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Giant Occipital Encephalocele: A Study of 5 Patients

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Abstract: ***Background: When the occipital encephalocele is greater than or equal to the size of the head, it is referred to as a giant occipital encephalocele (GOE). Giant occipital encephaloceles are rare lesions; because of their enormous size they pose a surgical challenge. This series consists of 5 patients with giant encephaloceles treated at our institute. Material and methods: This was a retrospective observational study. The patients' head size, sex, and age at presentation were all assessed. We also checked for any related neural tube defects. Imaging was utilized to both design the surgical treatment and check for accompanying brain anomalies. It was also assessed whether a ventriculoperitoneal (VP) shunt was required. Results: During the study period of 1 years, 5 patients of GOE were admitted in our department. Most patients were <1 year of age. Among them, two were male and the remaining three were female. Of 5 patients who underwent surgery, excision and repair of GOE were performed as primary surgery in 4 patients. Of those 5 patients, one patients underwent VP shunt placement after excision due to raised intracranial pressure (ICP). VP shunt was placed as initial surgery in one patients, which was followed by excision and repair after about 5 weeks. Those patients who underwent VP shunt placement as primary surgery showed a decrease in the size of the encephalocele. The content of the sac was only CSF in 4 patients and CSF and gliotic brain tissue in remaining 1 patient. Associated cranial malformations included microcephaly, Chiari malformation, corpus callosal agenesis, and hydrocephalus. Hydrocephalus was symmetrical in all of them. One patient expired in the follow-up. The exact cause of mortality could not be ascertained. The probable reason may be the associated other congenital diseases. Conclusion: GOE is an unusual entity with limited information about its management. Careful evaluation, accurate imaging of the patient, and care throughout the intraoperative and postoperative periods, with an emphasis on factors influencing the prognosis, may lead to favourable result.***

Keywords: neural tube defect, encephalocele, hydrocephalus, ventriculoperitoneal shunt

1. Introduction

A congenital neural tube defect (NTD) known as an encephalocele is defined by the herniation or protrusion of the intracranial contents (meninges, brain, and part of the ventricles) through a defect in the cranium (1) . Failure of the growing neural tube's cranial portion to close during the first few weeks of fetal life results in encephalocele. When an encephalocele is greater than the size of the head, it is referred to as a "giant encephalocele" (2) . Massive encephalocele and large encephalocele also known as giant encephalocele (3) . The occipital area is where giant encephaloceles are most commonly seen. (2) (3) . A rare condition, giant occipital encephalocele is often described in case reports. We are presenting our case report of five patients of GOE managed at Seth Sukhlal Karnani Memorial Hospital, Kolkata over time with an emphasis on practical problems faced in its management.

2. Materials and Methods

The neurosurgery department at our institute conducted this retrospective observational study from July 2021 to July 2022. Five patients with encephaloceles in all received treatment during the course of this 12 - month period. In this study, all GOE patients were included. Patients were assessed for head size, sex, and age upon presentation. All of the patients had MRI, CT, or both to evaluate any associated neural tube defects and plan for surgery. (**Figures No.1 and 2).** Patient's evaluation of encephalocele excision after being evaluated. Dissection of the pedicle was a part of the surgical procedure **(Figure 3a, 3b).** Excision of the gliotic brain tissue, careful cerebrospinal fluid (CSF) drainage (**Figure 4),** and closure of the defect removing the gliotic brain tissue, closing the wound, and the GOE management procedure **(Figure 5a and 5b).** Based on ventricular/biparietal ratio on a CT/MRI basis, a ventriculoperitoneal (VP) shunt was required. Based on this, VP shunting was carried out before the removal of the encephalocele if there was significant hydrocephalus. If the hydrocephalus was mild or moderate, excision was done, followed by monitoring. A repeat scan was done after 48 hours in the non - shunted group. VP shunting was carried out in the case that severe hydrocephalus progressed. In the event of nonprogression, patients were followed up on every six months or if an issue arose for the attendants. Patients received intravenous antibiotics that covered both gram - positive and gram - negative organisms in the event of skin infection or encephalocele ulceration. For wound healing, local cleaning and the use of topical antibiotic cream were utilized. An outpatient follow - up was used for the follow - up. In the case of a delayed visit to the outpatient department, a telephone call with the parents was used. For the first six months, patients were phoned every month or earlier if there was any evidence of a problem by the attendants.

