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Original Research Paper

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A CASE REPORT OF MENINGOMYELOCELE REPAIR

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ABSTRACT during the first 4 weeks of gestation. Early surgery should be performed to prevent rupture and infection preferably within 48 hours of birth. Positioning of the patient to secure the airway and the anesthetic approach is of prime importance. Proper positioning to protect the rupture of thin walled sac when supine and to avoid compression of cervical cord due to Arnold Chiari Malformation (ACM) is also required. Here, we report a case of 5 month male child weighing 8.5kg 7 kg for MMC repair having ACM type 3 with hydrocephalus.

KEYWORDS : Meningomyelocele, Cingenital Spinal Anomaly, Neural Tube Defect, Arnold Chiari Malformation, Hydrocephalus.

Introduction

Embryogenic neural tube fusion occurs during the first 4 weeks of gestation. Failure of fusion causes herniation of meninges with or without neural element: Meningomyelocele or Meningocele (1, 2).Folic acid helps to prevent neural tube defects when given from first trimester itself or even before planning a pregnancy. Arnold Chiari Malformation (ACM) and hydrocephalus are generally associated abnormalities with MMC. The clinical symptoms present as per the anatomical location of the defect. Various symptoms may occur as a result of tethering of spinal cord by sacral nerve root (3).

ACM is a downward displacement of the cerebellar vermin into upper cervical spinal canal and elongation of brain stem and 4th ventricle. Therefore when ACM is present with MMC and hydrocephalus, it requires special positioning during intubation, and also there should be no pressure on the exposed neural placode and a postoperative watch for stridor and apnoea (4).

So, the administration of anesthesia and the management in MMC is challenging. The anesthetic concerns include the pediatric age group, airway management, positioning, associated systemic co morbidities and post operative recovery after repair (5).

Case Report

A 5 month old male child presented with swelling in the back of neck and head region, which was size of lemon at birth and gradually increased to a size of tennis ball. There was no difficulty in breathing, no urinary symptoms or stool complaints were present. The swelling was diagnosed on Ultrasonography (USG) at 8 months of gestation child but presented at 5 months after birth for repair.

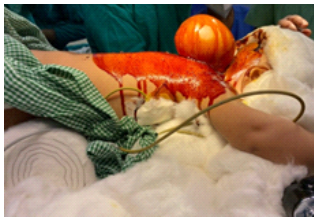
On examination, the swelling is in the cervico thoracic region posteriorly of size 5 × 7cms. MRI C-Spine showed splaying of posterior elements at the level of C7-D2 vertebral bodies with herniation of spinl cord, meninges and nerve roots through a defect of size 1.1 cm (craniocaudal) into a multi septated CSF filled sac in the cervicothoracic region of size of size 6.2 × 6.4 × 7.4 cms s/o cervical myelomeningolcele with significant tonsillar and approximately 5 mm medullary herniation into cervical spinal canal as described- features are consistent with Arnold Chiari malformation type 3.

He had normal vitals on presentation with an Hb of 9gm %, platelet count of 1.45 lac, blood sugar of 81 mg/dl and a normal range of liver and kidney functions. The child was posted for MMC repair. Difficult airway was anticipated due to the site of defect and difficulty in supine position of the patient. Operative procedure was explained to the parents and a written informed consent was taken. Nill Per Oral Guideline were given the night prior to surgery as follows. NPO solids – 6 hours, breast milk – 4 hours and clear fluids 2 hours. An i.v. line was secured using 24G i.v. cannula in the left forearm preoperatively.

On the day of surgery the child was premedicated using midazolam 0.5 mg/kg 30 min prior to surgery. When taken in the operating room, IV fluid was started using Isolyte-P at the rate of 6ml/kg/hr. All standard monitoring (SpO2, ECG, NIBP) were attached along with special monitoring like: core rectal temperature and the baseline parametes were recorded. Position for mask ventilation and intubation was decided to be a left decubitus position.

Preoxygenation was done using 100% oxygen with face mask of specific size on Jacson Reed's Circuit while premedication was given using i.v. atropine 0.01mg/kg and fentanyl 1mcg/kg. After 3 minutes, intravenous induction was initiated with Propofol 10mg i.v. and vecuronium 0.1mg/kg. after adequate muscle relaxation the child was intubated using uncuffed 4.0mm ID endotracheal tube. Once the position was confirmed using capnograph tracing and bilateral air entry, the tube was connected to the anesthesia machine (close circuit ventilator; Vt 8ml/kg; RR 18/min) and anesthesia was given using oxygen (40%) : N2O (60%) : Sevoflurane (1-2%) mixture. Prone position given for MMC repair (Figure 1), with proper size bolster under iliac crest and shoulder, keeping abdomen free for ventilation and prevention of venous congestion with proper pressure points care.

