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CASE REPORT

Giant Occipital Encephalocele: A Case Report and Literature Review

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Background: Protrusion of cerebrospinal fluid and meninges is called meningocele. Meningoencephalocele is a protrusion of neural tissue and meninges. The incidence of an Encephalocele is 1 in every 5000 live births. Anterior encephalocele is more common in men, while occipital encephalocele is seen in 70% of women. In a large encephalocele, the head size is small. Encephaloceles can occasionally be very large and are called giant encephaloceles. Occipital encephaloceles accounted for 80% to 90% of encephalocele cases in the western hemisphere. Encephaloceles vary in size and content. Various factors affecting the prognosis of patients with occipital encephalocele. Among them: extent, amount of brain tissue in the sac with or without Dural venous sinuses in the sac, with the brain or occipital lobe with hydrocephalus or presence of ventricles. Rarely, the sagittal and transverse sinuses are adjacent to the sac.

Case Presentation: This is a case of an 8-month old male infant born to a mother who had no regular antenatal care. After the baby presented with progressively increasing posterior head mass which is cystic, transilluminating, tender and size is 40cmX35cm imaging demonstrated small brain tissue mainly part of right occipital lobe with most of the sac being occupied by cerebrospinal fluid. Repair done and dysplastic brain tissue resected then healthy-looking brain tissue and Dural sinuses reduced to the skull, then patient stayed in the hospital and closely followed for hydrocephalus for seven days and discharged with no hydrocephalus and no neurologic deficit.

Keywords: occipital, encephalocele, agiant

Introduction

Encephalocele is a birth anomaly in which protrusion of brain tissue and meninges is caused by a defect in the skull. It is responsible for 10–20% of all dysraphism. Occipital encephalocele is caused by bone defects in the occipital bone which sometimes extend to the foramen magnum and the back of the atlas. Occipital encephalocele comprises 75–85% of lesions in the Western countries. There could be associated lesions like; Hypoplastic corpus callosum, hypoplastic cerebellum etc. Hydrocephalus can occur due to the stenosis of the aqueduct. Hydrocephalus also occurs after encephalocele resection due to alteration in cerebrospinal fluid flow dynamics.[1] MRI of the brain helps in identifying the contents of the sac including; brain tissue, ventricles, Dural venous sinuses and other vessels like PICA. Surgical repair is important to prevent infection, defect closure and protected brain growth, however hydrocephalus must be excluded before a patient is discharged.

Case Presentation

This is an 8-month-old male patient born to a para 3 mother. The mother had ANC follow-up, but it was irregular, and it was at a rural health center with a poor setup where she could not get obstetric ultrasonography. The delivery was at home with a traditional birth attendant. After delivery, the parents noticed a swelling on the occipital part of the head. It increased in size progressively to attain the current size. The parents went to the health center, where they referred them to the primary hospital, which then referred them to our center. The swelling had no discharge, and the patient has no fever, vomiting failure to suck, high-pitched cry, abnormal body movement, or lethargy. Upon examination of the patient, there is a giant 40cm by 35cm cystic, transilluminating to light, tender, occipital area mass on the area of the torcula of the sinuses (see Figure 1). There is