Procedure and outcomes

During the study period of 1 years, 5 patients of GOE were admitted in our department [Table 1]. Most patients were <1 year of age. Among them, two were male and the remaining three were female. Of 5 patients who underwent surgery, excision and repair of GOE were performed as primary surgery in 4 patients. Of those 5 patients, one patients underwent VP shunt placement after excision due to raised intracranial pressure (ICP). VP shunt was placed as initial surgery in one patients, which was followed by excision and repair after about 5 weeks. Those patients who underwent VP shunt placement as primary surgery showed a decrease in the size of the encephalocele. The content of the sac was only CSF in 4 patients and CSF and gliotic brain tissue in remaining 1 patient. Associated cranial malformations included microcephaly, Chiari malformation, corpus callosal

agenesis, and hydrocephalus [**Table 1].** Hydrocephalus was symmetrical in all of them. One patients expired in the follow-up. The exact cause of mortality could not be ascertained. The probable reason may be the associated other congenital diseases.

Table 1: Patients of giant occipital encephalocele

n	Age/sex	CT/MRI	Surgery	Outcome	Associated findings	Long-term follow-up
1.	2month/female	CT/MRI	VP shunt. Excisionafter5week	Uneventful	Chiari malformation	Delayed Milestone, Mental retardation
2.	1 months/male	CT/MRI	Excision	Post - operative, Surgical site infection	Nohydrocephalus	Delayed milestones, mental retardation
3.	1 month/female	CT/MRI	Excision	Uneventful	No hydrocephalus	Delayed milestone, Mental retardation
4.	3 month/male	CT/MRI	Excision	Uneventful	Chiari malformation	Delayed milestones, mental retardation
5.	2 month/female	CT/MRI	VP shunt. Excision after 2weeks	Post - operative, Surgical site infection, CSF leak	Microcephaly, hydrocephalus	Expired after 10days due meningitis

CT: Computerized tomography, MRI: Magnetic resonance imaging, VP shunt: Ventriculoperitoneal shunt, CSF:

Figure 1: Clinical photograph of a case of occipital giant encephalocele.

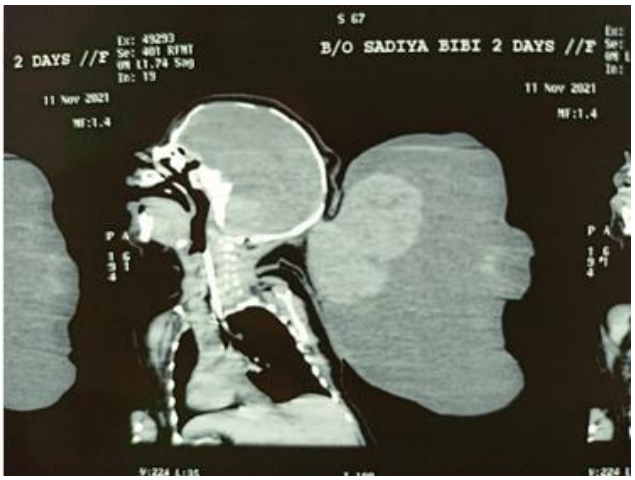

Figure 3 (a): MRI of occipital encephalocele.

Figure 2: Clinical photograph of a case of occipital giant encephalocele

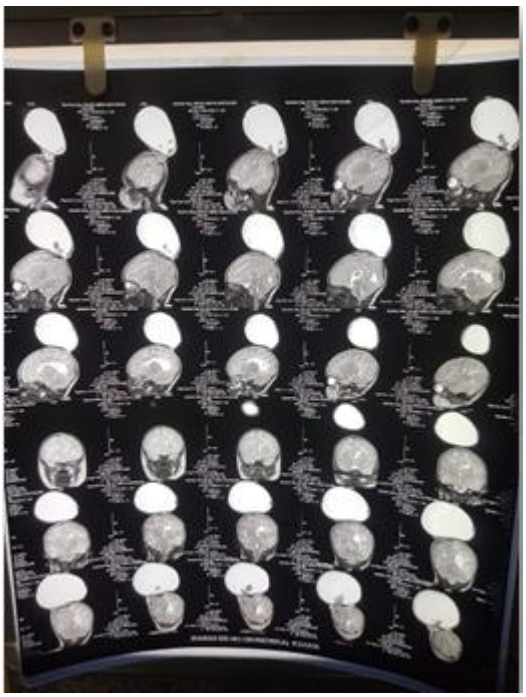

Figure 3(b): MRI of occipital encephalocele.

Figure 4: Cerebrospinal fluid (CSF) drainage

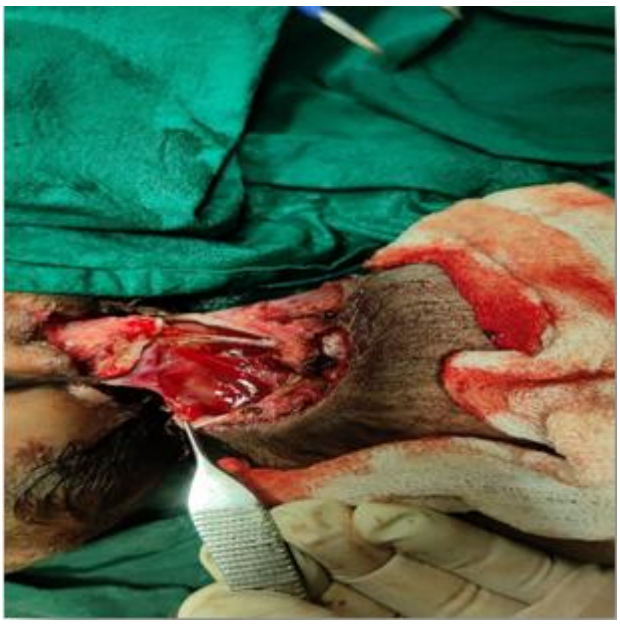

Figure 5 (a): Intraoperative photograph of a case of occipital encephalocele showing flap dissect

