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KEYWORDS: - encephalocele, myelomeningocele, obstructive hydrocephalus.

CASE REPORT: OCCIPITAL ENCEPHALOCELE WITH MYELOMENINGOCELE WITH OBSTRUCTIVE HYDROCEPHALUS: ANAESTHETIC CHALLENGES

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ABSTRACT

A 2yr old female admitted to neurosurgery with chief complain of swelling on posterior neck and posterior scalp since birth, which is progressively increasing in size of as compare to head. According to the patient mother, patient had swelling in back of scalp since birth, which was of small size about lemon at birth, which increased progressively to size bigger than head.

INTRODUCTION

Encephaloceles are classified as neural tube defects. The neural tube is a narrow channel in the developing fetus that allows the brain and spinal cord to develop. The neural tube folds and closes early during pregnancy (third or fourth week) to complete the formation of the brain and spinal cord. A neural tube defect occurs when the neural tube does not close completely, which can occur anywhere along the head, neck or spine. The lack of proper closing of the neural tube can lead to a herniation process which appears as a pedunculated (having a stalk-like base) or sessile (attached directly to its base without a stalk) cystic lesion protruding through a defect in the cranial vault referred as encephalocele. They may contain herniated meninges and brain tissue (encephalocele or meningoencephalocele) or only meninges (cranial meningocele). Encephaloceles containing tissue from the brain and spinal cord are called encephalo-myeloceles.

In the Posterior hemisphere, occipital encephaloceles constitute 80 to 90% of all encephaloceles. Classically, patient with encephaloceles are born with the swelling at birth. The size and content of the encephalocele are variable. In occipital encephalocele, a globular swelling is noticed over the occipital bone in the midline. The size of the encephalocele is hardly ever indicate of its content. Most of the encephaloceles are brilliantly trans illuminant on examination. However, when a large amount of gliosed brain tissue is present inside the sac, there may be variability in the degree of transillumination.

Usually, the head size is small. The larger the brain herniation, the smaller is the head size. Sometimes, the encephalocele may be very large and is called giant encephalocele.

We report to you a case of Occipital encephalocele with myelomeningocele with obstructive hydrocephalus posted for VP shunt.

Case Report:

A 2yr old female admitted to neurosurgery with chief complain of swelling on posterior neck and posterior scalp since birth, which is progressively increasing in size of as compare to head. According to the patient mother, patient had swelling in back of scalp since birth, which was of small size about lemon at birth, which increased progressively to size bigger than head.

Patient also has history of increase in head size as compared to age. Patient birth via normal vaginal delivery and no history of instrumentation during delivery.
O/E:

- Spontaneous eye opening
- Crying on examination
- Spontaneous movement of upper limb with weakness in both lower limbs.
- Obvious swelling at back of scalp and neck of about size 10x15 cms. with no any CSF leak.
- she had delayed motor milestones in the form of inability to hold her head.
- Breathing, feeding, and lower spine were unremarkable.
- Rest of the neurological examination was unremarkable.

She was weighing 4.5 kg before surgery. Head circumference 82 and the length was 102 cms. Child has anticipated difficult airway due to large due to large head, small mouth and no neck movement. Pre-operative lab investigations were within normal limits. Magnetic resonance imaging (MRI) revealed a large occipital encephalocele with obstructed hydrocephalous.

Anaesthetic management of children with giant encephaloceles present challenges with regard to patient positioning, airway management, temperature monitoring, and estimating blood and fluid loss. In our case, patient NBM,HRC confirmed, peripheral iv line with 24G iv canula secured, Standard monitors(PR, SPO2, NIBP) attached, iv fluid inj. RL(500ml)attached.

Pt. premedicated with inj. Midazolam 0.5mg, inj. Glycopyrrolate 0.008mg.

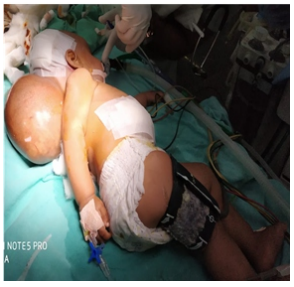

Figure1: Post-operative position of encephalocele under general anaesthesia before extubating.

