OMPHALOCELE: A CASE REPORT

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Abstract

An omphalocele is a herniation of abdominal content into the base of the umbilical cord. The gross appearance of omphalocele differs from that of gastroschisis in two important respects: A protective membrane encloses the malposition of abdominal contents and Elements of umbilical cord course individually over the sac and come together at its apex to form a normal-appearing umbilical cord.

This case report presents a female neonate with a large omphalocele containing the liver and intestinal segments, delivered via cesarean section at the University Clinical Center of Kosovo. Initial management focused on infection prevention, fluid balance, and respiratory stabilization. On the fifth day, a single-step surgical repair was successfully performed under general anesthesia, repositioning the herniated organs and closing the defect without complications. Postoperative recovery was stable, with gradual feeding tolerance and full respiratory adaptation. The neonate was discharged after 15 days in good condition.

Keywords: omphalocele, newborns, congenital.

Introduction

Omphalocele is a rare congenital malformation of the anterior abdominal wall, characterized by a hernial sac containing organs such as the liver and intestines. The incidence ranges from 1 in 4,000 to 1 in 10,000. It develops in utero, typically when the intestines fail to return to the abdomen by 11 weeks of gestation. Two-thirds of cases may involve abnormalities in the spine, heart, digestive, urinary, or limb systems. Omphaloceles vary in size: small ones typically contain only the intestine, while large ones may involve the liver, spleen, and gastrointestinal tract.[ ][ ]

Diagnosis- This anomaly is usually apparent. A ruptured omphalocele may be confused with a gastroschisis; both defects are characterized by exposed intestine, but infants with omphalocele do not possess an intact umbilical cord at the level of the abdominal wall to the left of the defect.[ ]

Management of omphalocele varies depending on the size of the defect and the organs involved. In cases of large omphaloceles, especially those containing the liver within the hernia sac, cesarean delivery is often recommended to prevent sac rupture. However, for smaller omphaloceles, vaginal delivery may be considered a reduction, even in stages over a lengthy period, which may be very difficult to achieve.[ ]

There are two main therapeutic groups: ruptured sac (requiring emergent surgery) and intact omphalocele (less urgent, with the sac protected until surgery). Nonoperative staged reduction has been described, and skin coverage may be used as a temporary measure.[ ][ ]

Prognosis is influenced by defect size and birth weight, with survival rates above 90% in the absence of additional malformations. [ ][ ]

This case report highlights the successful single-step surgical treatment of omphalocele.

Case Report

A female neonate was delivered via cesarean section at the University Clinical Center of Kosovo- Neonatology Clinic due to a prenatal diagnosis of omphalocele. The delivery was scheduled to prevent rupture of the hernia sac, given the large defect that contained portions of abdominal organs, including the liver. At birth, the neonate weighed 4,250 grams, had a length of 54 cm, and a head circumference of 37 cm. Initial Apgar scores were 7 and 8 at 1 and 5 minutes, respectively, indicating a moderately stable condition. Following birth,

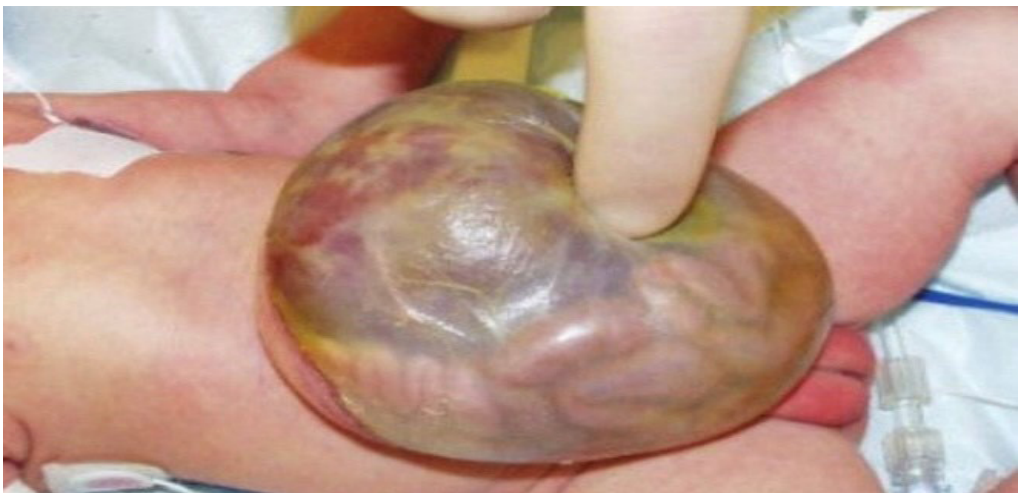

Figure 1. Omphalocele- the hernia sac and dressing (https://pmc.ncbi.nlm.nih.gov/articles/PMC1465128/pdf/annsurg01277-0124.pdf)

