

Case Report

Giant Encephalocoele: A Rare Case Report and Review of Literature

Abstract

Giant encephalocoeles are rare entities with only one case series and few case reports reported in medical literature. Encephalocoeles, which reach a size larger than the head size, are called Giant encephalocoeles. We report a case of a 6 month old child who had giant encephalocoele with delayed motor milestones in the form of inability to hold neck. Anesthetic implications include difficulty in securing air way due without undue pressure on the sac. She underwent VP shunt followed by excision of the encephalocoele sac. Patient is doing well at 1 year of follow up. Preoperative neurological status and amount of brain tissue herniating into the sac are the most important factors determining the long term prognosis.

Keywords: Anesthetic implications, giant encephalocoele, prognosis in encephalocoele

Introduction

Giant encephalocoele are rare entity and pose unique challenges to Neurosurgeons, Anesthetists, and Neuroradiologists. There is only one case series and few case reports about this in literature.^[1] We report to you a rare case of giant encephalocoele with delayed motor milestones which gradually improved following surgery.

Case Report

We report to you a 6-month-old child who presented with the swelling in the occipital region from birth which gradually increased in size [Figure 1]. It was a home delivery with no antenatal check-up in rural eastern India. It was a of 26 cm × 34 cm × 30 cm soft translucent, containing fluid. No e/o cerebrospinal fluid (CSF) leak. On examination, she had delayed motor milestones in the form of inability to hold her head. Breathing, feeding, lower spine were unremarkable. Rest of the neurological examination was unremarkable. She was weighing 4.5 kg before surgery. Magnetic resonance imaging (MRI) revealed a large encephalocoele with small amount of neural tissue herniating into the sac with gross hydrocephalus and cervicothoracic syrinx [Figure 2]. Anesthetic management of children with giant encephalocoels present challenges with regard to patient positioning, airway management,

temperature monitoring, and estimating blood and fluid loss.^[2] In our case, the encephalocoele was so big that it was not possible to intubate the child with the head supine as the giant encephalocoele limited head extension severely and also there was the risk of rupturing the sac with sudden uncontrolled third space volume loss. We were able to intubate the child by placing the child's head beyond the edge of the operating table and supported by an assistant [Figure 3]. On the operating table, encephalocoele was first drained with replacement of fluids to prevent volume loss. We did ventriculoperitoneal shunt followed by opening of the encephalocoele, excision of the redundant neural tissue, and primary closure of the dura. Skin was reconstructed. One year follow-up, she is able to hold her head, sit without support and mental milestones are grossly normal for her age.

Discussion

Giant encephalocoeles are rare phenomenon with only few case reports being reported.^[1] Only one case series has been reported until the date of 14 cases.^[1] They are known by different names such as giant massive or large encephalocoeles.^[1-4] Authors feel that they should be called real giant when the encephalocoele size reaches head size. Most of the children are neonates with chief complaints being enlarging swelling with difficulty to feed.^[1,4,5] They may have

**Vikas Naik,
Vinay
Marulasiddappa¹,
Mandya Appaji
Gowda Naveen,
S. Balaji Pai,
Pratham Bysani,
S B Amreesh**

Departments of Neurosurgery
and ¹Anesthesiology, Bangalore
Medical College and Research
Institute, PMSSY Super
Speciality Hospital, Bengaluru,
Karnataka, India

Address for correspondence:

Dr. Pratham Bysani,
Department of Neurosurgery,
1st Floor, 141, PMSSY Super
Speciality Hospital, Victoria
Hospital, K. R. Road,
Bengaluru - 560 002,
Karnataka, India.
E-mail: prathambyrani@gmail.
com

Access this article online

Website: www.asianjns.org

DOI: 10.4103/ajns.AJNS_87_18

Quick Response Code:



How to cite this article: Naik V, Marulasiddappa V, Gowda Naveen MA, Pai SB, Bysani P, Amreesh SB. Giant encephalocoele: A rare case report and review of literature. Asian J Neurosurg 2019;14:289-91.

