CASE REPORT

Giant Omphalocele: A Congenital Anomaly Containing Bowel Loops

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Abstract

Omphalocele is a rare birth defect with an incidence of 1 in 4000 – 7000 live births. Through a defect in the abdominal wall, abdominal organs protrude out into the base of the umbilical cord. Giant omphalocele forms due to failure of lateral in-folding of the embryo, exposing the abdominal cavity with organs covered only by a thin membrane called the omphalocele sac. Surgical repair is done in stages.

Keywords: Omphalocele, Abdominal Wall Defects, Giant Omphalocele, Gastrochisis

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Introduction

Omphalocoele is an embryological defect of the umbilical ring and medial segments of the two lateral abdominal wall folds that form the anterior abdominal wall during foetal growth.[1] Theories help to explain the reason, such as failure of the bowel to migrate into the abdomen by 10–12 weeks and the failure of the embryonic lateral folds to fuse in the midline.[3]

Case Report

A term female new born with protrusion of a sac through the anterior abdominal wall was brought to Neonatal ICU. Informants gave history of a normal home delivery to a Gravida-2 Para-1, thirty year old mother. The previous child was normal and two years old. The patient did not have any antenatal checkup. No family history of similar complaints. On examination the baby was emaciated and dehydrated, cry was feeble. A delicate sac containing the abdominal contents and the cord attached in the centre was seen protruding through anterior abdominal wall defect measuring 5-6cms, which later gave way exposing the bowel loops and stomach **(Figures 1 & 2)**. The attendants were counseled for staged surgical procedure and genetic evaluation; the attendants did not consent and was discharged against medical advice.

Figure 1. Photograph showing new born with ruptured giant omphalocele. Stomach and small bowel loops are seen protruding outside the abdominal cavity.

Discussion

Omphalocele is differentiated from gastrochisis by the presence of the omphalocele sac. Gastroschisis is protrusion of intestine through the small umbilical defect of the cord. A ruptured giant omphalocele has the intra-abdominal organs outside the abdomen without a covering membrane. Gastroschisis are usually seen in the first pregnancy of young mothers while omphaloceles develop in older women. The size of the defect range from 4 cm to more than 10 cm. A minor omphalocoele occurs when the defect is 4 cm or less, major or giant omphalocoele has 5 cm or larger defect.[4] The hernial sac may contain small and large bowel, stomach, liver, spleen, urinary bladder, uterus and ovaries. Based on the site of the defect omphaloceles are named as epigastric (classic omphalocoele) with cephalic fold defect, central type with a lateral wall defect greater than 4 cm and the hypogastric/caudal type with caudal fold defect.[5]

Prenatal ultrasound in the second trimester of pregnancy (>12weeks) and level II fetal ultrasound determines type of omphalocele with the abdominal organs in the umbilical sac and even whether the sac is ruptured (free floating

*See End Note for complete author details

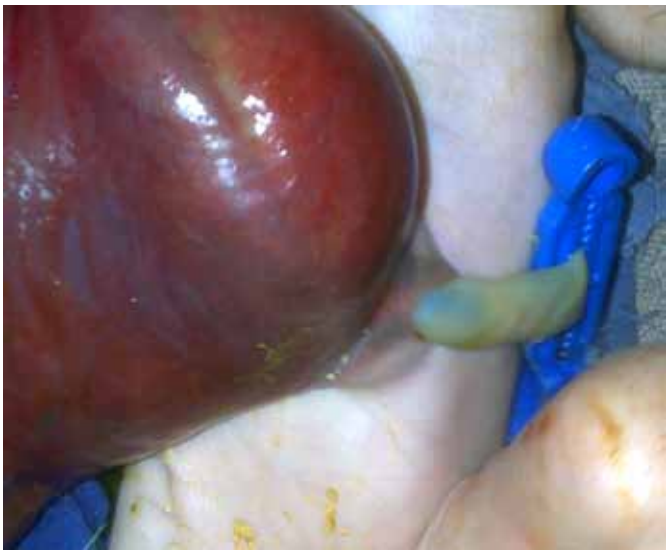

Figure 2. Photograph showing omphalocele sac with the contents and cord attached at the centre.

bowel or the liver outside of the abdomen). Other investigations done are amniocentic analysis to confirm chromosomal abnormalities, Ultrafast fetal MRI for central nervous system anomalies and Fetal echocardiogram to confirm heart defects. Delivery of foetuses with antenatally diagnosed abdominal wall defects varies. How et al[10] report that they can be safely delivered by the vaginal route and elective caesarean delivery be opted for obstetric indications. Fifty percent of babies born with omphalocele have genitourinary, brain, lung, spine, heart and gastrointestinal defects. The outcome depends upon the size of the herniation and the presence of defects.

In small omphaloceles primary repair is done by putting the herniated organs back into the abdominal cavity and closing. In Giant omphalocele with liver and other organs, a staged repair (Schuster procedure) is done where a mesh is sewn to the fascia and the muscle on each side of the omphalocele defect and the two pieces of mesh or silastic sheet are then sewn together over the defect creating a silo and the omphalocele sac remains intact, the mesh/silo is tightened over days or weeks to return the organs into the cavity and mesh is removed and closure done. This process provides time for the abdominal wall to stretch so as to enclose the viscera and promotes lung growth and expansion. Sometimes the protruding organs may prevent closure when a technique called "paint and wait" is employed, where the sac covering the omphalocele is painted with an antibiotic cream and covered with elastic gauze. This promotes epithelialization. Wrapping and component separation as done in adults for large ventral hernias may be tried. This provides extra length and mobility by separating the fascia and muscles, but there is a possibility of a hernia or defect that may develop at the site of repair.[7,8,9] Infants may have common complications such as feeding difficulties, bowel obstruction and gastroesophageal reflux and should be monitored.

Conclusion

Antenatal sonogram is a must for all pregnant women. They need to be counseled about the risks, outcome and possible modes of delivery and the need for surgical management well in advance when omphalocele is detected.

End Note

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Conflict of Interest: None Declared

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