The anaesthetic drugs according to the expected weight for a 02 yr. old (approximately 5 kg) were prepared. In order to optimize the patient's position during laryngoscopy and intubation, a large ring was placed below the patient's head. Child's body was laterally aligned to the head by using multiple folded OT sheets below it and a difficult airway cart was kept ready. Sevoflurane was used for induction with 100% oxygen. After confirmation of mask ventilation, fentanyl, thiopentone and scoline (dose according to weight) was administered. 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. Direct laryngoscopy was performed with No.0 Macintosh laryngoscope blade. We were able to intubate the child by placing the child's head laterally supported by an assistant with uncuffed endotracheal tube (ID 4.0) and tube was fixed after confirming adequate bilateral equal air entry.. Maintenance of anaesthesia was done with sevoflurane and intermittent doses of atracurium. Additional analgesia was achieved with paracetamol suppository. The ringer lactate was transfused for replacement fluid. Ventriculoperitoneal shunt was done to relieve intra cranial pressure. At the end of the surgery, neuromuscular blockade was reversed with neostigmine and glycopyrrolate. Child was extubated after she became fully awake and had spontaneous regular respiration.

Figure 2: Patient with occipital meningocoele and VP Shunt insitu. (original)

Postoperative risk of subdural effusion owing to over drainage of CSF was ruled out after obtaining a normal CT scan of head at day 3 and was discharged on day 5 of admission.

DISCUSSION

Cranial meningocele refers to the herniation of meninges through a bony defect in the skull and meningomyelocele contain neural elements along with the meninges in the herniated sac.[1] An encephalocele contains elements of brain tissue also within the sac. The defect is more often in the occipital bone.[2] Neonates with encephalocele can have associated sensory and motor deficits, hydrocephalus, and other associated congenital anomalies.

Airway management of neonates and pediatric population is difficult compared with adult due to anatomic variations. Short neck, large tongue and large head are some of the reasons why pediatric airway is difficult. Difficult airway cart with ventilation masks, tracheal tubes, airways, scope blades etc., should be kept ready. Devices for maintaining normothermia like circulating warm-air devices, airway humidifiers, fluid warming devices should be kept ready.[3]

Neonates with meningocele may have restricted neck extension and difficult airway, making intubation difficult. One more concern is positioning of the neonate for intubation and subsequent positioning during the procedure. Complications most commonly encountered during anesthetic management of meningomyelocele include bradycardia, bronchospasm, endobroncial intubation, hypoxemia and laryngospasm.[4]

- The perioperative anesthetic plan includes airway management, meticulous fluid administration, management of hypothermia and proper positioning of the baby. Care should be taken to avoid pressure on the sac. Intubation can be attempted in one of the ways described below:

- Lateral position[5]
- In the supine position after getting the child's head beyond the edge of the table with an assistant supporting the head[6]
- The baby can be placed on a pile of blankets and the sac protected in a dough-nut shaped support or held by an assistant.

Only after checking for adequate mask ventilation, NMB agents should be administered. Endotracheal tube should be carefully secured and taped.

Extubation in a difficult airway scenario, as in our case, should be carried out once the criteria are met with intact cough and gag reflex and the child is fully awake and breathing well. Causes for delayed recovery include hypothermia, inadequate reversal, prolonged effect of muscle relaxants and malnutrition.

Although CSF shunts are the standard means for treatment of hydrocephalus, they are prone to complications with 16% of shunts undergo revision within one month of insertion. The operative procedures involving shunt insertion may have a mortality rate of 31%. There is also a difficulty in the assessment of fluid deficits and drug doses which was managed by titration based on lean body weight. It is important to maintain of operating room temperature at 28 C along with warm IV fluids and warming blankets as increased surface area of head may lead to hypothermia. The surgical positioning may also be complicated.

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