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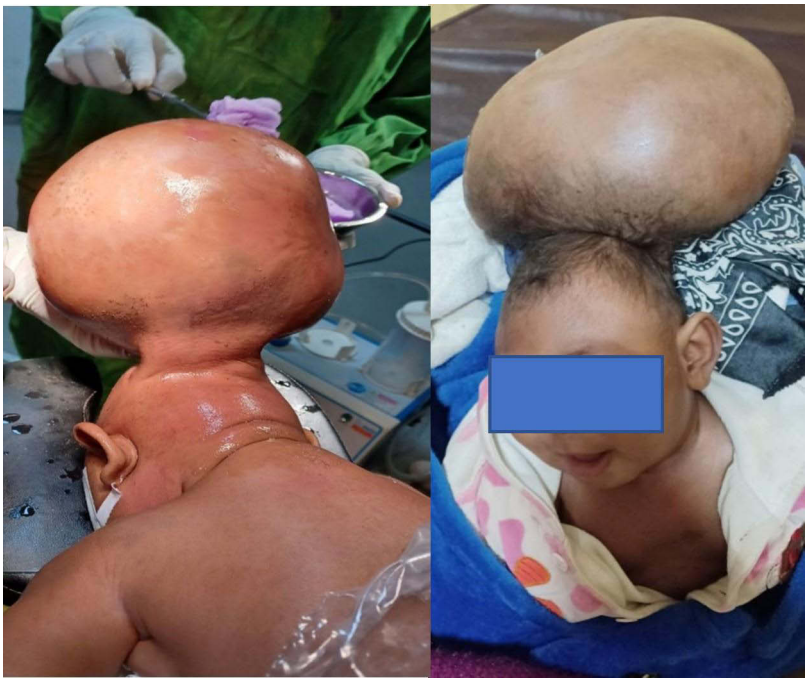

Figure 1 Preoperative patient photo.

a palpable bone defect at the sac's base. The anterior and posterior fontanels are not open. The baby is playful, alert moving all extremities, sucking well, and has stable vital signs. The patient was then investigated with a CBC, blood group, renal and liver function tests that were in normal range, and brain MRI revealed a huge occipital T1 hypointense and T2 hyperintense sac content with multiple flow voids showing torcula, superior sagittal sinus, and transverse sinus, and a part of right occipital lobe brain tissue with a bone defect at the base of the sac (see Figure 2). After getting informed written consent, the patient is put on the operating table with the head hanging beyond the operating table edge on the side of the anesthesia machine, with one

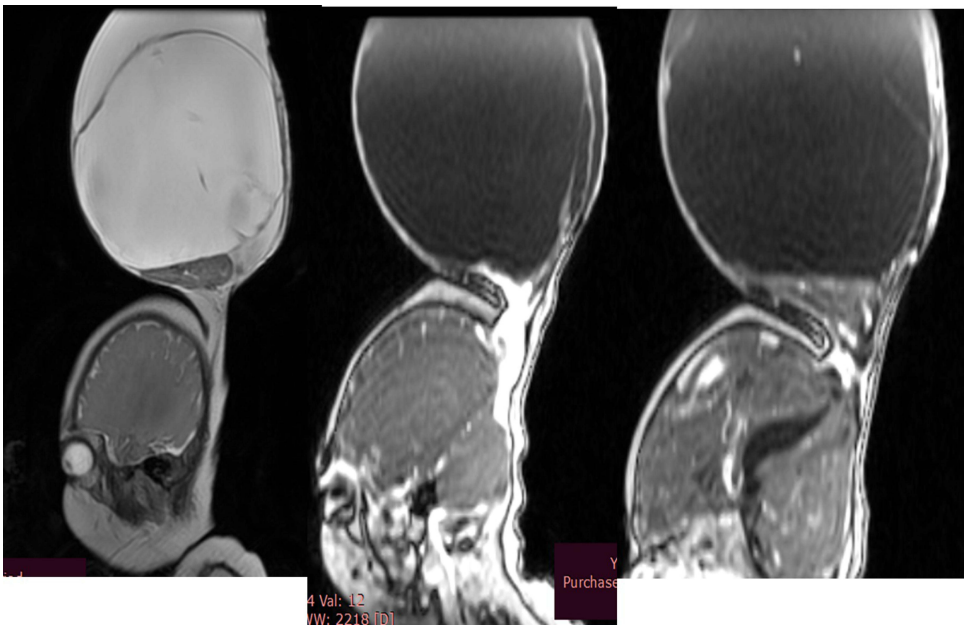

Figure 2 Preoperative patient MRI images: t2 hyperintense sac content which is a CSF with small t2 hypointense sac content which is a brain parenchyma, post contrast image showing the torcula, venous sinuses.

