

Giant Occipital Encephalocele: A Case Report, Surgical & Anesthetic Challenge

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Case report

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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 about 21 x 15 x 19 cm in size, who 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 did Surgical Excision and Repair with a good overall outcome.

Conclusion: Perioperative management of a Giant Occipital Encephalocele is a challenge for both anesthesiologist and neurosurgeon. Managing such a case demands search for other congenital abnormalities, expertise in handling airway and proper intraoperative care. Careful planning and perioperative management are essential for a successful outcome.

Background:

Encephalocele is a rare neural tube defect, occurring in 1 per every 5,000 births worldwide from which 70% are occipital.^[1] It is characterized by the herniation of several 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 larger than the size of the head.^[2]

Such malformations are dependent on their size of the sac, percentage of neural tissue content, hydrocephalus, infection, and other associated pathologies for a favorable neurological outcome. Preoperative neurological status of the patients and the amount of cranial contents herniating into the sac are the vital factors in determining the long term prognosis. The anaesthetic management of occipital meningoencephalocele presents a challenge due to difficulty in securing 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 with the history of a fall from his mother's lap. This baby boy had a huge swelling on the back of his head since birth and it increased in size over the period of time. Mother had no antenatal check-ups. Baby was delivered by

Cesarean section at term, in some local hospital and was taken to a local pediatrician for evaluation. He was not advised for neurosurgical opinion and the child was taken back to home. He grew up till the age of 4 months, when it became difficult for the mother to nurse the child and he fell from her lap and was taken to another nearby hospital who referred the child to our neurosurgery department.

On examination, the child appeared active. He had a huge occipital swelling measuring about 21 x 15 x 19 cm in size. (Fig. 1,A & B) The overlying skin was intact, though few areas of skin necrosis due to pressure were seen. No obvious traumatic injury noted. Swelling was cystic and transilluminant. Anterior fontanelle was lax and open, measuring about 3.5 x 2.5 cm. Child was moving all four limbs, power and tone were normal. With the aforementioned clinical findings, a diagnosis of Giant Occipital Encephalocele was made.

Routine investigations were normal. His Magnetic Resonance Imaging (MRI) Brain Plain was done which reported a Cerebrospinal Fluid (CSF) containing sac with brain parenchymal tissue herniating through a defect in posterior occipital bones in midline. It measured approximately 21 x 15 x 19 cm. Bilateral lateral and third ventricle were normal. So, they concluded it to be a huge meningoencephalocele without hydrocephalus. (Fig. 2, A & B)

Parents were counselled in detail about it and after taking informed written consent, we proceeded for surgical excision and repair. It was a great challenge for our anesthetist to intubate this child. Intubation was achieved in lateral position. After intubation, the child was kept in lateral position. Cleaned and draped while holding the encephalocele in our hands. Incision was given along the neck of sac and deepened through skin layers. Dissection done layer by layer and excised the sac along with dysplastic brain tissue. Part of occipital and cerebellar parenchyma was found in the sac. Dural venous sinuses were not involved. Dural layer was identified, brain tissue 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 suture. Valsalva maneuver performed. No CSF leak noted. Subcuticular layer closed with Vicryl (polyglactin 910) 3 – 0 suture. Skin closure done with Prolene (polypropylene) 3 – 0 suture. Child had smooth extubation and started to feed after 3 hours of surgery. There was minimal blood loss during the surgery, so 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 due to their characteristic swelling.^[2] The contents of occipital encephalocele mainly include meninges and occipital lobes. It may also comprise of ventricles, cerebellum, and brain stem. The factors influencing the outcome in the occipital encephalocele patients are site, size and herniation of the brain into the sac. Site, size, and the amount of brain herniated inside the sac determines the prognosis. The presence of brainstem or occipital lobe with or without the dural sinuses in the sac along with hydrocephalus also influence the outcome of the case.^[4]

The large-sized swellings may possibly have a remarkable brain herniation, an abnormality of the underlying brain and microcephaly. The clinical examination comprises the examination of its size, extent, amount of protrusion and its location along with the size of bony defect.^[2] The size of the head holds a significance importance for clinical suspicion of microcephaly or hydrocephalus and extracranial anomalies.^[2] MRI brain is the usual investigation of choice along with the three dimensional Computed Tomography (CT) that further helps in evaluating the deformity and hence the surgical procedure can take place as soon as possible to avoid further neurological deficits.^[5]

Giant encephaloceles are rare; surgical procedures are a challenging task for the anesthesiologists, as well as the neurosurgeons. The challenges present are mainly due to its complicated site, enormous size, associated bulging contents resulting in intracranial anomalies, intraoperative blood loss, and prolonged anesthesia.^[6] The major aim of the anesthesiologist is to avoid the 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 also includes the management of possible loss of large 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 immediate consideration and management has to be given to hypothermia, blood loss, and its associated complications. The surgery is advised to be done as soon as possible to avoid 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, comprises all the reported cases to the author's knowledge. (Table 1)^[3, 4, 9-21] The main 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 simple 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 used is done through ventricular volume reduction. It is a two-step technique; for start it increases the ventricular pressure inducing hydrocephalus followed by 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 made in the tentorium to create an infratentorial area for the herniated tissue to retract.^[7]

Table 1

CaseNo.	Year	Age	Site	Associated Findings	Surgery
1 ^[18]	2002	26 days	Occipital	No brain tissue in the sac, hydrocephalus	Excision; 2 nd 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 4 th 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	Under general anesthesia with

				weakness. There was no bladder and bowel dysfunction.	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	Craniosynostosis	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.
27-55 ^[19]	2017	5 months (average)	Occipital	Hydrocephalus (19 cases) and delayed	Excision and Repair

milestones (17 cases)					
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 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.