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com



Figure 1: Large occipital encephalocele



Figure 2: Magnetic resonance imaging showing large encephalocele with small amount of cerebellar tissue herniating with gross hydrocephalus and cervicothoracic syrinx

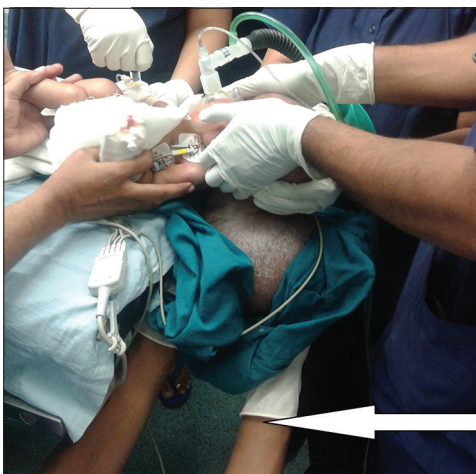


Figure 3: Assistant supporting the patient's head with the giant encephalocele as the head is positioned beyond the edge of the table for intubation

microcephaly, craniosynostosis, cleft lip or associated brain anomalies such as microgyria, polygyria, chiari

malformation, and hydrocephalus.^[1-6] MRI with magnetic resonance venography is the investigation of choice, with computed tomography to look for bony defects.^[1] One has to carefully examine the contents of the sac for any torcula or veins herniation. Most of the contents are atretic and can be excised however torcula and remaining viable tissue should be placed back weighing the risk of coming.^[6-9] Dura should be repaired with pericranium or artificial dura. Large bone defects to be covered with methylmethacrylate. Associated microcephaly craniosynostosis, hydrocephalus, chiari malformation, syrinx may require treatment.^[1,6-8] Postoperatively, one has to look for hypothermia, raised intracranial pressure, apnea, cardiac arrest, CSF leak, and infection.^[6-9] In our case, maybe child had not attained head holding due to the weight of the sac. She is now head holding with mild ataxic gait. Rest of the examination is unremarkable. Patient with a large amount of cerebrum, cerebellum and brain stem herniating into sac have a poor prognosis. Irrespective of the sac size patients with less amount of brain tissue in the sac and good preoperative neurological condition carry a good prognosis.^[7]

Conclusion

Giant encephaloceles are rare but challenging entities requiring multidisciplinary approach. Patients with less amount of brain tissue in the sac, no severe neurological deficits carry a good prognosis

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Mahapatra AK. Giant encephalocele: A study of 14 patients. *Pediatr Neurosurg* 2011;47:406-11.
2. Cevik B, Orskiran A, Yilmaz M, Ekti Y. Anesthetic management of a newborn with giant occipital meningoencephalocele: Case report. *Int J Case Rep Images* 2012;3:10-2.
3. Bozinov O, Tirakotai W, Sure U, Bertalanffy H. Surgical closure and reconstruction of a large occipital encephalocele without parenchymal excision. *Childs Nerv Syst* 2005;21:144-7.
4. Shokunbi T, Adeloye A, Olumide A. Occipital encephaloceles in 57 Nigerian children: A retrospective analysis. *Childs Nerv Syst* 1990;6:99-102.
5. Agrawal D, Mahapatra AK. Giant occipital encephalocele with microcephaly and micrognathia. *Pediatr Neurosurg* 2004;40:205-6.

6. Chapman PH, Swearingen B, Caviness VS. Subtorcular occipital encephaloceles. Anatomical considerations relevant to operative management. J Neurosurg 1989;71:375-81.
7. Agarwal A, Chandak AV, Kakani A, Reddy S. A giant occipital encephalocele. APSP J Case Rep 2010;1:16.
8. Mahapatra AK, Gupta PK, Dev EJ. Posterior fontanelle giant encephalocele. Pediatr Neurosurg 2002;36:40-3.
9. Mohanty A, Biswas A, Reddy M, Kolluri S. Expansile cranioplasty for massive occipital encephalocele. Childs Nerv Syst 2006;22:1170-6.