

CASE REPORT

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Giant occipital encephalocele: a case report, surgical and anesthetic challenge and review of literature

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Abstract

Background: An encephalocele is a congenital neural tube defect characterized by herniation of cranial contents through a defect in the cranium and is caused by failure of the closure of the cranial part of the developing neural tube. An encephalocele is termed as “giant encephalocele” when the size of encephalocele is larger than the size of the head. They depend on size of the sac, percentage of neural tissue content, hydrocephalus, infection, and other associated pathologies for a favorable neurological outcome.

Case presentation: We report a case of a four-month-old boy with a giant occipital encephalocele measuring 21 × 15 × 19 cm in size, which was a surgical and anesthetic challenge for us. Intubation was achieved in lateral position. Part of occipital and cerebellar parenchyma was present in the sac and bony defect was approximately 2.5 cm in occipital bone in midline. We performed surgical excision and repair with a good overall outcome.

Conclusion: Perioperative management of a giant occipital encephalocele is a challenge for both anesthesiologists and neurosurgeons. Managing such a case demands a search for other congenital abnormalities, expertise in handling airway, and proper intraoperative care. Careful planning and perioperative management are essential for a successful outcome.

Keywords: Giant, Encephalocele, Meningoencephalocele, Neural tube defect

Background

Encephalocele is a rare neural tube defect, typically occurring in one per every 5,000 births worldwide, of which 70% are occipital [1]. It is classified by the herniation of multiple cranial contents in the initial weeks of fetal life, through a defect in the cranium caused by inappropriate closure of the developing cranial part of the neural tube [2]. The size of an encephalocele varies from a few centimeters to an enormous swelling and is termed as “giant encephalocele,” when the size of encephalocele is more substantial than the size of the head [2].

Such cerebral malformations are dependent on their size of the sac, percentage of neural tissue content,

hydrocephalus, possible infections, and other associated pathologies for a favorable neurological outcome. Preoperative neurological status of the patients and the cranial contents herniating into the sac remain the vital factors in deciding the long-term prognosis. The anesthetic management of occipital meningoencephalocele presents a challenge due to apparent difficulty in satisfactorily establishing the airway while being in a prone position, blood loss, and perioperative care [3].

Case presentation

A 4-month-old baby boy presented to us through the pediatric emergency department following a fall from his mother's lap. This baby boy sustained a huge swelling on the back of his head since birth, and it increased in size over a period. There was no history of antenatal checkups and baby was delivered via cesarean section

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at term in a local hospital and was examined by a local pediatrician. He was not advised for any neurosurgical opinion and was discharged. The child referred to the neurosurgery department after a history of fall from his mother's lap due to the presence of a challenging swelling on his head.

On examination, the child was active. He had a huge occipital swelling measuring about $21 \times 15 \times 19$ cm in size (Fig. 1A, B). Swelling was cystic and transilluminates. The overlying skin was intact, though few areas of skin necrosis due to pressure were seen. No apparent traumatic injury 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. With the aforesaid clinical findings, a diagnosis of giant occipital encephalocele was established.

Routine investigations were normal. His magnetic resonance imaging (MRI) brain plain was done which reported a cerebrospinal fluid (CSF) containing sac with the brain parenchymal tissue herniating through a defect in posterior occipital bones in midline. It measured approximately $21 \times 15 \times 19$ cm. Bilateral lateral and third ventricles were normal. Consequently, it was concluded it to be a huge meningoencephalocele without hydrocephalus. (Fig. 2A, B).

Parents were counselled in detail about the diagnosis and after obtaining informed written consent, we proceeded to perform surgical excision and repair. It was a significant challenge for our anesthetist to intubate this child. Intubation was achieved in lateral position and was supervised in same position throughout. The surgical area was cleaned and draped while holding the encephalocele in hands. Incision was performed along the neck of the sac and deepened through

skin layers. Dissection done layer by layer and excised the sac along with dysplastic brain tissue was excised. Part of occipital and cerebellar parenchyma was discovered in the sac. Dural venous sinuses were not involved. Dural layer was precisely identified, and brain tissue was reduced through a bony defect of 2.5 cm size in occipital bone in midline and closure done with coated Vicryl (polyglactin 910) 4-0 sutures. Valsalva maneuver performed and no CSF leak was noted. Subcuticular layer was closed with Vicryl (polyglactin 910) 3-0 sutures. Skin closure was done with Prolene (polypropylene) 3-0 sutures. The child had an uncomplicated extubation and started to feed 3 h after the surgery. There was minimal blood loss during the surgery, hence blood transfusion was not done. The baby didn't develop hydrocephalus and was discharged after 3 days.

