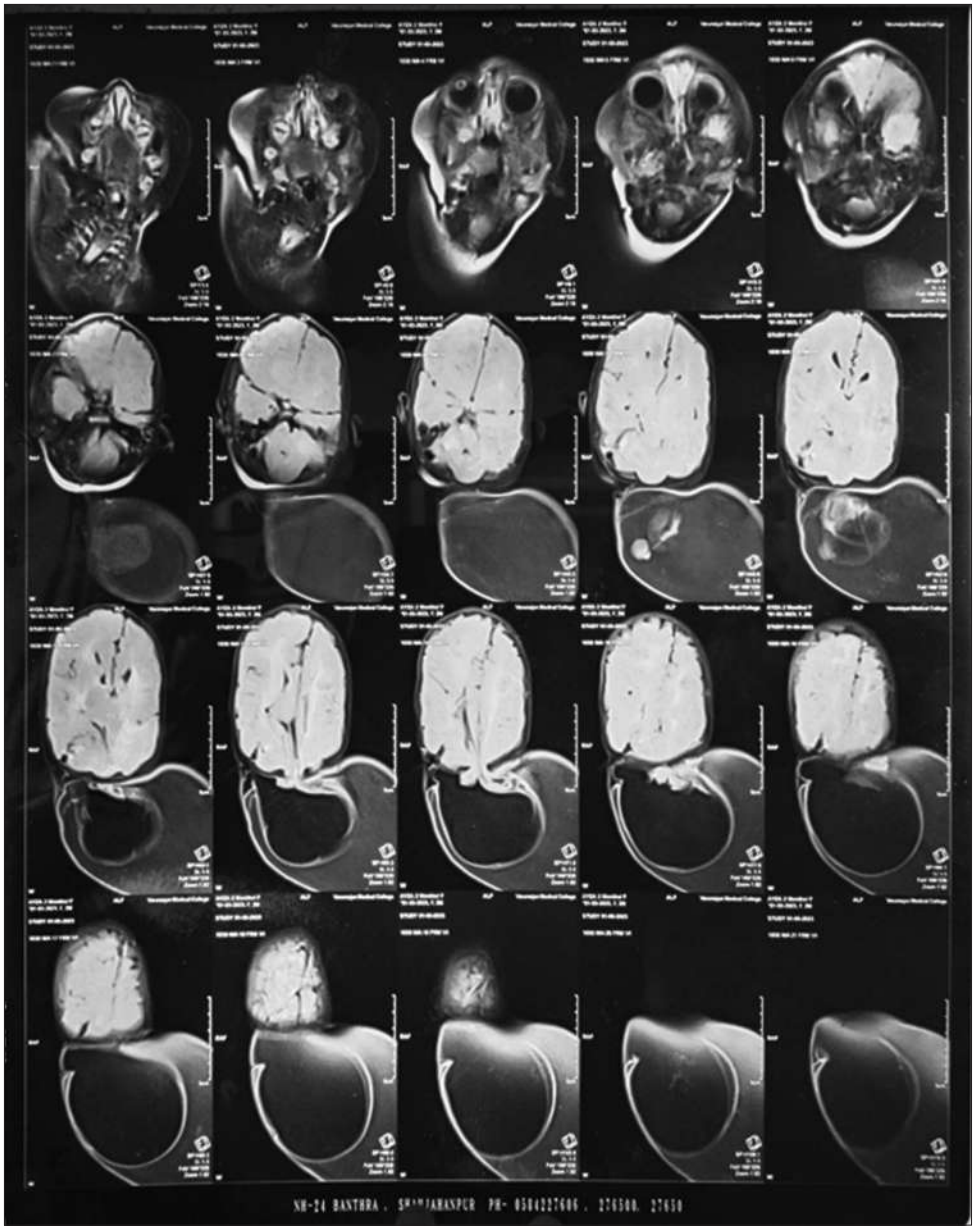

Figure 3: MRI brain suggestive of occipital encephalocele

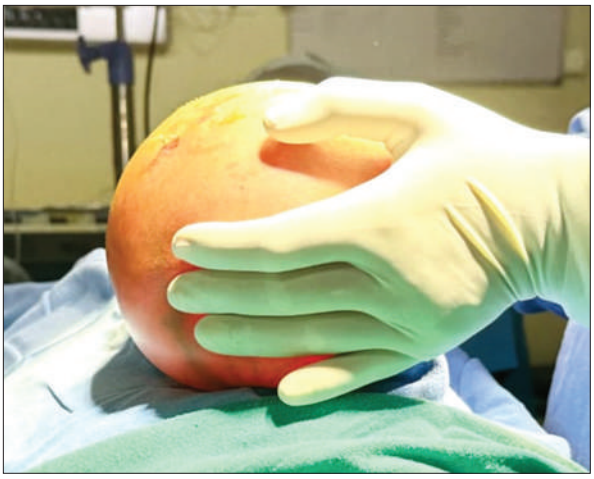

Figure 4: Patient was placed in prone position

Only a few cases of giant occipital encephalocele have been reported to date. The main goal is to illustrate the various and most often used surgical techniques for treating big occipital encephaloceles. The technique that is most commonly employed is sheer resection and dural repair. One surgical method that uses a mesh to make room for the protruding sac is expansion cranioplasty. Ventricular volume reduction is a further method that is frequently used. It is a two-step procedure that involves first raising ventricular pressure to cause hydrocephalus and then inserting a ventriculoperitoneal shunt.[7] The protruding tissue then moves back inside the skull as a result of the ventricles contracting. An incision is necessary in the brain's occipital and cerebellar parenchyma to treat the herniated tissue.

