Case Report

Early primary repair of giant omphalocele with favorable neo-natal outcome

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Abstract

Background: Omphalocele is a congenital midline abdominal wall defect characterized by herniation of abdominal contents (liver and bowel) into a membranous sac. We report a term female neonate with antenatally diagnosed large omphalocele with liver as content. Prenatal evaluation including karyotyping and whole exome sequencing was normal. A high-risk neonatal team attended delivery, and the sac was immediately protected to minimize fluid and heat loss. A temporary silo was placed to facilitate gradual reduction by gravity. The infant underwent elective primary repair on day 2 of life, with successful reduction of contents and closure of a 6 × 6 cm defect. The postoperative course was uneventful with early extubation, successful advancement to full feeds. The infant was discharged in stable condition after an 8-day NICU stay.

Key words: Omphalocele; Herniation; Cesarean section

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1. Introduction

Omphalocele is a congenital midline abdominal wall defect resulting from failure of return of midgut into the abdominal cavity during embryogenesis. It has a prevalence of approximately 2.6 per 10,000 births. It presents as a defect at the umbilicus covered by a membranous sac containing abdominal viscera, typically bowel and often liver, with the umbilical cord inserting at the apex of the sac. Approximately 80% of omphaloceles contain liver, and this has prognostic significance, as smaller non–liver-containing defects are more commonly associated with aneuploidy, while larger liver-containing omphaloceles are usually associated with euploid fetuses. Omphaloceles are variably classified as small (<5 cm) or giant when large or containing a significant portion of the liver. Associated structural anomalies are common and influence outcomes.

2. Case Presentation

A term female neonate (38 weeks, birth weight 3 kg, appropriate for gestational age) was born to a Rh-negative mother by elective lower segment cesarean section. Antenatal ultrasound revealed a large omphalocele containing liver. Prenatal evaluation with amniocentesis, including karyotyping and whole exome sequencing, was normal. A multidisciplinary team comprising obstetric, neonatal, and pediatric surgical specialists was involved in antenatal planning prior to delivery.

A high-risk neonatal team attended the delivery. At birth, the baby cried and delayed cord clamping was done. After stabilizing the airway and breathing, the omphalocele was covered with sterile moist gauze and enclosed in a sterile urobag to protect the contents and reduce insensible fluid loss (Fig 1). Careful examination confirmed the presence of liver and intestinal loops within the sac.

(a) **(b)** **c)**

Fig (1): a) Care in the delivery room focuses on respiratory management and protecting the omphalocoele contents. b) Initial NICU stabilization and protecting contents in a silo allowing gradual reduction by gravity. c) Post operative care after primary closure.

The infant was shifted to the NICU and stabilized. Intravenous fluids started. Pediatric surgical evaluation was performed, and a temporary silo was placed to facilitate gradual reduction of the contents by gravity (b). Preoperatively, glycerin enema was administered for bowel decompression. On day 2 of life, the baby underwent elective primary repair under general anesthesia. Intraoperatively, a 6 × 6 cm abdominal wall defect was noted with herniation of the left lobe of liver and small bowel loops. The sac was excised, adhesions were released, contents were gradually reduced, and primary closure was achieved following mobilization of surrounding structures. The intraoperative course was uneventful.

Postoperatively, the infant was electively ventilated and extubated within 24 hours (c). Baby required brief CPAP support for mild respiratory distress and was weaned to room air by day 6 of life. Enteral feeds gradually advanced, achieving full feeds (100 ml/kg/day) by postoperative day 3. Feeding progressed from gavage and paladai feeds to direct breastfeeding by day 7, with no feeding intolerance. Initial echocardiography showed a moderate patent ductus arteriosus (3 mm) and patent foramen ovale with left-to-right shunt, which resolved on repeat evaluation. Neurosonogram and abdominal ultrasound were normal. Hearing screening was passed bilaterally. The infant remained

clinically stable, gained weight (discharged weight 3.128 kg), and was discharged on full oral feed after an 8-day NICU stay, with advice for follow-up.

3. Discussion

Omphalocele requires meticulous antenatal, delivery room, and postnatal management to optimize outcomes. Antenatal identification and genetic evaluation are crucial, as associated anomalies and aneuploidy significantly influence prognosis. In this case, the presence of a liver-containing omphalocele with normal genetic evaluation supported a favorable outcome. Delivery room management focuses on protecting the sac, preventing hypothermia, and minimizing fluid loss. The use of sterile moist gauze and an impervious covering such as a urobag helps preserve sac integrity. Early involvement of the surgical team enables timely planning. Temporary silo placement, even in cases planned for primary closure, facilitates gradual reduction and prevents sudden increases in intra-abdominal pressure. Restoration of abdominal domain must be carefully balanced, as excessive intra-abdominal pressure may lead to reduced venous return, oliguria, and respiratory compromise. Postoperative care includes monitoring respiratory status and perfusion, with particular attention to urine output, pulse rate, and blood pressure as indicators of intra-abdominal pressure. Early initiation and advancement of enteral feeds contribute to recovery and improved outcomes.

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