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a physical examination revealed a significant abdominal wall defect consistent with a large omphalocele. The hernia sac was intact, covering abdominal contents without evidence of rupture or dissection. Vital signs included a respiratory rate of 42 breaths per minute and an oxygen saturation (SpO2) of 92% on room air. The neonate displayed a regular heart rate, with no immediate evidence of cardiovascular compromise. Due to the size and location of the omphalocele, initial management focused on preventing infection and minimizing fluid and heat loss. The omphalocele sac was carefully wrapped in sterile saline-soaked dressings covered with a plastic wrap to maintain moisture and provide a barrier against pathogens. Nasogastric suction was initiated to decompress the stomach and reduce the risk of abdominal distension, which could exacerbate respiratory or circulatory compromise. Intravenous (IV) fluids were started promptly to maintain adequate hydration and compensate for any potential fluid loss through the hernia sac and dressings.

The newborn had mild breathing difficulty shortly after birth, likely due to limited diaphragm movement from the abdominal defect. The infant was kept in an incubator with an oxygen hood for four days before surgery to ensure stable temperature, humidity, and oxygen levels. Post-surgery, one day of ventilatory support was provided to assist breathing. Following this, the infant was transitioned to CPAP for three hours to support respiratory adaptation, then continued with oxygen supplied via hood for three days, and finally moved to breathing ambient air.

Admission laboratory values showed hematocrit at 51%, leukocytes at 21.7 $x10^9$/L, and platelets at 235 $x10^9$/L. Biochemistry results indicated a urea level of 3.9 mmol/L and creatinine at 71.6 µmol/L. Blood gas analysis demonstrated a pH of 7.22, with PCO_2 at 64 mmHg, PO_2 at 32 mmHg, sodium at 132 mmol/L, potassium at 4.1 mmol/L, calcium at 1.3 mmol/L, and bicarbonate at 26.2 mmol/L, pointing to mild respiratory acidosis, likely from restricted diaphragmatic movement. Electrolytes and renal function remained stable within normal limits, indicating no metabolic issues. During routine examination, an echocardiogram revealed a restrictive ductus arteriosus. Additionally, an abdominal ultrasound confirmed the hernia sac's contents, verifying no other abdominal organ abnormalities, essential information for the surgical team's preparation.

On the fifth day, surgery was performed to close the omphalocele. The hernia sac contained the liver, cecum, ascending colon, appendix, and ileum. Under general anesthesia, these organs were carefully repositioned, and the defect was closed without any complications.

Postoperatively, the neonate was monitored in the NICU for signs of respiratory stability, infection, and wound healing. The surgical site remained intact, and gradual feeding was introduced three days after the operation progressing from nasogastric to total enteral milk without gastrointestinal complications. The neonate tolerated feeding well, and bowel function normalized promptly.

The newborn baby stayed 15 days in Neonatology Clinic, 10 days in NICU AND 5 days near the mother.

Discussion

In this case, the choice of cesarean delivery was crucial to minimize trauma and protect the exposed organs, particularly given the hernia's size and the involvement of the liver. Studies

