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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 be 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 encephalocele 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 encephalocele, prognosis in encephalocele*


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## Introduction

Giant encephalocoele are rare entity and pose unique challenges to Neurosugeons, 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 encephalocele 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 encepholocoeles.[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



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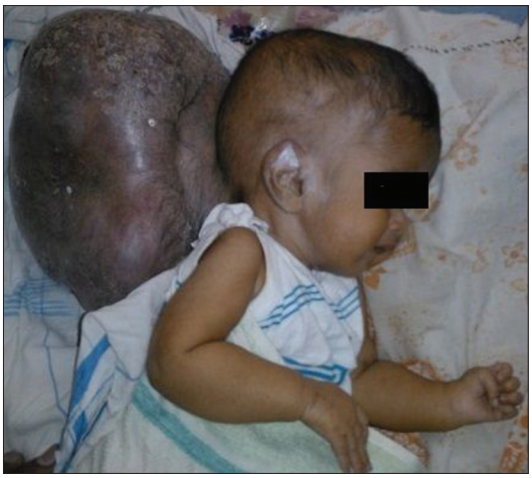

**Figure 1: Large occipital encephalocoele**

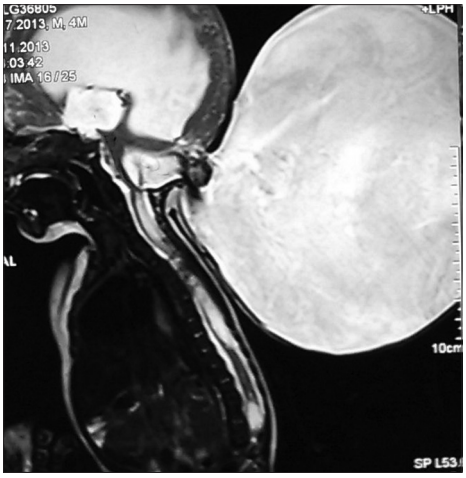

**Figure 2: Magnetic resonance imaging showing large encephalocoele with small amount of cerebellar tissue herniating with gross hydrocephalus and cervicothoracic syrinx**

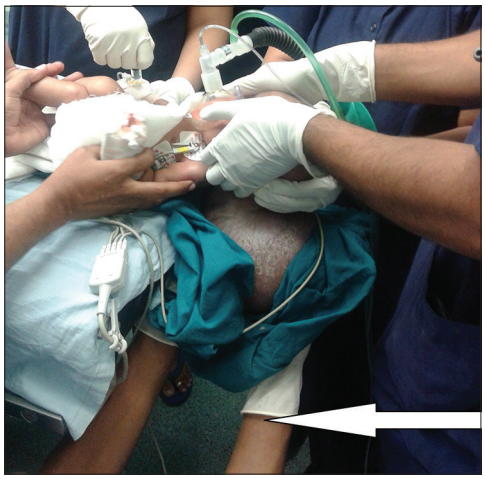

**Figure 3: Assistant supporting the patient's head with the giant encephalocoele as the head is positioned beyond the edge of the table for intubation**

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.

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### Conflicts of interest

There are no conflicts of interest.

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