Figure 1



Special care taken to keep the child warm with warm IV fluids and irrigating fluids, warm blanket and wrapping of limbs with cotton. Anesthesia was maintained using oxygen, nitrous, sevoflurane and intermittent vecuronium. The

surgical correction was started (Figure 2) which lasted 90 minutes and the repair was corrected (Figure 3). The patient remained stable and blood was replaced as per blood loss.

Figure 2

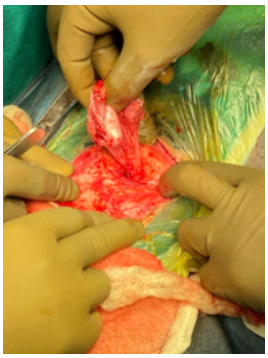
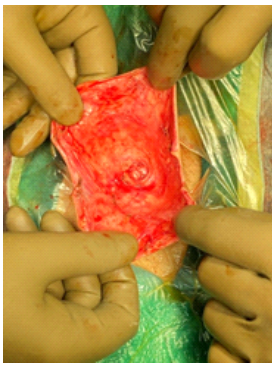


Figure 3



Reversal was standard with atropine and neostigmine, after adequate spontaneous respiration, eye opening and upper limb movements in supine position. For Postoperative pain relief, IV paracetamol and paediatric diclofenac suppository inserted, thereafter shifted to PICU for further management.

Discussion

MMC with an incident of 0.4 to 1 per 1000 live births is one the common malformations of CNS, with a leading cause of paralysis and lifelong disabilities including paraplegia, hydrocephalus, chiari malformation, incontinence, skeletal deformities and mental impairment. Folic acid should be supplemented ideally before conception or as soon as pregnancy is confirmed, to prevent neural tube defects. The timing of surgery is usually in the first 48 hrs after birth, to prevent rupture of sac, infection and neurological complications (6). About 1/3 rd of babies develop symptomatic chiari malformation where in cerebellar tonsils are displaced below the foramen magnum with elongation and compression of the brainstem and obliteration of cisterna magna , hence developing symptoms like inspiratory stridor, apnoea, swallowing dysfunction, bradycardia etc. This important associated abnormality gives us the indication for a very careful positioning during induction of anaesthesia, therefore avoid extension during laryngoscopy as it would lead to brainstem compression and ventilator dependence. 80-90% of MMC children have associated hydrocephalus, due to any contributory factor like aqueduct stenosis, 4th ventricle outlet obstruction, obliteration of posterior fossa , sub arachnoid space etc. Central venous access is an option to be considered as MMC repair requires blood replacement and IV fluids for a longer duration, especially with very fragile or no peripheral venous access in infants. Hypothermia in infants occur quite commonly under general anaesthesia, as it depresses the thermoregulatory response and autonomic control below defect is abnormal. Heat is lost from the core to the cooler peripheral tissue. This is compounded by the cold operating room. Thus prevention of hypothermia at the outset is important by very simple measures like adjusting the operative room temperature, wrapping of all extremities and head with cotton blankets, warm intravenous and irrigating fluids and inhaled anesthetics to be given through humidifier. Positioning of child for bag mask ventilation and intubation and similar precautions during extubation is a very important aspect of airway alignment management in MMC especially with associated ACM.

Reversal and recovery is equally crucial here. Extubation is to be done only when the child is awake and breathing well. The criteria for extubation are intact cough and gag reflex, a forced vital capacity in excess of 10ml/kg, an air leak around the tube, maintenance of oxygen saturation on spontaneous breathing, adequate reversal of neuromuscular blockade shown by sustained arm lift and spontaneous breathing. Post operative pain relief to be addressed as per standard institutional protocol.

Conclusion

Prevention of rupture of MMC sac is of prime importance. This further involves proper alignment and positioning for bag mask ventilation and intubation. Presence of Arnold Chiari Malformation poses a challenge during intubation and a risk of stridor /apnoea in the post-operative recovery. Paediatric airway management, intravenous access, correction of intra operative fluid and electrolyte imbalance, maintaining proper core temperature is a key to successful outcome of MMC repair.

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