Discussion

The meningeal membrane covering the giant encephalocele is either covered by a normal, dysplastic or a thin membrane. Clinically, the presentation of giant occipital encephaloceles is evident because of their characteristic swelling [2]. The contents of occipital encephalocele primarily include meninges and occipital lobes. It may also comprise of ventricles, cerebellum, and brain stem. The key factors influencing the outcome of patients with occipital encephalocele are site, size, and herniation of the brain into the sac. The presence of the brainstem or occipital lobe with or without the dural sinuses in the sac along with hydrocephalus also influences the outcome of the case [4].

The large-sized swellings may conceivably have a remarkable brain herniation, an abnormality of the underlying brain and microcephaly. The clinical examination incorporates the examination of its size, extent,



Fig. 1 A Giant occipital encephalocele. B Giant occipital encephalocele ($21 \times 15 \times 19$ cm in size)

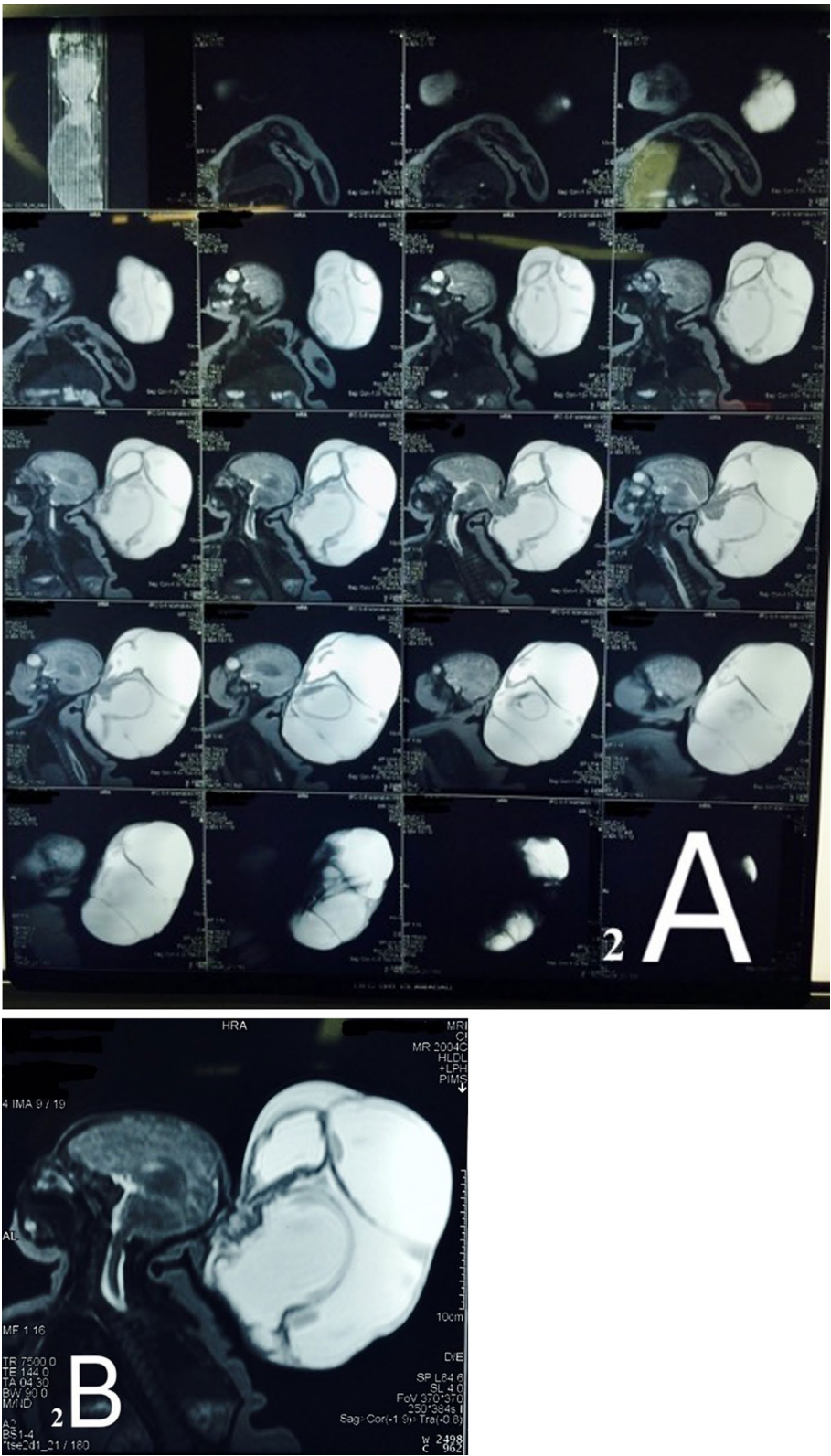


Fig. 2 **A** MRI T2 weighted image of Giant Occipital Meningoencephalocele. **B** MRI T2 weighted image showing a huge meningoencephalocele without hydrocephalus