Figure 5 (b): Intraoperative photograph of occipital giant encephalocele after excision of sac

3. Discussion

There are several cases of encephaloceles. Although rare, giant encephaloceles occur when the encephalocele reaches a large volume (4) . These conditions can be termed massive encephalocele (5) . Large encephalocele and giant encephalocele all of which, of course, refer to the same condition and present similar challenges (6) . They pose a great challenge to the neurosurgeon, neuroradiologist, and neuroanesthetist during surgery. This series highlights the clinical features and management problems of these patients.10%–20% of all dysraphisms are caused by encephalocele, a type of neural tube defect. Due to the size of the sac, the amount of neural tissue within, blood loss, and hypothermia, giant encephalocele poses a challenge. In contrast to the frontoethmoidal forms, which are more widespread in South and Southeast Asia, OEs are more common in the Western Hemisphere (7) . Other GOE - related defects include hydrocephalus, Dandy - Walker cyst, Chiari malformation, craniosynostosis, and microcephaly (3) . [**Table 1]** shows that many patients appeared following the neonatal period. When asked why the presentation was so late, the answers given were inadequate referral and cost concerns that inhibited early referral. In our center's catchment region, the cost element has become a major problem. Clinically, there is a massive cystic swelling on the back of the skull, posing a major health risk to the attendants. Neonatal transportation is difficult because of the massive occipital edema. We have seen some astute attendants who brought the newborn on a huge tray to prevent the chance of sac rupture. Larger size is a risk factor for sac rupture, as shown in two of the patients in this series. Because a rupture might predispose a patient to meningitis, preventive antibiotics can be life - saving. Pressure ulcers may also develop. The same event occurred to two other patients. These pressure ulcers may require early treatment to avoid skin infection. Intensive therapy may not be required in the event of GOE excision. The goal is to reduce the risk of postoperative infection of the incision site or the brain. Torsion of the encephalocele pedicle is a rare but significant cause of skin necrosis. (8) . Torsion of the pedicle is not often discussed, yet firsthand knowledge of this phenomenon may aid in avoiding its recurrence. A CT scan of the head is important since it is quick, affordable, and widely available. Furthermore, it may be able to delineate the size of the bony defect (3) .

MRI, on the other hand, is an excellent study for seeing the contents of the sac and its link to the venous sinuses. (9) (10) . The sac is sometimes surrounded by the sagittal sinus, torcular sinus, and transverse sinus. These can be assessed prior to surgical intervention using imaging modalities (11) . In these patients, endotracheal intubation is a major problem. To avoid any compression or rupture of the sac, the child cannot be positioned supine and must be in a lateral position for intubation. A posture in which the infant is placed at the end of the table with a hyperextended neck, as described in a few series, may be effective for intubation. It has been proposed that CSF from the encephalocele sac be aspirated prior to surgery to facilitate intubation (11) (12) . Other perioperative issues include serum electrolyte imbalance, which can develop during CSF draining, hypothermia, and blood loss. (9) (10) . Apart from that,

cardiovascular issues such as cardiac arrest, tachycardia or bradycardia, and hypotension may occur. Hypoxia, hypercarbia, and endobronchial intubation have all been reported as respiratory complications (12) . Inhalational anesthesia has been recommended as a desirable technique since it is easily washed off. Our anesthesiologists were able to intubate patients who were in a lateral posture. The surgical intervention technique in GOE is determined by several criteria, including the quantity of neural tissue in the sac, the state of the CSF pathway, the patient's neurological status, and the presence or absence of related congenital defects elsewhere in the body (11) (13) . Fortunately for us, the majority of our patients had mainly CSF as a GOE component. Aside from that, gliotic brain tissue may be removed with no distinct neurological deficit the size of the sac, neural tissue content, hydrocephaly, infection, and pathologies that accompany this condition are well recognized factors that impact the prognosis of individuals diagnosed with OEs. (9) .

4. Conclusion

GOE is an unusual entity with limited information about its management. Careful evaluation, accurate imaging of the patient, and care throughout the intraoperative and postoperative periods, with an emphasis on factors influencing the prognosis, may lead to favorable results.

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