Emergent Excision of a Massive Prenatally Diagnosed Sacrococcygeal Teratoma in a Newborn

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Case presentation:
A 24 year old G1P0 woman presented at 26 weeks gestation. Her fetus was diagnosed with a massive sacrococcygeal teratoma (SCT). Fetal MRI revealed a female fetus with a three vessel umbilical cord, polyhydramnios, and an SCT with cystic and solid components measuring 9cm in the craniocaudal dimension by 9.2 cm in the transverse dimension. There was no internal component, and no evidence of obstruction of either the gastrointestinal or genitourinary tracts. The fetus was followed closely with 13 echocardiograms in a 4 week period. They revealed no structural cardiac defects, mild cardiomegaly, and good ventricular function, with greatly increased but stable biventricular cardiac output of approximately 850 mL/kg/min (normal range 553±153 mL/kg/min). (1) There was dilation of both the inferior vena cava (IVC) and descending aorta. The IVC increased in size from 4.3mm to 6.3 mm over the next 4 weeks. Normal IVC diameter is 2.9 to 4.1 mm in fetuses 21-28 weeks gestation. (1) Despite these findings demonstrating a hyperdynamic state and greatly increased blood flow to the SCT, there was no evidence of hydrops fetalis.

Maternal polyhydramnios was treated with serial amnioreductions, but her uterus became more active, and she went into preterm labor at 30 weeks gestation. The baby was delivered by urgent cesarean section and taken immediately to an adjacent operating room which was prepared to receive the baby with both anesthesia and neonatology teams present for resuscitation. Estimated weight of the baby was 1.5 kg, Apgar scores were 1, 2, and 4 at 1, 5, and 10 minutes. Peripheral intravenous access was obtained. Intubation was difficult, and was successful only after multiple attempts by several experienced physicians with a 2.0 ETT. Chest X-ray was consistent with severe RDS. She received surfactant, atropine, and several doses of THAM and sodium bicarbonate. An umbilical venous line was placed, but the tip was in the liver; placement of the umbilical arterial line was not successful, so a radial arterial line was placed. Over the course of the resuscitation, changing color of the SCT suggested hemorrhage into the tumor. The blood gases improved slightly after surfactant, and excision of the tumor commenced. Tourniquets were placed at the base of the tumor, and dissection proceeded distal to the tourniquets. Visible blood loss did not seem excessive, but the hemoglobin levels did drop to 2-4 gm/dL, suggesting hemorrhage into the tumor and need for more compression of the arterial supply to the SCT. Precipitous drop in blood pressure and heart rate ensued, requiring intermittent chest compressions, red blood cell (RBC) and fresh frozen plasma (FFP) transfusion, and infusions of dopamine and epinephrine. (Figure 1)
The tumor was eventually debulked and the skin was closed. Additional time was spent in continuing resuscitation of the infant, who was in a state of disseminated intravascular coagulation as evidenced by bleeding from multiple non operative sites. At the end of surgery, after over 4 hours in the operating room, this 1.5 kg infant received 290 mL RBC, 200 mL FFP, 200 mg calcium gluconate, and 72 mcg epinephrine. These were in addition to continuous infusions of dopamine and epinephrine. Estimated blood loss was 250 mL, or two blood volumes.

The infant was transported to the neonatal intensive care unit, where over the course of 3 hours, she continued to ooze from the operative site. More RBC’s, cryoprecipitate, platelets, recombinant factor VII, calcium, and epinephrine were given. She was placed on a high frequency oscillating ventilator. Twice during this resuscitation, the infant became bradycardic and required chest compressions with epinephrine and atropine. Her prothrombin time was 31s, activated partial thromboplastin time >150s, INR 3.25, and fibrinogen 761 mg/dL. Her hemoglobin levels dropped from 17 to 6 mg/dL in 45 minutes. Pulmonary hemorrhage was also noted, and after discussion with the family, morphine and midazolam were given for comfort and care was withdrawn.

Autopsy revealed multifocal organ congestion and hemorrhage, intrapelvic hemorrhage, skin hemorrhage and petechiae, and both pleural and pericardial effusions. The SCT measured 14x 12x 8 cm and weighed 965 gm with large blood filled cysts. There were mesenchymal, neuroglial, and epithelial cell types represented, with no evidence of malignancy.

Discussion

Prenatally diagnosed SCT has a much higher mortality rate than SCT diagnosed in the neonatal period. (1,2) Evaluation and comprehensive care for these complex patients requires an experienced center and a multi-disciplinary team with the ability to perform fetal MRI, fetal echocardiography, and frequent maternal evaluation. Close communication between obstetricians, pediatric surgeons,
anesthesiologists, and neonatologists is vital. Two operating rooms with multiple teams of physicians and nurses are needed. One OR must have an anesthesia, surgical, and nursing team for the maternal surgery, and another OR is needed for the newborn with separate anesthesia, surgical, nursing and neonatology teams. The infrastructure to easily perform in utero surgery, cesarean section, and neonatal surgery must be in place, as management continually evolves with the progress of the disease.

References:
2. Benachi et al., J Pediatr Surg. 2006 Sep;41(9):1517-21