Table 1 Overview of past reported cases of occipital encephalocele

Case No	Year	Age	Site	Associated Findings	Surgery
1 [18]	2002	26 days	Occipital	No brain tissue in the sac, hydrocephalus	Excision; second surgery; VP shunt
2 [18]	2002	3 months	Occipital	Hydrocephalus	VP shunt; surgical excision and repair
3 [18]	2003	10 days	Occipital	No associated findings	Surgical excision and repair
4 [18]	2003	9 months	Occipital	Polygyria; microgyria; craniostenosis	Surgical excision and repair; craniotomy; cranioplasty by autologous bone
5 [18]	2004	2 ½ months	Occipital	Hydrocephalus	VP shunt; surgical excision and repair
6 [18]	2005	4 months	Occipital	Hydrocephalus; craniostenosis	Surgical excision and repair; craniectomy
7 [18]	2005	5 months	Occipital	Hydrocephalus; craniostenosis	Surgical excision and repair; craniectomy and cranioplasty
8 [18]	2006	3 months	Occipital	No hydrocephalus; craniostenosis	Craniectomy; excision; cranioplasty by methyl methacrylate
9 [18]	2007	1 month	Occipital	Hydrocephalus	VP shunt; Surgical excision and repair
10 [18]	2008	1 year	Occipital	Hydrocephalus	Surgical excision and repair. VP shunt 4th day
11 [18]	2008	4 years	Occipital	No associated findings	Repair of encephalocele
12 [18]	2009	4 months	Occipital	Microcephaly; craniostenosis; small bony defect	Surgical excision and repair; craniectomy
13 [18]	2009	20 days	Occipital	Microcephaly	Surgical excision and repair; patient had intraoperative hypothermia
14 [12]	2010	4 months	Occipital	No associated findings	Sac was opened. The herniated brain tissue looked redundant. The sac was reduced in size, sufficient enough to accommodate the healthy-looking brain tissue. The skin was closed with interrupted sutures
15 [4]	2014	14 days	Occipital	History of limb weakness. There was no bladder and bowel dysfunction	Under general anesthesia with prone position excision and repair of sac was done
16 [3]	2014	1 month	Occipital	No associated findings	The dysplastic portion of the occipital lobe was removed, and tight dural closure was achieved in two layers
17 [11]	2014	6 day	Occipital	Delayed cry at birth, hypotonic with decreased spontaneous movements and had respiratory distress	Dysplastic cerebellar tissue with a herniated sac was excised, and the primary closure of the defect was performed
18 [17]	2016	7 months	Occipital	Craniostenosis	Excision and repair of sac with strip craniectomy of sagittal and lambdoid suture
19 [15]	2016	40 days	Occipital	Persistent elevation of serum potassium levels	The child underwent complete surgical excision and repair of the meningoencephalocele
20 [20]	2016	< 1 month	Occipital	Posterior cerebellar artery and basilar artery into the defect	Excision and repair
21–22 [20]	2016	< 1 month	Occipital	No associated findings	Excision and repair
23 [20]	2016	< 1 month	Occipital	No brain tissue inside the sac, hydrocephalus	Excision and repair
24 [13]	2017	2 months	Occipital	No associated findings	A circumferential incision was given on the swelling with meticulous dissection. The protruded portion of cerebellum was excised, and the dural closure was done
25 [16]	2017	1 day	Occipital	No associated findings	The sac with dysplastic brain tissue was excised and closed in a single layer using continuous suture
26 [16]	2017	1 month	Occipital	No associated findings	The sac was dissected out and the dysplastic brain tissue was excised. The sac was closed in a single layer using continuous suture, skin and subcutaneous tissue was closed

Table 1 (continued)

