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ANAESTHETIC MANAGEMENT OF A HUGE OCCIPITAL ENCEPHALOCELE IN A NEONATE

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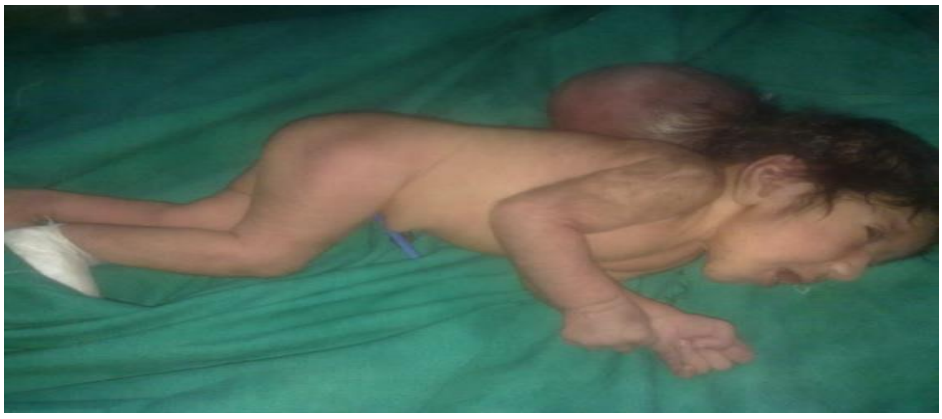
I. INTRODUCTION

Anaesthesia to newborn poses many challenges to the anaesthesiologist and that for occipital encephalocele adds to it. It includes securing a difficult airway, prone position and blood loss. We describe a successful management of huge occipital encephalocele for excision.

II. CASE HISTORY

7 day old female baby of 3 kg presented with large cystic swelling in occipital region for surgical excision. Baby was born at 37 weeks of gestation by elective caesarean section for breech presentation and presence of large cystic swelling on back of head. No +ve medical history was present in mother. She consumed folic acid tablets from 6 weeks of conception. No h\o teratogenic drug consumption. Diagnosis of large cystic swelling suggestive of encephalocele was done in USG scan of 1st trimester and condition being surgically treatable, pregnancy was continued. Baby cried immediately after birth . Examination was normal. Head circumference was 34 cm. A huge pedunculated fluctuant cystic swelling measuring approximately 12×10×9 cm arising from occipital region which was intact and covered with skin. Baby was moving all 4 limbs normally and no sensory deficit. The neck was very short, webbed with low hair line. No other congenital anomaly was obvious. All routine investigations were normal .CT brain showed 10.6 cm × 9.2 cm × 8.2 cm well defined extracranial lesion with neural tissue within confirming the diagnosis of occipital encephalocele connected to cistena magna through bony defect of size 6mm. Herniation of vermis through foramina magna and high arched tentorial leaflets suggestive of Chiari malformation II. Written informed consent with high risk explained to surgeons and relatives was obtained. Difficult intubation anticipated and LMA, jet ventilation, cricothyroidotomy cannula and tracheostomy was kept ready.



I.V line was secured with 24 G catheter and inj atropine 0.1 mg was given. Precordial stethoscope attached and child made supine by placing the baby on the platform, made by keeping a pillow and flat OT table cusion obove the pillow

.Simultaneously the sac was carefully placed in doughnut shaped support created with sterile towel cloth matching the height of encephalocele sac. So now the baby is placed like normal supine position. Child is induced with sevoflurane 3%-4% and intubated with macintosh blade no. 00 with portex no. 3 uncuffed tube in neutral position of head by senior ansaesthesiologist with Cormack Lehane grade I. Endotracheal tube position confirmed. Throat was packed for additional stability of tube in prone position. After proper fixation of tube baby made prone with extreme care to prevent accidental extubation and rupture of huge neural placode .Pressure on abdomen was avoided and pressure points including eyes were protected. Local infiltration was given at base of pedunculated swelling by surgeon with 3 ml of 1% lignocaine. Anaesthesia was maintained with N2O : O2 50:50, sevoflurane 0.6- 1%, fentanyl 1µg/ kg and IPPV with Jackson n Ree's circuit. Muscle relaxation was achieved with inj Atracurium 1mg i.v. Dystrophic meninges were exercised after draining the sac .Procedure lasted for 40 min .I.V. Isolyte P given@ 4ml/kg/hr. Additional 15 ml given to replace blood loss which was approximately 5-6ml . Body temperature was maintained by covering rest of the body with warm blanket, infusion of warm i.v. fluid n keeping the OT temp between 23-25 degree. Neuromuscular blockade was reversed with neostigmine and atropine and baby extubated in supine position after removing throat pack fully awake. Baby was moving all 4 limbs normally, analgesia was achieved with inj Tramadol 1mg/kg i.v. 8 hrly. Postop period was uneventful with no neurological deficit n baby was discharged on 8th postop day.