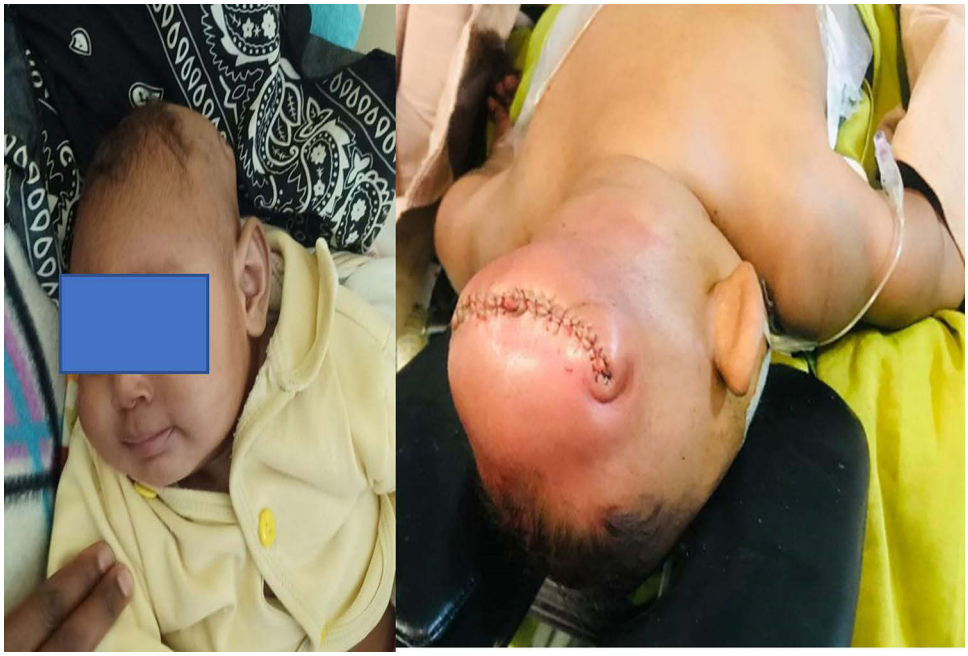

Figure 3 Postoperative patient photo.

anesthetist holding the head and the sac hanging beyond the table, and is intubated with the first attempt. After intubation, the patient was put in a prone position with the aid of a horseshoe Mayfield. Hair shaved and cleaned with soap and water, and one assistant holding the mass with a sterile gloved hand, the skin is then cleaned and draped. Initially, we made a transverse skin incision, drained the CSF, and the sac collapsed. Then, dysplastic brain tissue was resected, and venous sinus, tumor, torcula and other viable-looking brain tissue were gently reduced to the skull gently. Dura dissected, trimmed, trimmed and closed as a watertight primary. We did not recommend cranioplasty in a giant occipital encephalocele with a high risk of postoperative hydrocephalus; hence, we did not close the bone defect. After that, the skin was trimmed and closed in two layers (Figure 3). Postoperatively, the patient stayed in the neurosurgery ward for seven days with no hydrocephalus and no neurologic deficit. The patient is then followed at an outpatient clinic every 2 weeks with no surgery-related or neurologic complications.

Discussion

A giant encephalocele was first described in 2002 by Mahapatra. He defined a giant encephalocele as being as large as the head. Giant encephalocele is a very rare disease. The challenge for neurosurgeons originates from the contents of the sac, perioperative blood loss, and perioperative hypothermia.[2] Encephalocele occurs when there is herniation of the meninges and neural tissue via the skull defect. Occipital encephaloceles are defined as giant by other authors when they are larger than the head of the patient.[3] The occipital area is the most common (75% of patients). Followed by fronto-ethmoidal (13–15%), parietal (10–12%), and sphenoid areas. Occipital encephalocele is associated with significant morbidity and mortality.[4] Folic acid supplementation, antenatal and prenatal care, and the legal permission of abortion in developed countries have led to a reduction in encephalocele and other neural tube disorders. The incidence of neural tube defects and large encephaloceles is higher in developing countries due to the above factors and many other genetic and environmental reasons. The true cause of encephalocele is not clear and can be caused by many factors, including genetics and the environment. People with a family history of neural tube defects are more likely to have an encephalocele. Some investigators have suggested that nonenvironmental factors (hyperthermia, aflatoxin, genetics, maternal malnutrition, or other environmental factors) influence encephalocele formation. Even some toxins or diseases can play a role in its emergence.[3] It occurs as a result of the failure of the cranial part of the neural tube and usually occurs in the first 22–28 days of development. Some authors argue that encephaloceles are not neurulation defects and must not be classified as NTDs. The major challenges of treating patients with giant encephalocele include the

