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Omphalocele: a Case Report

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Abstract

A neonate with giant omphalocele was posted for repair on 3rd day of life. Anesthesia management was challenging as liver and intestines herniated through the anterior abdominal wall defect. Anticipating post-operative respiratory embarrassment, the child was kept on ventilatory support for two days, recovered fully.

Keywords: Gaint Omphalocele, Abdominal Wall Defects

1. Introduction

Omphalocele is defined as a midline abdominal wall defect of the herniated viscera covered by a membrane consisting of amnion on the outer surface and peritoneum on the inner surface. This is also known as exomphalos. Incidence of omphalocele ranges between 1.5 and 3 per 10,000 births[1]. Children with omphalocele have very high (50%–70%) incidence of associated anomalies[1]. In live born babies the incidence of associated anomalies is lower because those who have multiple and serious anomalies are born dead[2]. The etiology of omphalocele is unknown, but maternal factors may play important role in this condition.

2. Case History

A 23 year old woman, primigravida delivered vaginally a full term male child weighing 2.5 kg. Baby cried well immediately after birth. As there was swelling over umbilical region, the baby was immediately transferred to our hospital for further management. Baby was seen by pediatric surgeon and diagnosis of omphalocele was made. Laboratory investigations were done: hematocrit - 53.1%, total count – 23600, liver enzymes, serum electrolytes, coagulation profile were within normal limits. Ultrasound examination of abdomen revealed omphalocele with herniation of portion of liver, gall bladder and intestines. 2D echocardiogram showed 3mm ventricular septal defect with bidirectional shunt, with mild pulmonary stenosis. Baby was operated on third day of life. After proper preparation and premedication, patient was induced with Inj. Ketamine 1.5 mg/kg and intubated with endotracheal tube no 2.5 plain, under Inj. Atracurium 0.5 mg/kg. Patient was maintained on sevoflurane with oxygen. Nitrous oxide should be avoided in order to prevent further bowel distention. Adequate care was taken to prevent hypothermia, as all neonates are prone to hypothermia because of their large surface area to body weight ratio. Third space losses may be significant and require replacement with cristaloids and colloids. Blood loss must be monitored throughout surgery and should be replaced if required. Intraoperative period and surgical procedure were uneventful. Baby was shifted to the NICU and put on ventilator anticipating the risk of respiratory embarrassment due to raised intraabdominal pressure.

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After five days of surgery, baby was weaned off from mechanical ventilation; adequate respiration was confirmed and patient extubated. Patient was discharged after 10 days.

3. Discussion

Paediatric surgeons and anaesthetist plays important role in the management of anterior abdominal wall defect (omphalocele). The midgut herniates outward through the umbilical ring and continues to grow at the time of normal development of the human embryo. The midgut returns back into the abdominal cavity at the end of 11th week of gestation. At the same time normal rotation and fixation occurs, with closure of the umbilical ring. The omphlocele is a condition in which the intestine fails to return into the abdominal cavity, and infant is born with abdominal contents protruding directly through the umbilical ring[3]. Omphalocele is associated with major or minor malformation in more than 50% cases. The most common is cardiac anomalies. Other anomalies includes musculoskeletal, gastrointestinal, and genitourinary. The triad of Beckwith-Wiedemann syndrome is (omphalocele, hyperinsulinemia and macroglossia). Mortality is 30%, cardiac defect and prematurity being the major causes.

The incidence of herniation of intestines into the cord is approximately 1:5000 live births, while herniation of liver and intestines occurs in 1:10000 live births, with a male predominance[4]. Antenatal ultrasound examination can identify an infant with an omphalocele as early as 12th gestational week[5]. Alpha-Fetoprotein (AFP) levels are raised in these patients. Preconceptional multivitamin use is associated with 60% reduction in risk of non syndromic omphalocele[5].

The new born with an abdominal wall defect offers many challenges to anaesthetist and has unique presentation. Heat loss from exposed abdominal content causes hypothermia. Management includes fluid and electrolyte maintenance, prevention of sepsis, gastric distention and cardio-respiratory stability. Surgical management includes primary closure (small omphalocele) or staged closure (giant omphalocele) to repair the defect. A prostheticsilo placement for gradual reduction and skin flap, tissue expanders and external compression with bandages is required for giant omphalocele[6]. Immediate post operative complications are increased intra abdominal pressure and respiratory instability due to diaphragmatic elevation. Other complications include necrotizing enterocolitis, prolonged ileus, respiratory distress, feeding intolerance, gastroesophageal reflux and neurodevelopmental delay[7]. Complications occur mostly with giant omphaloceles.

Advances in surgical therapies, anaesthetic techniques and nursing care have improved the outcome for infants with omphalocele. Survival rate is 75% to 95% for infants with small omphalocele[7]. Infants with associated anomalies and giant omphalocele have the poorest outcome.

4. Conclusion

Surgical management of giant omphalocele is a challenging case for pediatric surgeon and anesthesiologist. Key point managing this case includes temperature, electrolytes maintenance and cardiorespiratory stability intraoperatively.

Postoperative ventilation is necessary to prevent respiratory embarrassment.

5. References

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