Case No	Year	Age	Site	Associated Findings	Surgery
27–55 [19]	2017	5 months (average)	Occipital	Hydrocephalus (19 cases) and delayed milestones (17 cases)	Excision and repair
56 [21]	2018	1 day	Occipital	No associated findings	No associated findings
57 [14]	2019	4 months	Occipital	No associated findings	The encephalocele was excised and the dura was identified, and a watertight closure was carried out
58 [9]	2019	6 months	Occipital	Delayed motor milestones in the form of inability to hold her head	Excision of the redundant neural tissue, and closure of the dura done and skin was reconstructed
59 [10]	2019	10th day	Occipital	No associated findings	A transverse incision was given over the parietal mass followed by surgical excision and repair
60 [6]	2019	3 months	Occipital	The child had a delayed milestone with poor feeding and absent neck holding. ECHO showed the presence of a small 5 mm ASD with L → R shunt	Posterior paramedian incision was given, encephalocele sac along with some brain tissue was excised followed by primary duraplasty

amount of prominence, and its location along with the size of bony defect [2]. The size of the head holds a significant importance for clinical suspicion of microcephaly or hydrocephalus and extracranial anomalies [2]. MRI brain remains the usual investigation of choice along with the three-dimensional computed tomography (CT) that further helps in evaluating the deformity [5].

Giant encephaloceles are rare; surgical procedures represents a challenging task for the anesthesiologists, as well as the neurosurgeons. The challenges present are principally due to its complex site, enormous size, associated bulging contents resulting in intracranial anomalies, intraoperative blood loss, and prolonged anesthesia [6]. The supreme aim of the anesthesiologist is to prevent premature rupture of the encephalocele intraoperatively. The occipital site of the encephalocele causes a hindrance with the intubation as due to a difficult airway caused by the restriction of the neck movement. This further leads to an inability of having an optimal tracheal intubation position [3].

The operative procedure additionally includes the management of possible loss of copious quantities of CSF causing superimposed electrolyte imbalance. Infants with encephalocele can develop sudden hypothermia due to dysfunction of autonomic control below the present defect [3]. Thus, urgent consideration and management must be given to hypothermia, blood loss, and its associated complications. The surgery is advised to be done as promptly as possible to escape life threatening complications such as central nervous system (CNS) infections, respiratory distress, aspiration pneumonia, irreversible impairment of vagus nerve, and hypothermia [5].

The table below incorporates all the reported cases to the author's knowledge. (Table 1) [3, 4, 9–21] The key aim is to portray the different and most commonly used surgical procedures in the management of giant occipital encephaloceles. The most frequently used method is sheer resection and dural repair. Expansion cranioplasty is one of the surgical procedures that consists of a mesh to provide room for the protruded sac. Another technique commonly practiced is executed through ventricular volume reduction. It is a two-step technique which first increases the ventricular pressure inducing hydrocephalus followed by insertion of a ventriculoperitoneal shunt. The ventricles then contract, and the protruded tissue repositions itself inside the cranium. For the herniated cerebellar and occipital parenchyma, an incision is required in the tentorium to establish an infratentorial area for the herniated tissue to retract [7].

Modern-day neurosurgical techniques along with neuroimaging and neonatal intensive care with neurological facilities have markedly improved morbidity and mortality rate in the management of encephaloceles

[8]. Postoperatively, complications such as hypothermia, raised intracranial pressure (ICP), apnea, cardiac arrest, CSF leak, and infection can arise and hence need to be managed effectively. However, in our case no complications were present. Alongside all present difficulties, intubation and anesthetic management in our patient were consistently achieved.

Conclusions:

Perioperative management of a giant occipital encephalocele is an extensive team challenge for both anesthesiologists and neurosurgeons. These patients encounter a difficulty in attaining the supine position. Endotracheal intubation is achieved in the lateral position. The prognosis is crucially dependent on the extent of the invasion of cranial contents into the sac. Because of the herniation of parts of the brain, there is a significant rise in the difficulty level of the surgical procedure. There is habitually a more remarked risk of infection involved in giant encephaloceles usually due to CSF leakage. To minimize superimposed complications, carrying out the operative procedure at a primitive age is immensely beneficial.

Abbreviations

CSF: Cerebrospinal fluid; MRI: Magnetic resonance imaging; CT: Computed tomography; CNS: Central nervous system; ICP: Intracranial pressure.

Acknowledgements

Not applicable.

Authors' contributions

SZ and AK wrote the manuscript. AK is involved in the surgical management of the patient. Both authors read and approved the final manuscript.

Funding

No funding was required for this work.

Availability of data and materials

All data are within the article.

Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

Written informed consent was taken from the mother of the child for publication of case report and accompanying images.

Competing interests

None of the authors has any conflict of interest to disclose. We confirm that we have read the Journal's position on issues involved in ethical publication and affirm that this case report is consistent with those guidelines.

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Received: 23 August 2021 Accepted: 4 October 2021

Published online: 28 November 2021

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