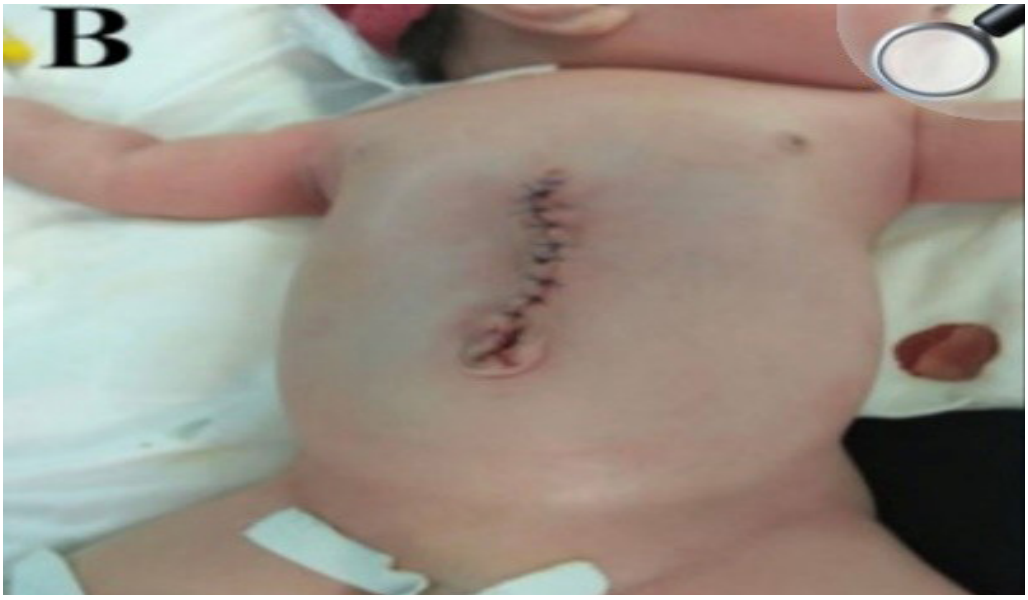

Figure 2. Post-Surgical Condition of Omphalocele Repair in Neonate. (University Clinical Center of Kosovo – Clinic of Neonatology)

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show that cesarean delivery can reduce the risk of sac rupture, especially when the liver is included[ ]

The initial management strategy involved covering the hernia sac with sterile saline dressings and plastic wrap, preventing both infection and fluid loss. This aligns with best practices, highlighting the importance of minimizing contamination risks. Additionally, nasogastric decompression prevented abdominal distention, helping to reduce respiratory strain.

Respiratory support in this case was minimal, with only a brief period of CPAP and oxygen therapy, indicating adequate pulmonary function. Respiratory challenges are common in large omphalocele cases, often due to pulmonary hypoplasia or diaphragmatic displacement, yet this neonate adapted well to spontaneous breathing.

Primary closure of the abdominal defect was achieved successfully. When possible, this is often preferred over staged repair, as it reduces infection risks and accelerates recovery.

The neonate's postoperative course was favorable, with no significant complications, allowing for early feeding and recovery. This case emphasizes the benefits of early enteral nutrition, which supports gut function and reduces the need for parenteral nutrition, thereby avoiding associated risks[ ]

Conclusion

eThis case highlights the importance of a multidisciplinary approach in managing neonates with omphalocele. The successful surgical intervention and careful postoperative monitoring ensured an optimal outcome for this neonate, with minimal respiratory support requirements and a rapid recovery. The case supports early surgical repair and appropriate perioperative care as effective measures for achieving positive outcomes for congenital abdominal wall defects.

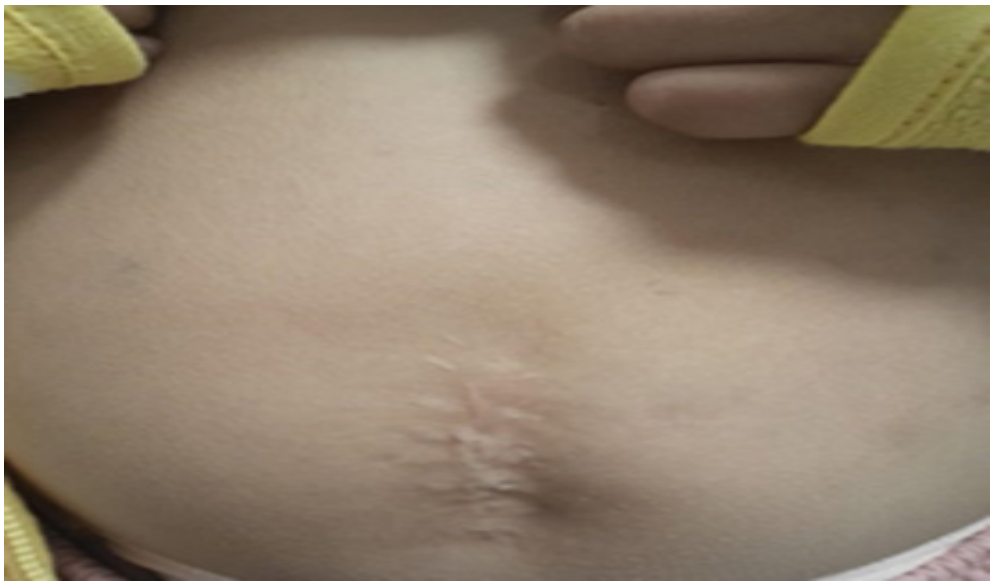

Figure 3. . Healed Surgical Scar Following Omphalocele Repair at Five Year Follow-Up.

Table 1. Neonatal Management of Giant Omphalocele

Surgical Techniques for Closure of Giant Omphalocele
Intraperitoneal tissue expander for greater intra-abdominal space
Progressive stretching of the abdominal wall
Muscle tissue expanders and translation of muscles
Sequential sac ligation
Vacuum-assisted closure
Bilateral longitudinal fibroperitoneal-aponeurotic transposition

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