Giant Encephalocele: A Rare Case Report and Review of Literature

Abstract
Giant encephaloceles are rare entities with only one case series and few case reports reported in medical literature. Encephaloceles, which reach a size larger than the head size, are called Giant encephaloceles. We report a case of a 6 month old child who had giant encephalocele with delayed motor milestones in the form of inability to hold neck. Anesthetic implications include difficulty in securing airway 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

Introduction
Giant encephalocele 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 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 encephalocele with small amount of neural tissue herniating into the sac with gross hydrocephalus and cervicothoracic syrinx [Figure 2]. Anesthetic management of children with giant encephaloceles present challenges with regard to patient positioning, airway management, temperature monitoring, and estimating blood and fluid loss.[2] In our case, the encephalocele was so big that it was not possible to intubate the child with the head supine as the giant encephalocele 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, encephalocele was first drained with replacement of fluids to prevent volume loss. We did ventriculoperitoneal shunt followed by opening of the encephalocele, 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 encephaloceles 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 encephaloceles.[1-4] Authors feel that they should be called real giant when the encephalocele 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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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.\cite{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.

**References**


