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Case Report

A rare case of an omphalocele major with associated tetralogy of Fallot in a newborn

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ABSTRACT

Omphalocele major (OM) is a surgically challenging condition which owing to its large defect size and other congenital anomalies, is associated with poor outcomes. Complications while managing it are not uncommon. But here, we report a case of OM in a neonate who was successfully managed in a multidisciplinary approach by a team of neonatologist, surgeons and anesthetist.

Keywords: Omphalocele, Abdominal wall defect, Neonate

INTRODUCTION

An omphalocele is an abdominal wall defect which is congenital.[1] During the 6th week of intrauterine life there is physiological herniation of the intestine into the umbilical cord which gets reduced by the 12th week after 270-degree counter clockwise rotation. Omphalocele occurs when there is a defect in abdominal wall closure.

The abdominal contents protrude through an open umbilical ring. It is covered by a membrane consisting of peritoneum on the inner surface, amnion on the outer surface, and Wharton's jelly between the layers. The content of the sac includes mainly intestinal loops and sometimes liver. It is often associated with other congenital anomalies like chromosomal and cardiac defects.[2,3]

CASE REPORT

A one-day old neonate boy weighing 2.25 kg delivered vaginally at term presented to a large mass over abdomen. On examining the neonate, it was found to be a 12×10×7 cm lump protruding out from the umbilical region of the abdominal wall, containing bowel loops, covered with a thin, semi-transparent sac, and no overlying skin (Figure 1). On ultrasonography it was confirmed that patient has omphalocele major with bowel loops and liver protruding out in a sac. 2D echo revealed tetralogy of Fallot in an otherwise healthy child. On a chest X-ray characteristically boot shaped heart was visualised (Figure 2). After accessing new born for surgical fitness, decision was made to operate for it. Under general anesthesia, the sac of Omphalocele was opened up precisely without injuring the underlying viscera. Congested liver and small bowel loops were found to be protruding through the defect of 5×5 cm in the anterior abdominal wall (Figure 3).

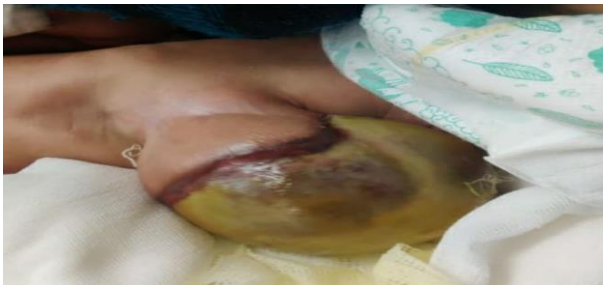

Figure 1: Pre-operative image of new born- showing lump of 12×10×7 cm containing abdominal viscera protruding through the defect in the anterior abdominal wall.

Liver was covered with hot saline mops for 10 minutes to reduce the congestion and edema, which facilitated easy reduction of it. Then, all the contents were reduced without difficulty. Later skin flap closure was done. Postoperatively the patient was monitored for the signs of abdominal compartment syndrome in the neonatal intensive care unit by neonatologist and team of operating surgeons. Patient's postoperative course was uneventful and he was discharged after suture removal on 14th day after surgery.

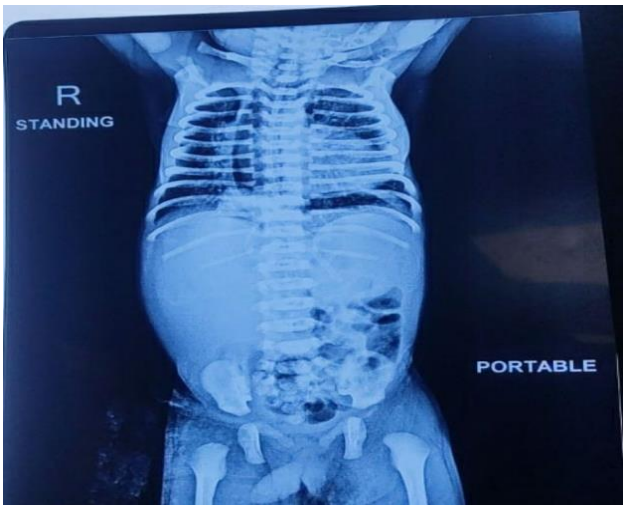

Figure 2: X-ray showing typical boot shaped heart shadow.

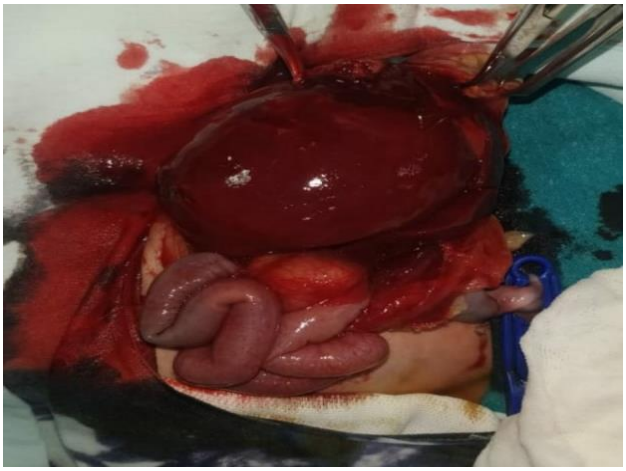

Figure 3: Intra-operative image showing edematous and congested liver with bowel loops after opening the omphalocele sac.

DISCUSSION

The incidence of omphalocele is around 1 in 3000 to 10,000 live births.[4] It can be classified into two types based on the size of fascial defect: omphalocele minor (defect size <5 cm) and omphalocele major (O) (defect size ≥5 cm). [1]

Both surgical and non-operative delayed closure methods to manage major omphaloceles have been reported. Surgical methods include primary closure and staged closure while non-operative delayed closure approach comprises of application of topical medications to promote neo-epithelialization and later on interval repair of remaining ventral hernia.[5]

Primary closure often leads to abdominal compartment syndrome. It occurs due to the size difference between sac contents and abdominal domain.[4,5] It can present post-operatively as increased intra-abdominal pressure, bowel obstruction, hemodynamic instability, bowel ischemia or respiratory distress. To avoid this complication, staged closure has been implicated in which multiple operations are required prior to final abdominal wall fascial closure. The placement of tissue expanders, skip flaps or mesh is also included in this technique.[5,6]

The intra-operative intravesical pressure assessment can help in excluding abdominal compartment syndrome and deciding upon the choice of closure.[7]

Regarding the outcome, a study by Vachharajani et al shows that major omphalocele have poorer prognosis compared to minor omphalocele.[8] Also, study by Akinkuotu et al shows that it is associated with the presence of concomitant anomalies including cardiac anomalies requiring surgery in the neonatal period, congenital diaphragmatic hernia, and chromosomal anomalies which significantly increased mortality and pulmonary morbidity.[9]

CONCLUSION

In regard to major omphalocele associated with cardiac anomalies, morbidity and mortality remains high. Despite that fact, with the help of a multidisciplinary team approach involving neonatologists, surgeons, anaesthetists, timely decision making and interventions in this case has shown exemplary outcomes.

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