Modern day neurosurgical techniques along with neuroimaging and neonatal intensive care with neurological facilities have greatly 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 anaesthetic management in our patient were successfully achieved.

Conclusion:

Perioperative management of a Giant Occipital Encephalocele is an extensive team challenge for both anesthesiologist and neurosurgeon. These patients have a difficulty in attaining the supine position. Endotracheal intubation is achieved in the lateral position. Prognosis is mainly dependent on the extent of the invasion of cranial contents into the sac. Due to the herniation of parts of the brain, there is a significant rise in the difficulty level of the surgical procedure. There is always a higher risk of infection involved in giant encephaloceles usually due to CSF leakage. In order to avoid superimposed complications, carrying out the operative procedure at an early age is highly beneficial.

Abbreviations

CSF: Cerebrospinal Fluid

MRI: Magnetic Resonance Imaging

CT: Computed Tomography

CNS: Central Nervous System

ICP: Intracranial Pressure

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.

Availability of data and materials:

All data is within the article.

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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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.

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References

1. Stephanie A, Black JA, Galvez MA, Rehman. Alan Jay Schwartz; Images in Anesthesiology: Airway Management in an Infant with a Giant Occipital Encephalocele. *Anesthesiology*. 2014;120:1504.
2. Ghritlaharey RK. A Brief Review of Giant Occipital Encephalocele. *Journal of neurosciences in rural practice*. 2018;9(4):455–6.
3. Pahuja H, Deshmukh. Shubhda & palsodkar, supriya & lande, surabhi. (2015). Anaesthetic management of neonate with giant occipital Meningoencephalocele.. *International Journal of Research in Medical Sciences*. 31. 1. 10.5455/2320–6012.ijrms20150165.
4. Nath HD, Mahapatra AK, Borkar SA. A giant occipital encephalocele with spontaneous hemorrhage into the sac: A rare case report. *Asian journal of neurosurgery*. 2014;9(3):158–60.
5. Velho V, Naik H, Survashe P, Guthe S, Bhide A, Bhople L, Guha A. Management strategies of cranial encephaloceles: A neurosurgical challenge. *Asian J Neurosurg*. 2019;14:718–24.
6. Ganeriwal V, et al. Giant meningoencephalocele with Arnold-Chiari type III malformation and anaesthetic challenges: A rare case report. *Saudi journal of anaesthesia vol*. 2019;13(2):136–9. doi:10.4103/sja.SJA_616_18.
7. Alwahab A, Kharsa A, Nugud A, Nugud S. Occipital Meningoencephalocele case report and review of current literature. *Chinese Neurosurgical Journal*. 2017;3(1).
8. Rehman L, Farooq G, Bukhari I. Neurosurgical interventions for occipital encephalocele. *Asian J Neurosurg*. 2018;13:233–7.
9. Naik V, Marulasiddappa V, Gowda Naveen MA, Pai SB, Bysani P, Amreesh SB. Giant Encephalocele: A Rare Case Report and Review of Literature. *Asian journal of neurosurgery*. 2019;14(1):289–91.
10. Antunes M, Pizzol D, Calgaro S, Di Gennaro F, Colangelo A. Case report Giant Encephalocele: successful management in limited-resource settings. *EuroMediterranean Biomedical Journal*. 2019;14:114–6. 10.3269/1970-5492.2019.14.26.
11. Bulut M, 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):316–9.
12. Agarwal A, Chandak AV, Kakani A, Reddy S. A giant occipital encephalocele. *APSP Journal of Case Reports*. 2010 Jul;1(2):16.
13. Kumar V, Kulwant SBhaikhel, Saurabh S, Chauhan R. Giant Occipital Meningoencephalocele in a Neonate: A Therapeutic Challenge. *Journal of Pediatric Neurosciences*. 2017;12:46. 10.4103/1817-1745.205655.
14. Murthy PS, Kalinayakanahalli Ramkrishnappa SK. Giant Occipital Encephalocele in an Infant: A Surgical Challenge. *Journal of Pediatric Neurosciences*. 2019 Oct-Dec;14(4):218–21.

