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A case of traumatic rupture of a giant omphalocele and liver injury associated with transverse lie and preterm labor

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ABSTRACT

Perinatal omphalocele rupture is a rare occurrence. We present a case of a baby delivered at 35 weeks with a known giant omphalocele, transverse lie, and the omphalocele downward in the birth canal who suffered rupture of the omphalocele and liver injury around the time of delivery. The pregnancy was complicated by one day of preterm labor, preterm premature rupture of the membranes, and the omphalocele was the presenting part. Despite pulmonary hypertension, rupture of the omphalocele, and a significant liver injury, individualized management with decompression of the liver hematoma allowed successful early closure with mesh followed by delayed reconstruction.

Omphalocele is a rare congenital anomaly affecting approximately 1 in 5000 live births [1,2]. In contrast to gastroschisis, omphalocele is associated with congenital anomalies and increased mortality [3,4]. Surgical management of an intact omphalocele is performed after initial resuscitation and stabilization of the infant and workup of other congenital anomalies has been completed [5]. However, rupture of the omphalocele is associated with increased mortality risk and must be addressed more urgently [6]. With loss of the sac covering, the infant experiences increased insensible fluid losses and the exposed organs such as the liver and spleen are more susceptible to injury [7]. We present a case of a 35-week gestation infant with a ruptured giant omphalocele and subsequent subcapsular liver hematoma (Grade III) after preterm labor. A staged management strategy was successful in achieving a successful outcome.

1. Case report

A 38-year-old G5P0040 with a history of three previous spontaneous abortions and one previous ectopic pregnancy presented at 34 weeks and 6 days gestation with a complaint of abdominal pain. The baby was known to have a giant omphalocele. Given the complex history, she was transported by air to a tertiary care obstetrics facility. On presentation, the mother had stable vital signs and review of systems was otherwise negative except for contractions. However, preterm premature rupture of membranes (PPROM) was confirmed by sterile vaginal exam that showed copious meconium-stained fluid at cervical os. The fetal heart rate was reassuring. Previous prenatal ultrasound confirmed a back-up transverse lie with the unruptured giant omphalocele as the presenting part. The decision was made to proceed with delivery because meconium was noted in the amniotic fluid; caesarian delivery was felt to be necessary due to the transverse lie of the child.

Intraoperatively, the uterus was opened. The back was up and the omphalocele was in the birth canal. Upon extraction, it became apparent that the omphalocele was ruptured. The delivery was difficult due to the impaction of the omphalocele deep into the maternal pelvis. After delivery, the infant was promptly intubated and suctioned without evidence of meconium in the endotracheal tube. Apgar scores were 2 at 1 min, but 9 at 5 min. Positive-pressure ventilation was given via the endotracheal tube and spontaneous respiration began soon thereafter.

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Examination revealed the omphalocele to be ruptured with a large defect at the 7 o'clock position with the umbilical cord still directly attached to the omphalocele sac (Fig. 1; video link). The abdominal wall opening was small (4 cm) yet the entire gastrointestinal tract including the liver, spleen, and pancreas was exposed. A large subcapsular hematoma involving the entire anterior surface of the liver, approximately 70% of the fully exposed liver (grade III liver injury).

The baby was transferred to the neonatal intensive care unit (NICU) on conventional ventilation and was started on empiric antibiotics. Immediate pediatric surgical consultation was requested for management of the ruptured giant omphalocele and liver injury. After establishment of intravenous access, the surgical team was mobilized to the bedside as the degree of pulmonary hypertension prevented transfer to the operating room. Surgery began approximately 2 h after delivery.

The omphalocele sac was densely adherent to the liver, so no attempt was made to separate the sac from the liver due to the risk of bleeding. The entirety of the small and large bowel was able to be reduced into the abdomen given the neonate's adequate abdominal domain. Most of remaining sac was then taken off of the skin. Because of the large size of the subcapsular hepatic hematoma, it was felt that the safest course of action was to place a Silastic silo over the abdominal contents. This would also allow a complete cardiac workup and evaluation of the pulmonary hypertension. The majority of the silo was subfascial with the exception of the cephalad portion adjacent to the liver. The hepatic veins were very superficial (close to the anterior abdominal fascia) so the fascia could not be freed at this point. The silo was carefully sewn to the fascia using interrupted PDS sutures at the cephalad portion and running sutures was used to attach the remainder of the silo. After surgery, the patient remained in extremely critical condition from respiratory distress solely secondary to pulmonary hypertension.

After the neonate stabilized over the next 36 h, the ventilator settings were low and there was no evidence of hepatic dysfunction or coagulopathy, so decision was made to attempt mesh closure of the abdominal defect. Initial attempts were made to gently reduce the liver into the peritoneal cavity, however, this was limited by the size of the subcapsular hematoma. Therefore, the omphalocele defect was extended inferiorly in an attempt to provide adequate space. Despite this the liver was still unable to be completely reduced into the abdomen. Finally, the subcapsular hematoma was decompressed allowing complete reduction. The portion of the omphalocele sac adherent to the liver was not removed due to risk of bleeding.

