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Anaesthetic Management of a Newborn with Giant Occipitalmenigocele, Cleft Lip & Palate and Cardiac Abnormality

Monal Ramani[1], Deepshikha[2], Prachi Agnihotri[3]

[1]Associate Professor

[2]Third Year Resident Doctor, MD Anaesthesia

[3]Third Year Resident Doctor, MD Anaesthesia

Abstract: ***Introduction: Meningocele is herniation of csf and meninges through a congenital defect in the skull or vertebral column. Anesthetic management is challenging due to difficult airway, prone position ,associated congenital abnormality, hypothermia and fluid loss. Case Report: Neonateinduced with sevoflurane in lateral position and for intubation lifted from table, swelling and pelvis supported by other anesthesiologist. Prone position given with care. Total 90 ml PCV and 150 ml crystalloid given. Warmer placed to prevent hypothermia .Conclusion: Vigilant evaluation with careful management result in safe and successful result.***

Keywords: menigocele, cleft lip and palate, DTGA

1. Introduction

Meningocele is a cystic swelling filled with cerebrospinal fluid but not contain neural tissue.It is midline sac like protrusion of either cerebral or spinal meninges through a congenital defect in the skull or vertebral column. Here, we present a case of newborn with giant occipital meningocele with cleft lip and palate and cardiac abnormalities.

2. Case Report

A 2 day old female neonate weighing 3kg born by caesarian section in civil hospital,ahmedabad presented with giant occipital meningocele of approx 15×7×16 cm.The baby also had cleft lip and palate and 2D echo suggestive of dextro transposition of great arteries(DTGA) with large ASD, maintaining saturation of 90% on oxygen hood. On air saturation was 60-65%. The baby was referred to pediatric surgery department for further management.

Figure 1: Newborn with huge occipital swelling and cleft lip and palate

General physical examination:

The newborn presented with large cystic swelling over the occipital region. She also had cleft lip and palate. Cyanosis noticed over her finger and toes. The pulse rate was 160/min regular rhythm and adequate volume. On airway examination we found that neck extension and mouth opening were inadequate.

Systemic examination

On systemic examination respiratory system showed bilateral equal air entry and was clear. Cardiovascular system also normal on auscultation. Perabdomen examination was soft no organomegaly detected.

3. Investigation

NCCT brain- approx 7×13mm defect in occipital bone in midline forming approx 15×7×16 cm sized sac with septation suggestive of occipital meningocele. Small posterior fossa with inferior displacement of cerebellar hemisphere &vermis of upto8 mm into upper spinal canal suggestive of Arnold Chiari malformation. Lateral ventricles have characteristic upturned apices giving Viking helment appearance. Finding suggestive of p/o Arnold chiari3 malformation with corpus callosum agenesis. 2D echo-dextro transposition of great arteries with large ASD, situssolitus, levocardia.

Other lab investigation were within normal limits.

Pre operative Preparation

- Pediatric stylet
- Different size pediatric mask
- Proper size endotracheal tube and laryngeal mask airway
- Blood reserved in blood b

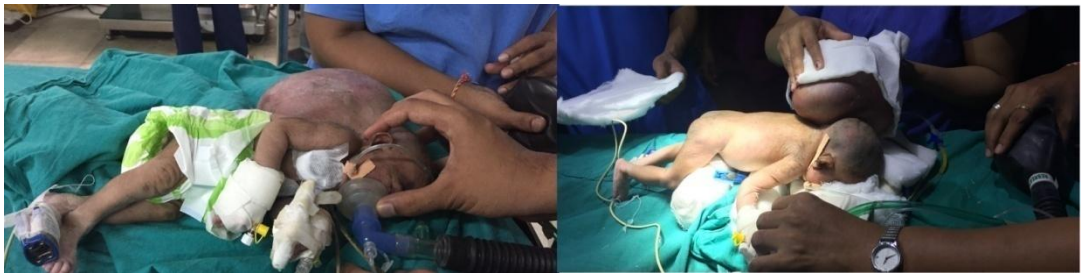

Figure 2: (a) mask ventilation in lateral position (b)final position after intubation

Baby could not be placed in supine positionso mask ventilation & induction done in lateral postion.(figure 2(a))Inj .Glycopyrrolate 0.004mg/kg, emset 0.15mg/kg IV, injparacetamol 15mg/kg and sevoflurane (8%) used for induction. Intubation first tried in lateral position but it failed due to inadequate exposure. After proper mask ventilation, intubation again tried in supine position with baby lifted up from the table and swelling and pelvis supported by other anesthesiologist .Baby finally got intubated with ET no. 3 mm after proper fixation of tube, baby made prone with extreme care to prevent accidental extubation and rupture of sac(figure2(b)). Pressure on the abdomen was avoided and pressure point including eyes were protected. Ventilation carried out by Jackson and Rees circuit and anaesthesia maintained by oxygen and sevoflurane (3%).Baby was covered properly after final positioning and a warmer placed at the foot end to prevent hypothermia.. Total 90 ml PCV & 150 ml crystalloid given to replace the fluid loss. The surgery lasted for 3hrs and 20 minutes. Intra op period was uneventful. After surgery neonate was not extubated and shifted to NICU electively on T-PIECE with O2 4 lit/min. She was vitally stable maintaining saturation 99%.

4. Discussion

The term cephalocele refers to a defect in the skull & dura with extracranial extension of intracranial structures. Meningocele is protrusion of only meninges.

Incidence is 1 in 5000.[1]Associated common congenital defect include club foot, hydrocephalus, exstrophy of bladder, prolapsed uterus, klippel- feil syndrome & congenital cardiac defect.[2]

Difficult airway management, prone position, protection of neural placode, assessment of volume status & prevention of hypothermia are the major challenges encountered by the anesthesiologist.[3]Mask ventilation &inubation may be attempted in lateral position[3] or in alternative approach including to place the child in supine position on a platform of rolled up blankets while one assistant temporarily supports the head or placing the child head beyond the edge of the table with assistant supporting it.[4]We used the similar technique. We used inhalational anesthesia with adequate mask ventilation for intubation.
Intensive monitoring is required to estimate the blood loss and to replace it adequately. Removal of a large quantity of csf carries volume and electrolyte imbalance which need to be corrected perioperatively.

We report a very rare case of triad of giant meningocele with DTGA and ASD & cleft lip and palate. Associated congenital abnormality further added to the challenges. Despite difficulties, intubation &anaesthetic management in our patient successfully achieved.

5. Conclusion

Anesthetic management of newborn with large occipital meningocele is challenging due to presence of difficult airway, prone position, associated congenital abnormality, hypothermia and fluid loss replacement. Vigilant evaluation with careful management result in safe and successful result.

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Author Profile

Dr. Monal Ramani (Associate professor) BJ medical college, Ahmedabad)

Dr. Deepshikha (third yr resident doctor, MD Anaesthesia, BJMC Ahmedabad)

Dr. Prachi Agnihotri (third yr resident doctor, MD Anaesthesia, BJMC Ahmedabad)