psychosocial problem of having a child with a "monstrous outlook" or "two heads" and the nursing care. This is most often seen in developing countries and can occasionally be attributed to sinfulness. The preoperative difficulties include day-to-day care, the risk of rupture with local infection or meningitis, ventriculitis, sepsis, the perioperative risk of anesthesia, blood loss, fluid shift with third space loss, and neurologic deficit. Care is generally not easy, especially in growing countries with poorly equipped operating rooms and ICU setups.[5] Isolated encephaloceles are sporadic in their occurrence most of the time. The types of encephalocele are sincipital (fronto-ethmoidal), basal (transsphenoidal, spheno-ethmoidal, trans-ethmoidal, and sphenoorbital), and occipital. Anterior encephaloceles have an equal sex ratio, while 70% of occipital encephaloceles are seen in females.[6] Patients present usually with posterior head cystic mass in the first week of life, with or without rupture of the sac, failure to suck, CSF leak, meningitis, sepsis, high-pitched cry, change in mentation, macrocephaly if there is hydrocephalus or microcephaly if there is large brain tissue in the sac, seizures, particularly for those with concomitant CND and other system malformations and anomalies. Congenital heart disease should be ruled out before surgery (Table 1). During pregnancy, encephalocele can be picked up

Table 1 Previous Case Reports with a Giant Occipital Encephalocele

Year	Authors	Imaging Contents and Size	Age/Sex	Presence of Hydrocephalus and Shunt	Cranioplasty Done	Outcome	Intraoperative and Postoperative Incident and Complications
2022	H.D. Nath et al[4]	SIZE-45X21cm CONTENTS-occipital lobe, choroid plexus, gliotic brain, 1100cc CSF	14 days Sex-not mentioned	Not mentioned	Not done	Good	No complication
2005	Lt col bipin et al[3]	Size-17cmX10cm Associated brain anomalies and CSF	3 days old / female	Not mentioned	Done with calvarial split	Good	No complications
2012	Hukum sing et al[1]	Size-22cmX13cm Contents-CSF and small brain tissue	5-month old/male	Postoperative hydrocephalus shunted	Not done	Good	Post op hydrocephalus
2019	Davendran et al[6]	Size-44cm Contents- cerebellum Associated anomalies	1-day old/ female	Postoperative hydrocephalus shunted	Not done	Good	Post op hydrocephalus
1992	Anthony et al[8]	Not mentioned	Not mentioned	Postoperative hydrocephalus shunted	Done with tantalum mesh	Good	Post op hydrocephalus
2021	Khizar et al[9]	Size-21x15x19cm Contents-Brain tissue	4 months/ male	No hydrocephalus	Not done	Good	No complication
2022	Durga et al[10]	Size-20x15x17cm CSF and brain tissue	1 month/ male	Post operative hydrocephalus shunted	Not mentioned	Good	Post op Hydrocephalus
2019	Vikas et al[11]	Size-26x30x34cm CSF, small brain, hydrocephalus, syrinx	6 months/ female	Preoperative hydrocephalus shunted	Not mentioned	Good	No complication
2008	Yasir et al[12]	Ulcerated 38cmx60cm	1-day old/ male	Not mentioned	Not mentioned	Good	Not mentioned
2005	Arvind et al[7]	Gangrene, ruptured 15x10cm	14 days/ male	No hydrocephalus	Not done	Good	No post op complication
2023	Harsh et al[2]	Ulcerated, dark, twisted large	2 days/ male	Not mentioned	Not done	Good	No complication