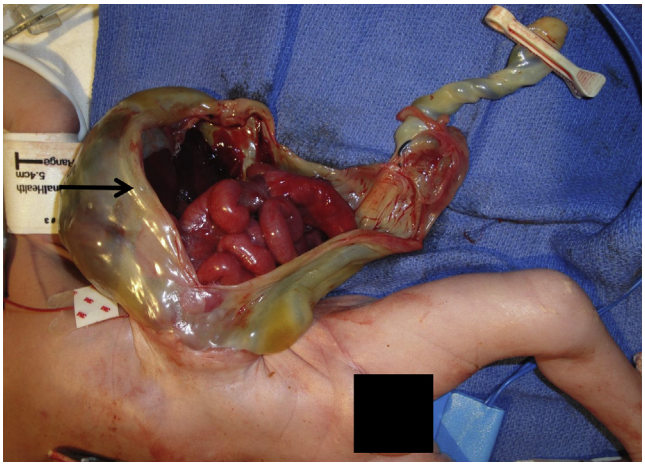

Fig. 1. Omphalocele rupture with umbilical cord in continuity with the omphalocele sac with subcapsular hepatic hematoma (arrow).

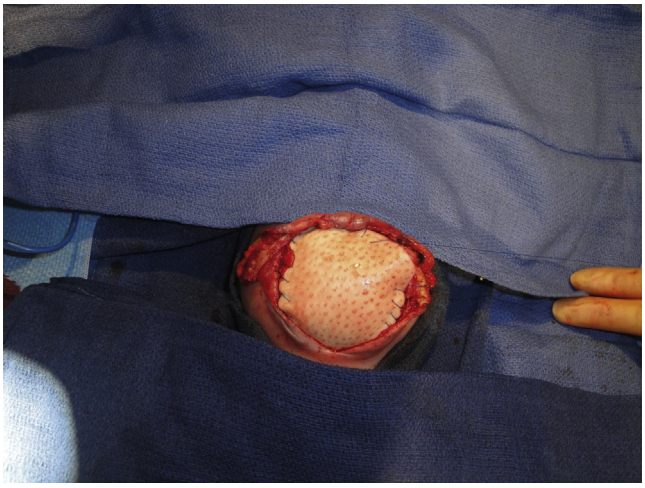

Fig. 2. Strattice™ closure of abdominal wall defect.

Following decompression of the hematoma, the liver was completely reduced into the abdomen. The fascia was inadequate for primary closure, so the decision was made to use a biologic mesh for coverage (Strattice™ Reconstructive Tissue Matrix, LifeCell Corporation, Bridgewater, NJ). Skin flaps were made after fascial mobilization and the mesh was sewn onto the fascia with 3-0 polypropylene sutures (Prolene®, Ethicon US, LLC, Somerville NJ). The mobilized skin flaps were closed over the mesh (Figs. 2 and 3). The patient tolerated the procedure well and was returned to the NICU.

Continued workup for associated anomalies revealed an interrupted inferior vena cava (IVC) with azygous continuation, atrial septal defect, and moderate pulmonary hypertension on echocardiography. The child was started on total parenteral nutrition (TPN) postoperatively. He was extubated on postoperative day 3 and nasogastric feedings with breastmilk were trialed unsuccessfully. Due to prolonged ileus, TPN was required for 41 days until full nasogastric feeds could be obtained. He also had bilateral inguinal hernias that were repaired at 2.5 months of age. The child was discharged to home 3 months postoperatively on sildenafil for pulmonary hypertension. A laparoscopic gastrostomy was ultimately done at 13 months of age due to persistent oral feeding

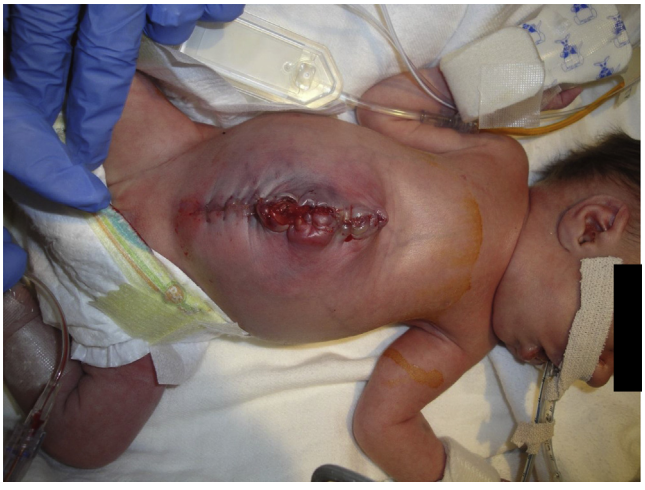

Fig. 3. Postoperative image of complete skin closure over the Strattice™ mesh.