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Case report

Surgical management of a massive omphalocele in a newborn: A case report study

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ABSTRACT

Introduction and importance: Omphalocele is a rare congenital defect in the abdominal wall, affecting about 1 in 5000 to 10,000 newborns. It occurs when abdominal organs protrude through an opening at the base of the umbilical cord. Treating massive omphaloceles is highly challenging, requiring innovative and staged surgical methods to avoid complications like increased intra-abdominal pressure and potential organ damage.

Case presentation: A female newborn with a 12.5 cm omphalocele was delivered emergently via cesarean section due to fetal distress. The exposed organs, including parts of the intestines and liver, were covered by a thin membrane. To minimize complications, a staged approach was opted for: first, a silo was placed to gradually reduce the herniated organs, followed by closure of the abdominal wall with absorbable sutures and biologic mesh.

Clinical discussion: Omphalocele in newborns is a serious congenital defect where abdominal organs protrude through the umbilical cord, covered by a membrane. It requires urgent medical care to prevent complications like respiratory distress and infections. Treatment typically involves a team of pediatric surgeons and staged surgeries to repair the defect and ensure the infant's long-term health.

Conclusion: The successful staged surgical method of silo reduction and biologic mesh for this massive omphalocele highlights the need for personalized surgical planning and multidisciplinary care. Six months later, the patient is thriving, showing no signs of recurrence or complications.

1. Introduction

Omphalocele is a rare congenital abdominal wall defect that occurs in approximately 1 in 5000 to 10,000 live births [1]. It is characterized by the herniation of abdominal organs, typically the liver, intestines, and occasionally other viscera, through an opening in the abdominal wall at the base of the umbilical cord [2]. Omphaloceles can be classified as small, medium, or large based on the size of the defect and the extent of the herniation [3].

Management of omphaloceles, especially massive defects, presents significant surgical challenges. Immediate closure of a large omphalocele sac can lead to increased intra-abdominal pressure, respiratory compromise, and potential organ damage [4]. Various surgical strategies have been described to address this issue, including staged repair, use of prosthetic materials, and delayed closure techniques [5,6].

This case report details the effective surgical treatment of a newborn with a large omphalocele using a staged approach. It offers important perspectives on the clinical decisions and surgical methods used to safely

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and successfully address this challenging congenital condition. The work has been reported in line with the SCARE 2023 Criteria [7].

2. Case presentation

2.1. Background

The female newborn was the product of an uncomplicated pregnancy until the third trimester, when routine ultrasound revealed a large abdominal wall defect. The decision for an emergency cesarean section was prompted by signs of fetal distress observed during the mother's labor. Upon delivery, the omphalocele measured an impressive 12.5 cm in transverse diameter, which is considered massive and poses significant surgical and medical challenges (Fig. 1).

2.2. Investigations

The physical examination upon admission to the NICU revealed a stable newborn with good respiratory effort and strong vital signs, despite the significant abdominal anomaly. Blood tests were conducted to assess the baby's overall health and to check for any signs of infection or electrolyte imbalances. An abdominal ultrasound was crucial in confirming the extent of the omphalocele and in ensuring that there were no other associated anomalies, such as obstructions or malformations within the gastrointestinal tract. Karyotyping was essential to rule out chromosomal abnormalities, which are known to be associated with omphaloceles in some cases. Additionally, the baby underwent a comprehensive screening for other congenital anomalies, which included a 2D echocardiogram to assess cardiac structure and function, abdominal ultrasound to evaluate gastrointestinal integrity, and a comprehensive metabolic panel to check for metabolic disorders.

2.3. Management

Given the size of the omphalocele and the significant risk of complications associated with immediate surgical intervention, the surgical team decided on a staged approach. The initial management focused on protecting the omphalocele sac to prevent rupture, which could lead to infection and other complications. While the newborn was stable at the time of admission, the presence of a massive omphalocele poses significant risks, including potential respiratory compromise due to increased abdominal pressure and the anatomical challenges presented by the defect. Therefore, respiratory support was implemented as a preventative measure to ensure adequate oxygenation and to monitor the infant closely for any changes in respiratory status. This proactive approach is critical in managing high-risk cases like this one, where the clinical situation can evolve rapidly. The respiratory support provided was primarily low-flow supplemental oxygen via nasal cannula, which minimized the risk of aerophagy and did not significantly increase intraabdominal pressure. This choice allowed for adequate oxygenation while avoiding the complications associated with more invasive ventilation modes.

The first stage of the surgical management involved the placement of a silo on the day of life two, a technique that allows for the gradual reduction of the herniated organs back into the abdominal cavity. The silo, made of silicon, was carefully placed over the omphalocele and secured in position. Over the next several days, the silo was gradually tightened, reducing the size of the opening and allowing the organs to be slowly repositioned into the abdomen. This process was closely monitored with serial abdominal ultrasounds and clinical assessments. We utilized ultrasounds as a precautionary measure to monitor bowel viability and intra-abdominal pressure in the context of the massive omphalocele. While clinical indicators such as NG tube output and urine output were essential for guiding our management, the ultrasounds provided an additional layer of monitoring to detect any subtle changes that could indicate potential complications.

Once the viscera were fully reduced into the abdominal cavity, the second stage of the surgery was performed on day of life ten. Under general anesthesia, the abdominal wall was closed primarily, which means that the muscles and fascia were reapproximated without the need for a prosthetic material to bridge a gap. However, given the large defect, a biologic mesh was used to reinforce the repair and to provide additional support to the abdominal wall. The use of a biologic mesh reduces the risk of hernia recurrence and allows for tissue ingrowth, promoting a more natural healing process. The additional role of mesh includes providing reinforcement to the abdominal wall, which helps prevent hernia recurrence, particularly in high-risk patients. Mesh acts as a scaffold that supports scar tissue formation, enhancing the stability of the repair over time.