early with the aid of ultrasound, maternal serum alfa fetoprotein, and amniocentesis.[7] Routine investigations, including CBC, organ function tests, transfontanel ultrasonography, and echocardiography, have to be done for all patients. Before surgical intervention, imaging has to be done to identify the bone defect, its exact location, and the contents of the sac. Of all the imaging modalities, brain MRI is the safest, with no radiation exposure for these patients. On encephalocele MRI, characteristic signals of neural tissue are demonstrated, including the presence of hydrocephalus, the presence of other brain malformations, surgical planning, and CSF in the sac. MR venography characterizes the Dural venous anatomy and if they are contained in the sac and see other vessels like PICA. CT mainly demonstrates the bone margins, the bone defect, and the brain tissue and sac content. CT is not usually indicated because of both the radiation exposure and its poor value in delineating soft tissue disorders. Transfontanel ultrasonography helps to see sac contents in resource-limited settings and helps in the detection of hydrocephalus and screening obstetric Ultrasonography will identify encephaloceles in the fetus.[6] A visual-evoked response is used to decide whether the sac has functional tissue from the visual cerebral cortex.[6] Delivery of a neonate with a giant encephalocele by cesarean section is recommended by most authors to avoid labor-related complications for both mother and neonate.[4] The counseling and the decision should involve family members and other health personnel. The surgical treatment of these children requires special consideration of peri-intubation challenges and surgical principles. Removal of a significant amount of CSF can result in volume and electrolyte abnormalities that need to be treated urgently.

Giant occipital encephaloceles are challenging to position for intubation. The occipital encephalocele poses a challenge by limiting the range of neck extension and flexion. Therefore, using laryngoscopy and getting the glottic opening are difficult. Due to the risk of compression and rupture, patients cannot be easily put in the supine position. There are various techniques for intubation, including intubating in a lateral position, placing the baby's head above the margin of the operation table, and raising the baby off the table with the help of two assistants.[4] Cleaning and draping can be performed by holding the mass with either forceps for small ones or a sterile gloved hand for large ones, which facilitates better cleaning.[7] Generally, patients should be positioned prone, and temperature and other patient conditions should be monitored. In patients with encephalocele, drainage of cerebrospinal fluid (CSF) prior to incision may assist in dissection. A transverse incision is ideal for round encephaloceles with small occipital defects. A vertical skin incision is recommended by the authors in patients with encephalocele extending above and below the posterior fossa.[8] Most often, we must be careful during occipital encephalocele dissection as brain tissue, Dural venous sinuses, and other neurovascular structures could be the contents of the sac. Preservation of neurovascular tissue is usually preferred as much as possible. Dura must be repaired for watertight closure.[8] Duraplasty can be done using a pericranial graft. Bone defect closure should not be attempted in infants and young children.[8] The most commonly used techniques are encephalocele resection and primary Dural repair. Expansion cranioplasty is the other surgical technique that uses a mesh to create room for the protruded sac. Another technique commonly used is ventricle size. It is a two-step procedure that first increases the ventricular pressure by inducing hydrocephalus, followed by the placement of a VP shunt. The ventricles then reduce in size, and the herniated tissue retracts itself back inside the skull. Occasionally, an incision is made in the tentorium to release an infratentorial area of the protruded tissue.[13] The surgical planning and decision depend on different issues, including cosmesis, skin lesion, imminent rupture, meningitis, necrosis, size, herniated tissue, ventricles, torcula, or sinus, clinical condition of the patient, hydrocephalus, associated CNS anomalies, and associated congenital anomalies in other parts of the body. It is advised to excise only the dysplastic tissue and replace it with healthy neurovascular structures. Pathological examination revealed a large encephalocele with degenerative cerebral cortex, which was thought to be dysfunctional. Surgical procedures have been described in the literature to protect the cortex from further damage. Surgery for a patient with a large occipital encephalocele with a large amount of brain tissue protruding into the sac will be very difficult. The head is small and the tissue of the brain is very large; therefore, it is difficult to reduce all cystic elements. Another difficulty is that intracranial blood vessels can enter and exit the sac to feed the brain parenchyma. Therefore, the patient's MRI and MRV should be thoroughly studied before surgery to avoid damage, and cerebrospinal fluid should be removed as quickly as possible during surgery to facilitate surgery. Most of the dysplastic content has to be resected; however, healthy neurovascular tissues need to be replaced.[13] If major blood vessels are injured, brain resection can lead to infarction. If there is functional brain tissue in the sac, techniques such as "cranioplasty" and "ventricular volume reduction" can be used to preserve the protruding brain.[4] Most patients show significant tissue gliosis, and excision is recommended. The dura mater should be closed watertight with pericranial tissue.[10] The dura mater is osteogenic; bone defect healing can occur over time, and cranial split grafts and cranioplasty are not usually necessary[1]

