CASE REPORT

Meningomyelocele Repair in a Premature Newborn with Hydrocephalus: Anaesthetic Confronts and Management

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Abstract:

Deficit of neural tube closure in the initial phases of intrauterine development leads to a gamut of abnormalities ranging from spina-bifida occulta, a relatively benign condition, to encephalocele and meningomyelocele, an anomaly in vertebral bodies, spinal cord and sometimes involving brainstem (in cervical meningomyelocele). Meningomyelocele is the most common nonlethal malformation in the spectrum of neural tube deficits. The intrinsic challenges associated with the latter disorder warrants tailor-made approaches for providing anaesthesia to the requisite therapeutic surgical interventions. Pediatric patients pose a set of natural barriers because of their ever budding and maturing neuro-physiological status, apart from the central neural disease process. Hence, in order to provide optimal neuro-anaesthetic care, the anaesthesiologist must have the knowledge of the outcomes of various pharmacologic interventions on cerebral aerodynamics apart from his professional experience in pediatric neuroanaesthesia. The current case report accounts for a challenging anaesthetic management in a premature newborn having hydrocephalus and lumbosacral meningomyelocele, presented for surgical repair within four hours of delivery.

Keywords: Hydrocephalus, Meningomyelocele, Neural Tube Closure, Neuroanaesthesia, Premature Newborn, Surgical Repair.

Introduction:

Meningomyelocele (MMC) has been depicted as "the most intricate and treatable congenital anomaly compatible with life" [1]. It is the most common nonlethal malformation in the spectrum of neural tube deficits. Development of central nervous system occurs in three phases-neurulation, canalization and retrogressive differentiation. During neurulation, neural tube folds upon itself to form a groove and then fuses to form the neural tube. Defects in neural tube occur during neurulation [2]. Failure in early stages leads to total dysraphism within the brain and spinal cord. Failure in canalization leads to spina bifida: myelocele exposes neural tissue; MMC exposes meninges along with neural tissue while meningocele contains meninges only. MMC can occur anywhere throughout the spinal column but is most commonly seen in lumbar region. Globally, the annual incidence is 0.4-1 per 1000 live births and it also varies with environmental and genetic factors [1-4]. Apart from the neural tube defect, several associated anomalies for example hydrocephalus, syringomyelia (a fluid-filled cyst within the spinal cord), hip dislocation concern the anesthetists. Mostly, along with hydrocephalus concomitant Arnold-Chiari II malformation (downward displacement of the cerebellum to brainstem with medullary kinking) is also seen

which can cause cervical cord and brainstem compression during neck extension required for intubation [4]. Managing the airway in premature newborns with hydrocephalus is a crucial issue as they show a diminished response to hypoxia and increased susceptibility to post operative apnoea [5]. Even direct pressure to the exposed neural placode can increase the complications [4], so proper positioning of neonates during induction of general anaesthesia and surgery must always be considered. In addition, these patients have a higher incidence of associated other congenital anomalies like Arnold Chiari malformation, Meckel syndrome, Dandy-Walker syndrome, Aqueductal stenosis etc [6]. Usually neurological manifestations of MMC are not very apparent at the time of birth but early surgical interventions prevent future neurological deterioration [7].

Closure of MMC within 72 hours of birth is recommended to prevent meningitis. In utero repair of the defect is an alternative in less than 20% of pregnancies but also depends on the technical feasibility and surgical services availability. Currently, palliative surgical care is mostly offered to correct the defect [8]. Owing to paediatric age group, associated congenital anomalies and co morbid conditions surgical management of MMC is demanding, not only for administration of anaesthesia, but also for providing peri-operative care [9]. Here we report a case report on successful anaesthetic management of a premature newborn presenting with hydrocephalus and lumbosacral meningomyelocele.