15. Agrawal AC, Umamaheshwara RV, Hegde KV, Suneetha P, Kolikipudi DS. Giant high occipital encephalocele. *Romanian Neurosurgery*. 2016;30:122–6.
16. Kaif D, Ahmad D, Kumar Singh D, Pandey D. Giant Occipital Encephalocele – A Report of Two Rare Cases. *Journal of Medical Science clinical Research*. 2017;05(02):18162–5.
17. Dasgupta S, Saha S, Baranwal S, Banga M, Sandeep B. A rare case of giant occipital encephalocele with craniosynostosis. *Asian Journal of Medical Sciences*. 2016;7(4):98–100.
18. Mahapatra A. K: Giant Encephalocele: A Study of 14 Patients. *Pediatr Neurosurg*. 2011;47:406–11.
19. Mahajan CMD, DM*; Rath GP, MBBS, MD, DM*; Bithal PK, MD*; Mahapatra AK. MS, MCh† Perioperative Management of Children With Giant Encephalocele: A Clinical Report of 29 Cases. *Journal of Neurosurgical Anesthesiology*: July. 2017;Vol. 29(- Issue 3):322–9.
20. Ozdemir N, Ozdemir SA, Ozer EA. Management of the giant occipital encephaloceles in the neonates. *Early Hum Dev*. 2016 Dec;(103):229–234.
21. Kanesen D, et al. Giant Occipital Encephalocele with Chiari Malformation Type 3. *Journal of neurosciences in rural practice* vol. 2018;9(4):619–21. doi:10.4103/jnrp.jnrp_103_18.

Figures



Figure 1

A, Giant Occipital Encephalocele B, Giant Occipital Encephalocele (21 x 15 x 19 cm in size)

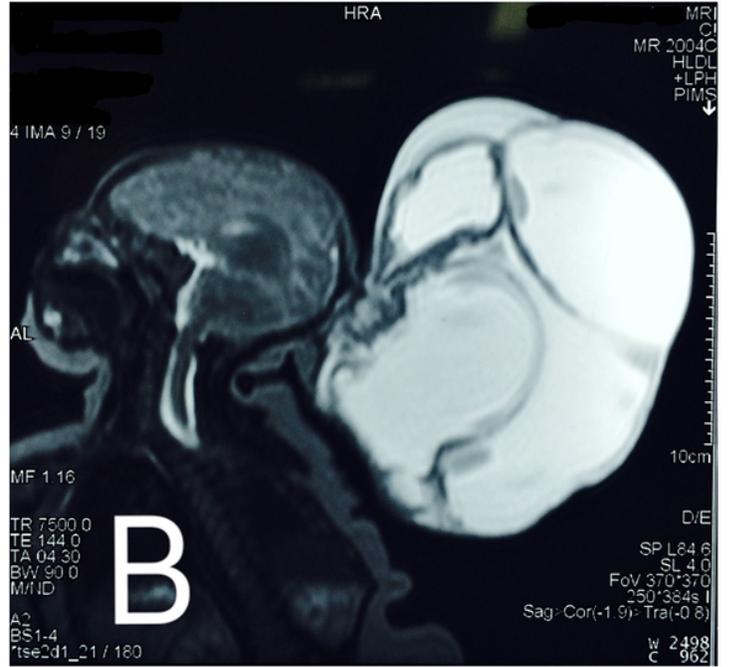
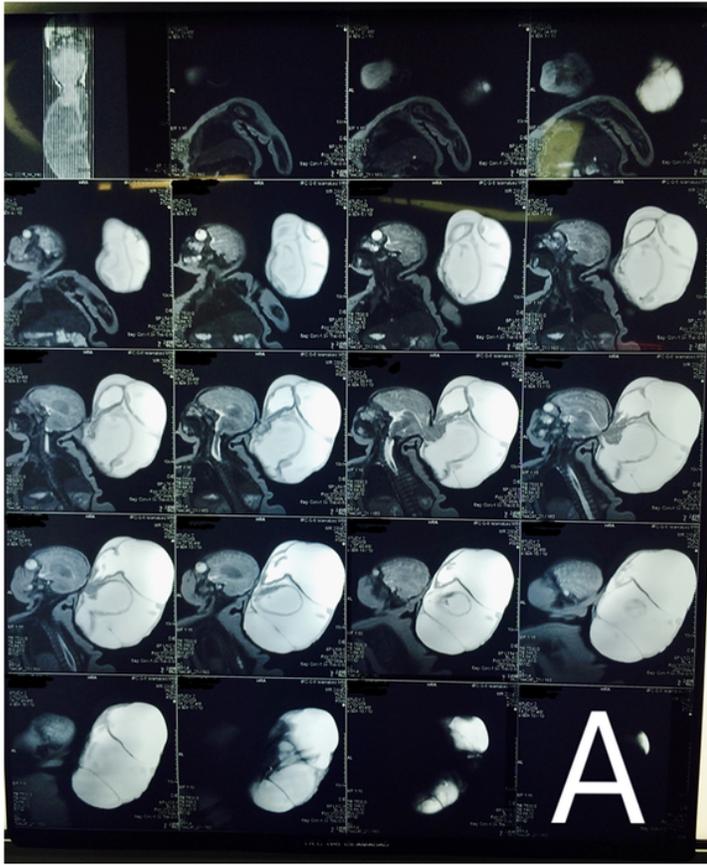


Figure 2

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