(Table 1). Cranioplasty may not be necessary if the skull is small. Cranioplasty can be done to close the defect for aesthetic reasons and to protect the brain. This can be done during surgery or later, after the encephalocele has been removed. In the literature, methyl methacrylate, hydroxyapatite bone cement, demineralized bone matrix, tantalum mesh, autografts (such as ribs), or autologous calvarial bone grafts have been used. In our patient, cranioplasty was not performed because of the small bone defect. When the skin is insufficient for initial closure and a flap or advancement is required, a multidisciplinary team of plastic and reconstructive surgeons always achieves appropriate results.[1] No aesthetic problems were observed in the occipital bone in the postoperative period. Intraoperative blood loss and cerebrospinal fluid loss, hypothermia, bradycardia, and cardiac arrest are the most important complications in large occipital encephalocele surgery. Blood loss from large cysts and loss of cerebrospinal fluid can cause electrolyte disturbances and hemodynamic instability. Such patients may develop hypothermia due to large surface exposures.[4] Note hypothermia, increased intracranial pressure, apnea, cardiac arrest, cerebrospinal fluid leak, and postoperative infection perioperatively.[10] For large occipital encephaloceles, the outcome is often poor, even with surgery, with a high risk of death and complications, as well as mental and/or physical disability. In particular, patients with larger encephaloceles having cerebellum, cerebrum, brain, and ventricles in the sac have a poor prognosis.[10] Clinically significant hydrocephalus occurs mostly postoperatively (Table 1). The size of the encephalocele and the amount of neural tissue found in the brain sac are associated with prognosis.[4] The neurological outcome depends on the presence or absence of hydrocephalus, while the cognitive level is usually related to the absence of the brain in the sac. Mahapatra et al In their series, 67% of all patients with massive occipital encephaloceles reported normal development, while 33% showed abnormal growth.[4] Hydrocephalus occurs in 60% to 70% of posterior encephalocele patients and requires VP shunt surgery. Large occipital encephaloceles and small head with large brain tissue have a bad prognosis.[1]

Conclusion

A giant occipital encephalocele is not a common disease. It is challenging for anesthesiologists, neurosurgeons, and plastic and reconstructive surgeons. Because it is multifactorial, including environmental and genetic factors, fortification of diets with folic acid has decreased the incidence in developed countries. The bone defect and its relationship with the torcula should be well identified and planned before surgery. Intubation at the edge of the operating table is one of the most commonly used techniques. Cranioplasty is not needed mostly as the dura is osteogenic in infants; skin closure may need the involvement of a plastic and reconstructive surgeon; and strict follow-up for worsening of previously existing hydrocephalus or new-onset postoperative hydrocephalus is recommended.

Abbreviations

CBC, complete blood count; CNS, central nervous system; MRI, magnetic resonance imaging; CT, computed tomography; US, ultrasonography; OT, operating room; CSF, cerebrospinal fluid; MRV, magnetic resonance venogram; maternal serum alfa fetoprotein (MSAFP); PICA, posterior inferior cerebellar artery; ANC, antenatal care.

Informed Consent

Informed written consent is provided by participants' parents to have the case details and any accompanying images published.

Disclosure

The authors declare that they have no conflicts of interest.

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