Case Report:

A 25 year old female underwent emergency caesarian section for twin pregnancy at 35 weeks of gestation due to premature rupture of membranes. Her prenatal Ultrasonography (USG) revealed hydrocephalus in second twin. During emergency caesarian section surgery, first live female baby was delivered at 0848 hours and second live female baby was delivered one minute later. The second baby's Appearance, Pulse, Grimace, Activity, and Respiration (APGAR) score at 1 min was 8/10 and at 5 min it was 10/10. This female baby had large head with features suggestive of hydrocephalus and examination also revealed a lumbosacral meningomyelocele. Birth weight was 2.0 kg. Immediately, after the delivery, the baby was shifted to Neonatal Intensive Care Unit (NICU) and was examined by neurosurgeon, for further management. Non-contrast Computerized Tomography (NCCT) scan brain was also done, which suggested hydrocephalus and lumbosacral MMC with no other obvious anomalies. It was decided to do Ventriculo-Peritoneal (VP) shunt procedure followed by surgical repair of the MMC. Hence, the new born was shifted to operation theatre after four hours of delivery. The baby was kept nil per oral after the delivery, with ongoing maintenance intravenous fluid (N/5) being given in NICU (till the time she was shifted to operation theatre).

On examination of the baby, her heart rate was found to be 144 per min, respiratory rate was 50 per min and electrocardiogram was within normal limits. The cry of the baby was good. High risk consent and consent for post-operative ventilation was obtained from the parents. Monitors used were ECG, SpO_2, (NIBP), skin temperature and capnography and baby was placed on operating table with a warmer placed underneath.

The neonate was pre-medicated with injectable glycopyrrolate dose of 5 mcg/kg, injection midazolam of 0.03 mg /kg and injection fentanyl of 4 mcg intravenously. Baby was pre-oxygenated and induction was done with injection thiopentone 10 mg and sevoflurane 1 mg Minimum Alveolar

Concentration (MAC). Subsequently, intubation was done using size 0 miller blade and a 2.5mm uncuffed endotracheal tube. To get the ideal supine position, gauze bandages were kept below the shoulder, chest and abdomen to compensate for the large head and the swelling in the lower back. After confirmation of the endotracheal tube placement, injection atracurium 1 mg intravenously administered and anaesthesia was maintained with oxygen and air (50% each) and sevoflurane 0.8 MAC. After the VP shunt procedure which lasted for 35 min, the baby was shifted to prone position with careful precautions being exercised for MMC repair.

During surgical repair, dystrophic meninges were excised and transfixed followed by a water tight dural closure. The recovery was smooth with mobility maintained in all the four limbs. The total blood loss during the procedure was 12 ml, which was well managed with IV Fluid (N/5). The total amount of IV fluid given during the surgery was 50 ml with surgery lasting for 150 min. Muscle relaxation was maintained with injection atracurium top ups (0.5 mg each) and intravenous paracetemol 30 mg was given for analgesia.

Post-surgery, neuromuscular blockade was reversed with injection neostigmine 100 mcg and injection glycopyrrolate 20mcg and the baby was extubated. Throughout the perioperative period, baby's vitals were stable. Baby was then shifted to NICU for further monitoring. Postoperatively also, vitals of the baby were stable with a good cry and movement of all four limbs. Ryles's Tube [6FG] feed was started after 06 hrs of surgery. Intravenous antibiotics and RT feeds were continued for 10 days post-surgery. Gradually, breast feeding was started and the baby was subsequently discharged to home after 15 days of surgery.

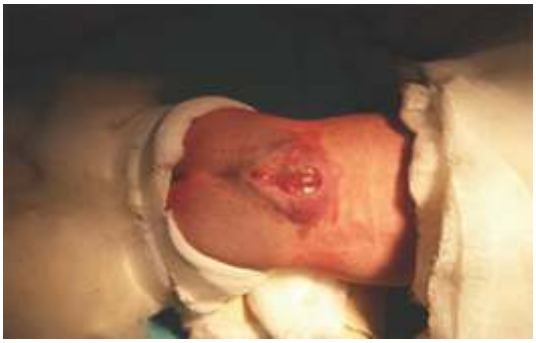

Fig. 1: Pre-operative MMC

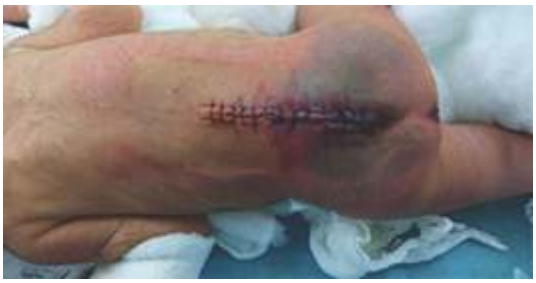

Fig. 2: Patient on Post-Operative Day 7th

Discussion:

