Giant Occipital Meningoencephalocele: Anaesthetic Implications
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Abstract
Encephalocele is a broad term representing herniation of cranial contents through a congenital defect in the cranium. If only cerebrospinal fluid (CSF) and meninges herniate, it is termed as a meningocele. A meningoencephalocele is herniation of both neural elements and meninges. Anaesthetic challenges in management of occipital meningoencephalocele include securing the airway with intubation in lateral position, intraoperative prone position and its associated complications, careful securing of the endotracheal tube and accurate assessment of blood loss. We report a case of a giant occipital meningoencephalocele and discuss its anaesthetic implications.

CASE REPORT
A 3 day-old female neonate presented with a large cystic swelling at the back of the neck and was posted for surgical excision of the swelling. Baby was delivered by cesarian section and had cried 2 min after birth. At birth APGAR score was 11 and weight 3.5 kg. The swelling measured 20 x 16 cm arising from posterior part of the head. (Fig.1)

Figure 1
The neonate was active and there was no other discernible congenital anomaly.

There was no neurological deficit. Magnetic resonance imaging (MRI) brain showed giant occipital meningoencephalocele with occipital lobe and part of brainstem herniating in the swelling. (Fig.2)

Figure 2
There was no other associated congenital abnormality and cardiac and respiratory system examination was within normal limits. The associated neurological abnormality was Chiari Type II malformation.

After attaching cardio scope and pulse oxymeter for monitoring purpose the neonate was induced with sevoflurane in lateral position. After confirming adequate
mask ventilation, muscle relaxation was achieved with succinylcoline 2 mg/kg and the neonate was intubated by senior anesthesiologist in lateral position only. Confirming endotracheal placement of tube and after proper tube fixation, the baby was put into prone position with extreme care taking to prevent the accidental extubation of the tube. Anaesthesia was maintained with N\textsubscript{2}O:O\textsubscript{2}:50:50, sevoflurane 1-2\%, atracurium infusion 0.5 mg/kg/hr and fentanyl infusion 2 mcg/kg/hr. Surgical resection included much of the occipital lobe and part of the brainstem along with the swelling. Total blood loss of 100 cc was adequately replaced with 100 cc of blood. Postoperatively, after assessing the brain matter resected, we decided to put the patient electively on a ventilator after adequately warming the neonate to desired set temperature. However due to extensive and unavoidable resection of much of the occipital lobe and brain stem neonate could not make it up and expired on 5\textsuperscript{th} postoperative day.

**DISCUSSION**

Meningoencephalocele is hernial protrusion of part of meninges and neural elements in a sac.\textsuperscript{1} Reported incidence is 1 in 5000 live births.\textsuperscript{2} Children with meningoencephalocele, are likely to have varying degrees of sensory and motor deficits. Associated congenital defects\textsuperscript{2} includes club foot, hydrocephalous, extrophy of bladder, prolapsed uterus, Klippel-Feil syndrome and congenital cardiac defects.

Major anaesthetic challenge,\textsuperscript{2,4,5} in management of occipital meningoencephalocele is securing the airway. Awake tracheal intubation\textsuperscript{2} in lateral position may be performed in these patients to avoid pressure on the sac. Anaesthesia may also be induced in supine position with sac protected by elevating it on a doughnut-shaped support. Although long acting non-depolarizing muscle relaxants may be used to facilitate tracheal intubation, these are usually avoided after consulting with surgeon, if he has to use a nerve stimulator to identify functional neural elements.

We mask ventilated the child in lateral position. (Fig.3) Finding no difficulty in mask ventilation, succinylcoline was given and child was intubated by senior anesthesiologist in lateral position only. As nerve stimulation was not sought by the surgeon, we maintained relaxation with atracurium throughout the surgery.

Although meningoencephalocele can be associated with both upper and lower motor neuron dysfunction, succinylcoline does not elicit a hyperkalemic response.\textsuperscript{6} These neonates however may have an abnormal ventilatory response to hypoxia and hypercarbia.

Children with meningoencephalocele have an increased incidence of latex allergy,\textsuperscript{7} which can manifest as intraoperative cardiovascular collapse and bronchospasm.

Attention has to be given to blood loss, maintenance of body temperature, prone position (Fig.4) and its associated complications and careful securing of the endotracheal tube.

**Figure 3**

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**Figure 4**
The persistence of fetal circulation increases the blood loss in posterior fossa surgery. So intensive monitoring is required to estimate the blood loss and replace it adequately.

The neurological prognosis in such children depends on the amount of neural tissue that has herniated through the sac. The neural tissue is often dysplastic and gliotic but the presence of microcephaly with a large posterior encephalocele containing significant brain tissue is a predictor of poor neurological outcome. The decision regarding surgery is dependent on various factors including the amount of neural tissue in the sac, other congenital anomalies, etc. The decision must involve the family and other medical personnel.

In our case due to extensive herniation of occipital lobe and some part of brain stem in the sac, prognosis had already been sited as very poor by the neurosurgeon. However, even after explaining every thing to the parents regarding postoperative mortality in such cases, they had given consent to get the child operated as last hope.

CONCLUSION
Managing a case of meningoencephalocele includes looking for other congenital abnormalities, expertise in handling airway, and intraoperative care mainly involving proper positioning and blood loss replacement.

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