Contemporary neurosurgery methods, neuroimaging, and neonatal intensive care with neurological facilities have significantly reduced

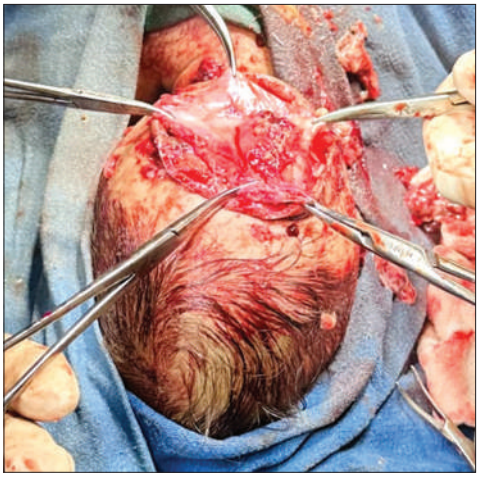

Figure 5: Excision of gliosed brain tissue

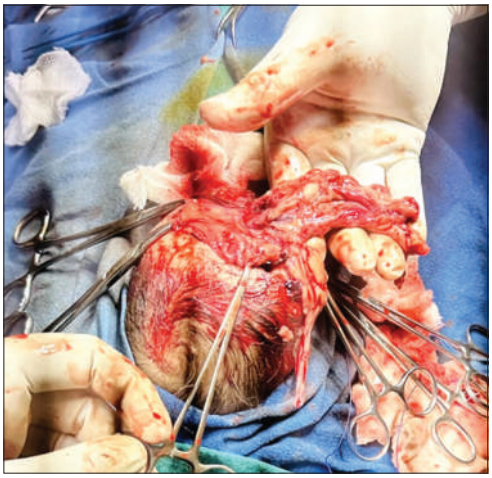

Figure 6: Identification of defect and herniated brain matter after excision of extra skin and duramater

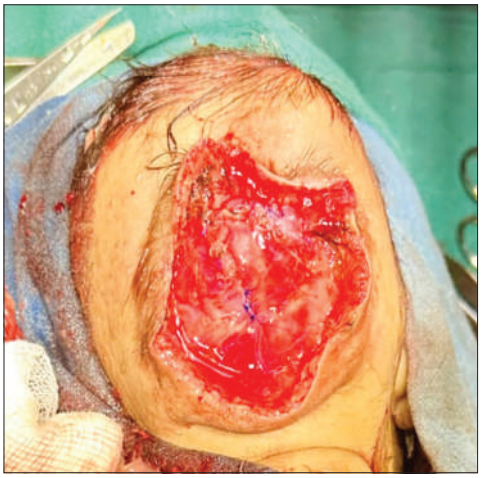

Figure 7: After repair of duramater

morbidity and fatality rates.[8] Post-operative complications might include infection, elevated intracranial pressure, apnea, cardiac arrest, and hypothermia so need to be effectively controlled. However, in our instance, there were no issues. Furthermore, despite our patient's current difficulties, intubation and anesthetic management were regularly accomplished.

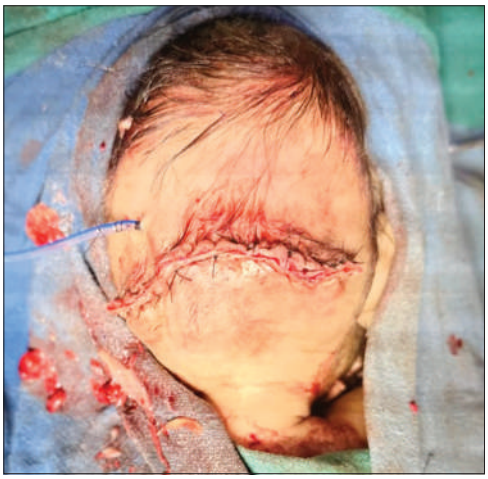

Figure 8: Skin closure after placement of drain

CONCLUSION

Managing a massive occipital encephalocele during surgery requires a large-scale collaborative effort from anesthesiologists and neurosurgeons. These patients have trouble getting into the supine position. In the lateral position, endotracheal intubation is accomplished. The degree of the cerebral contents' invasion into the sac is a key factor in the prognosis. The difficulty level of the surgical process has significantly increased due to the herniation of some brain regions. Infection risk is frequently increased in big encephaloceles, which are typically brought on by CSF leaking. It is incredibly beneficial to perform the operation at a young age to reduce unforeseen difficulties.

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