Globally, MMC is considered as one of the leading and most common cause of infantile paralysis associated with several congenital anomalies and problems like hydrocephalus, paraplegia, mental impairment, incontinence, sexual dysfunction and Chiari II malformations [10, 11]. Chiari malformations (Type I, II, III and IV) are mainly structural defects in the cerebellum which usually results from downward displacement of the cerebellar tonsils through the foramen magnum (the opening at the base of the skull), sometimes causing non-communicating hydrocephalus as a result of obstruction of Cerebrospinal Fluid (CSF) outflow. However, vigilant antenatal screening procedures and oral folic acid supplementation in antenatal cases has reduced the incidence of MMC drastically [10, 11]. Comprehensive preoperative evaluation of all the organ systems in newborns is

necessary as embryological formation of all these systems had occurred concurrently along with the malformed neurological system [3].

It is imperative that MMC repair should be performed in the first 48 hours of life as delayed surgery increases the risk of infection [12]. Mostly, MMC is located in the lumbosacral region, but in rare instances it may be seen in cervical region and has got better neurological prognosis than lumbosacral MMC [13, 14].

Hydrocephalus is the most common anomaly associated with MMC and is found in almost 85-90% cases of MMC. This occurs either due to aqueductal stenosis or ventricular outlet obstruction or posterior fossa obliteration [15, 16]. Clinically, hydrocephalus causes increase in the total head circumference of the newborn so the need of surgery depends largely on the degree of increase in Intracranial Pressure (ICP), most common being the VP shunt surgery. Also, it exaggerates the risk of hypoxia and post-operative apnoea in newborns [17].

Out of all, general anaesthesia is preferred for paediatric neurosurgeries as it avoids an increase in intracranial pressure during the surgery and lessens significant hemodynamic instability. Intravenous induction is the preferred technique in case of raised ICP and can be performed either with thiopentone or propofol with or without a neuromuscular blocking agent. However, in cases without an IV access, inhalational induction can be done with sevoflurane [18].

Generally, neurosurgeries in children are performed in difficult surgical positions which impart certain challenges to the anaesthetist. Firstly in Ventriculoperitoneal (VP) shunt surgery patient is kept supine with head turned to opposite side for surgical access and ETT is also secured on the opposite side which may lead to loosening of adhesive tapes due to oral secretion while MMC repair is done in prone position, it is particularly important that the patients abdomen should be kept free. Excessive pressure on abdomen can hamper ventilation and venacaval compression can cause increased bleeding due to increased epidural venous pressure. Also improper padding in prone position can lead to congestion of face and tongue and pressure on the eyes may cause optic nerve damage.

Along with all the routine monitoring techniques skin temperature monitoring is also advisable in children as less subcutaneous fat and greater surface areas compared to adults make them more susceptible to hypothermia leading to increased vulnerability to apnoea, bradycardia, hypotension, acidosis and delayed recovery from neuro-muscular blockade [19].

All the routine monitoring was done in our patient and normothermia was maintained using hot air warming blanket, warm intravenous fluid and wrapping the head and extremities with cotton rolls. Normovolemia and thus hemodynamic stability is maintained by providing calculated intravenous fluid replacement. Although blood was reserved preoperatively but not used. The process of recovery is crucial as well. Rapid awakening is desired for early neurological assessment and hemodynamic stability. Extubation should be done only when the child is awake and breathing well [20].

Our case was different being the single young anaesthesiologist managing the case, a premature low birth weight newborn imposing challenges both for anaesthesia and surgery and strict adherence to basics in maintenance of temperature, oxygenation, fluid therapy and hemodynamic stability.

Conclusion:

Newborns are prone for anaesthetic complications, hence, safe anaesthetic management can be done by understanding the age related pathophysiology and dealing with the challenges and problems of ventilation, positioning, fluid management and temperature regulation. The case is presented for its rarity and successful management.

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