The surgical team ensured that the closure was performed without tension to prevent potential complications such as wound dehiscence or impaired blood flow to the abdominal organs (Fig. 2).

2.4. Outcome

The patient's postoperative course was uncomplicated. She was extubated on postoperative day 3 and transitioned to nasal cannula oxygen. Enteral feeding was initiated on postoperative day 5 and advanced as tolerated. The patient was discharged home on postoperative day 21 in stable condition. At the 6-month follow-up visit, the patient was thriving, with normal growth and development. Physical examination revealed a well-healed abdominal scar, and the patient had no evidence of recurrent omphalocele or other complications.

3. Discussion

3.1. Background

Omphalocele in newborns represents a complex congenital condition where abdominal organs protrude through the umbilical cord, encapsulated by a transparent membrane. This anomaly, occurring during fetal development, requires immediate medical attention post-birth due to the risk of associated complications such as respiratory distress, infections, and gastrointestinal issues [8].

The exposed viscera included segments of the small and large bowel, as well as the liver, all of which were covered by a thin, intact membranous sac. This sac, derived from the peritoneum and amnion, provided a protective barrier against amniotic fluid during gestation and continued to protect the organs after birth. Treatment typically involves a coordinated effort between pediatric surgeons, neonatologists, and other specialists to provide optimal care for the newborn, often involving staged surgical interventions to address the defect and ensure the best possible long-term outcome for the infant [9].

The surgical management of a massive omphalocele in a newborn presents a unique set of challenges that require a multidisciplinary approach and meticulous surgical technique. This case report highlights the successful management of a 12.5 cm omphalocele, which is notably larger than the average size, making the surgical intervention more complex. The staged approach utilized in this case, involving the initial placement of a silo followed by definitive abdominal wall closure, is a testament to the adaptability and innovation required in pediatric surgery.

3.2. Surgical technique and innovations

The use of a silo in the management of omphalocele is a well-established technique that allows for the gradual reduction of herniated viscera into the abdominal cavity. This method is particularly beneficial in massive omphaloceles, where the risk of primary closure leading to increased intra-abdominal pressure and its associated complications, such as respiratory distress and potential organ ischemia, is significantly higher. The gradual reduction facilitated by the silo technique



 $\textbf{Fig. 1.} \ \ \textbf{The newborn baby with an huge omphalocele.}$



Fig. 2. Newborn baby recovering after successful omphalocele surgery.

minimizes these risks and allows for the abdominal wall to stretch and accommodate the viscera more naturally [10].

The definitive closure of the abdominal wall using a combination of absorbable sutures and a biologic mesh is another critical aspect of the surgical management. The use of biologic mesh in pediatric surgery has gained popularity due to its ability to reduce the risk of hernia recurrence and to facilitate tissue ingrowth, promoting a more robust repair. In cases of massive omphaloceles, where the abdominal wall defect is extensive, the reinforcement provided by the mesh is invaluable in preventing future complications [11].

3.3. Multidisciplinary care

The successful outcome of this case is also a reflection of the comprehensive, multidisciplinary care provided to the newborn. The involvement of neonatologists, pediatric surgeons, anesthesiologists, and nursing staff ensured that all aspects of the patient's care were addressed. The meticulous monitoring and support of the newborn's respiratory and nutritional status during the perioperative period were crucial in facilitating a smooth recovery [12].

3.4. Wound care

Effective care of the surgical wound after repairing a massive omphalocele is vital for preventing complications and ensuring proper healing [13]. The wound should be kept clean and dry, with sterile dressings applied to safeguard the incision from infection. Regular assessments of the wound are necessary to check for any signs of redness, swelling, or discharge that could suggest an infection [14,15]. Pain should be managed with suitable analgesics to keep the patient comfortable during recovery [16]. Moreover, adequate nutrition and hydration are important for promoting healing [17]. Caregivers should be instructed on recognizing early warning signs of complications and the significance of follow-up visits to monitor the wound's healing and the infant's overall recovery [18,19]. This thorough approach helps facilitate effective healing while reducing the likelihood of negative outcomes [20].

3.5. Implications for practice

This case report underscores the importance of a tailored surgical approach in the management of massive omphaloceles. It emphasizes the need for careful preoperative planning, the selection of appropriate

surgical techniques, and the importance of postoperative care in achieving successful outcomes. The use of innovative techniques such as silo reduction and biologic mesh reinforcement should be considered in the armamentarium of pediatric surgeons dealing with similar cases [21].

4. Conclusion

The case presented here demonstrates the effectiveness of a staged approach, utilizing silo reduction and biologic mesh reinforcement, in achieving a successful outcome. It highlights the importance of a multidisciplinary team in the care of these complex patients and provides valuable insights into the management of similar cases in the future.

Author contribution

Fatemeh Marivani (Corresponding author), Navid Faraji, Rasoul Goli and Niloofar Bagheri: study concept, data collection, writing the paper. Robab Choopani and Negar Mirzaei: writing the paper, reviewing and validating the manuscript's credibility.

Consent

Written informed consent was obtained from the patient's parents/ legal guardian for publication and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Ethical approval

Ethical clearance was not necessary by Research Committee of Urmia University of Medical Sciences as the format of this paper is a case report.

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The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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