Airway Management Of A Large Giant Occipital Encephalocele In A Neonate - An Anaesthetic Challenge

Dr. Chavi Sethi, Dr. Roopesh Kumar, Dr Shruti Gaur

1Assistant Prof, Dept of Anaesthesia, MLB medical college Jhansi.
2Prof and Head, Department of Anaesthesia, MLB medical college Jhansi.
3Junior Resident, Department of Anaesthesia, MLB medical college Jhansi.

Background - Encephalocele is a term which represents herniation of meninges and cerebrospinal fluid through the congenital defect in the cranium. Anaesthetic challenges in the management of neonates with encephalocele include airway management and proper positioning of the neonate without pressure on the meningocoele sac so as to prevent the rupture of the membranes. Associated congenital anomalies also can cause anaesthesia and procedure related complications. Other difficulties include performing a difficult airway case in an unfamiliar environment outside operation theatre. We report a case of 6 day old neonate with occipital meningocoele posted for MRI brain and the successful anaesthetic management.
INTRODUCTION:
The term Encephalocele refers to a defect in the skull and the dura with extracranial extension of intracrani al structures. In southeast Asia the incidence is approximately 1 in 5000 live births. Occipital encephaloceles can vary from a small pedunculated swelling with a narrow neck to an extremely large swelling. In one study up to 16% of the occipital encephaloceles were more than 20 cm in diameter [1]. In giant occipital encephaloceles the size of the swelling is larger than the size of the head from which they arise, and because of their enormous size these pose a surgical challenge and anaesthetic challenge [2]. We report this case to highlight the difficulties in the management of giant occipital encephaloceles.

CASE REPORT
A two months female baby presented with progressively increasing swelling over the occipital region since birth. The baby was born at 37 weeks of gestation by normal delivery with a birth weight of 2500 gms. There was no abnormality on physical examination except for a large cystic mass in the occipital region. It was larger than the size of the head (Fig. 1). The skin over the swelling was stretched but well formed. The anterior fontanelle was closed. Baby was able to track objects and light and pupils were reactive. Routine hematological and biochemical investigations were reported as normal. A MRI report SE T1 AND FSE T2 weighted axial and sagittal images .T1 and T2 of brain demonstrated the encephalocele with evidence of herniation of very thin looking redundant brain tissue into the sac (Fig.3,4). MRI images also revealed a significant defect of the occipital bone.
Figure 1 and 2: Clinical photograph showing giant occipital encephalocele and translumination present in swelling.

Figure 3: MRI brain showing large encephalocele sac with protrusion of the contents.

Anesthetic management-

The baby was kept nil per oral for 4 hours on the day of surgery. We planned intubation in lateral position as the encephalocele was bigger than the patient’s head, positioning of the head was anticipated to be difficult. The child’s head was positioned maquet pillow in lateral position. After attaching the standard monitors (ECG, PULSE OXYMETRY, NIBP), child was induced with sevoflurane and 100 oxygen. Glycopyrolate (0.04mg / kg), Ondansetron (0.1 mg /kg), fentanyl (2 microgram/kg), propofol (2mg /kg), succynyl choline (2mg / kg) were given. And the mask ventilation was considered adequate.

Laryngoscopy was attempted with MAC 1 BLADE. The attempt of laryngoscopy was failed[3]. The repeat attempt of laryngoscopy with MILLER blade in right lateral position also failed. Both the attempts of laryngoscopy were very quick so there was no desaturation noted. Since the prior attempts of laryngoscopy in lateral position failed, the intubation was attempted in supine position with the head supported on the table and the encephalocele hanging at the edge. The neck was supported by one assistant, laryngoscopy was done and 4mm of ETT was introduced.
Surgery was planned to reduce the size of the swelling as well as its contents. During surgery the baby was positioned in lateral position. A circumferential incision was placed over the sac, and the neck was dissected out. Sac was then opened. The herniated brain tissue looked redundant. However, near to opening in the skull there was evidence of normal looking arterioles and veins on the surface of the occipital lobe with normal sulci and gyri pattern. The sac was reduced in size, sufficient enough to accommodate the healthy looking brain tissue. The skin was closed with interrupted sutures (Fig. 4). There was evidence of persistent cystic collection in the occipital region with mild dilatation of ventricles. Child remained well in follow up and monitored for fluid collection and the requirement for a shunt.

**DISCUSSION**

Encephaloceles account for 10 to 20% of all craniospinal dysraphisms and 70% of occipital encephaloceles occur in females [4]. These lesions are usually covered either with normal skin, dysplastic skin or a thin, distorted meningeal membrane. The large sized swellings may have significant brain herniation, abnormality of the underlying brain, microcephaly and ventriculomegaly. Such patients usually have poor prognosis. Encephaloceles with a small amount of dysfunctional tissue are conventionally treated by excision of the herniated brain tissue and repair of the dural defect. The surgical management of children with large defect along with herniation of a considerable proportion of brain matter into the sac, at times can be extremely difficult. Large encephalocele distort the normal anatomy, making airway management difficult [5]. It places the neck in extreme flexion and obscure the sight of laryngoscopy. So elevating the body and shoulders by towels is necessary in order to facilitate laryngoscopy.

There may be considerable blood loss during repair which leads to hypotension not responding to blood and fluid replacement. Sudden loss of CSF from sac may be one of the reason for hypotension [6].

Hypothermia is one of the frequent complications, which corresponds to surface area of area exposed, age of the patient and use of cold solutions during surgery and these hypothermic neonates are prone to develop bradycardia, apnea, hypotension, acidosis, prolonged recovery from neuromuscular block, impairs platelet functions lead to more incidence of wound infections. It can prevented by warmers in OT, use of warm fluids covering the child with cotton and drapes.

Cushing response that is intracranial hypertension and bradycardia is a complication which can be prevented by avoiding hypoxia and hypercapnia, as it causes cerebral vasodilation and may worsen raised intracranial pressure especially in combination with laryngoscopy and airway manipulation. Patients with giant encephalocele and large amount of brain tissue in the sac usually die either shortly after birth or as a result of operation. In such patients, it is generally impossible to foretell whether the infant will die quickly or will continue to live for many months or years, as size of the encephalocele itself is not a guide to prognosis. Ultimate result depends on the amount of normal brain tissue left inside the skull after the operation [7]. Surgery thus just facilitates nursing of the baby.

In contrast to the previous case reports where the neonate had poorer prognosis (because of larger
lesions and significant brain tissue within the sac) this infant was neurologically well developed. Furthermore less functional tissue in the sac made the surgical excision of the sac easy and safe.

REFERENCES