III. DISCUSSION

Encephalocele is hernial protusion of neural elements in sac. Meningocele is protrusion of only meninges. Incidence is 1 in 5000 [1] and common in lumbosacral region. Early excision is recommended to prevent infection and rupture. Anaesthesiologist encounters many challenges during excision of meningocele or encephalocele in occipital region including difficult airway management, prone positioning, protection of neural placode, assessment of volume status and prevention of hypothermia.[3,4]

Common congenital anomalies associated may be club foot, hydrocephalus with Chiari's malformation, extrophy of bladder, prolapsed uterus, Klippel feil syndrome and cardiac defects. [2]

Anaesthesia may induced with intravenous or inhalational method. Intubation can be done awake in lateral position or supine position by placing the neural placode in doughnut shaped support.

Other method is placing the head at the edge of the table, supported by one person and elevating the body off the table while supporting the pelvis by other .[7] Another method was described by Mowaffi is placing the baby supine on platform made by silicon supports kept one above other till the height matches with encephalocele sac and head was supported in hollow cushion protecting sac.[5] We used similar technique. We made the baby supine on flat platform made by a pillow and flat foam cushion of OT table placing above the pillow. So that height of the platform matched height of encephalocele sac.

We also protected neural placode in doughnut shaped support prepared with sterile towel cloth, height of which was also matching height of sac. So baby was now supine like routine supine position. This method is more useful than others as single person can manage the airway, lateral position not required and pressure on sac is avoided. We intubated the child

in neutral position of head to avoid injury to cervical spine as child was having very short webbed neck with low hair line suggestive of Klippel feil syndrome. But the diagnosis was not confirmed by MRI preoperatively.
Klippel feil syndrome was independently described by Maunice Kippel and Adre Feil in 1992. They described patients who had a short webbed neck, decreased range of motion and low hair line.

Nagib et al described 3 types.[6]

Type I – 2 sets of fused vertebrae with open intervening sac that can sublux gradually or with acute trauma.
Type II – involves craniocervical anomalies leading to increased mobility at craniocervical level. It is associated with Arnold Chiari malformation.
Type III – fusion of one or more levels with associated spinal stenosis.

Common association of encephalocele with type II Chiari malformation is probably because of CSF loss in sac resulting in inadequate stimulus for development of structures in posterior fossa leading to upward herniation of vermis and downward herniation of brainstem. Therefore signs of brainstem dysfunction may be associated like poor feeding or swallowing with weak gag, lack of pharyngeal coordination, stridor due to vocal cord abnormalities, apnea or abnormal breathing pattern due to structural derangement in pontomeduallary respiratory centre, abnormal response to hypoxia n hypercarbia may be associated. Non depolarizing muscle relaxant atracurium was used after consulting the surgeon. It may need to be avoided if the surgeon plans to use nerve stimulator to identify functional neural elements. Though encephalocele can be associated with both upper n lower nerve dysfunction suxamethonium does not elicit hyperkalemia. [8] We avoided use of suxamethonium. These children exhibit sensitivity to latex and may manifest intraoperative cardiovascular collapse and bronchospasm. We performed latex agglutination test preoperatively was negative.
Meticulous attention has to be given while making the baby prone, maintenance of body temperature, volume replacement.

Due to risk of apnea and abnormal breathing pattern postop O2 supplementation in hood is recommended.

Prognosis depends of amount of neural tissue herniated in sac which is excised. Prognosis needs to be discussed with family.

IV. CONCLUSION

Anaesthetic management of occipital encephalocele is challenging due to presence of difficult airway, prone positioning for surgery, associated with neonatal anatomy, physiology and pharmacology. Vigilant evaluation with careful management results in safe and successful results.

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