Abdominal (subdiaphragmatic) Bronchopulmonary sequestration

A GxxxPxxx pregnant woman presented for the routine second-trimester fetal anatomy assessment at xxx weeks of gestation. On the scan, a well-circumscribed, homogeneous, hyperechoic solid mass with small cystic areas within it was noted in the left suprarenal region. Focused assessment showed a shifting in the mass during fetal breathing movements, the so called 'sliding sign'. Colour Doppler imaging revealed a feeding artery from the abdominal aorta in several conventional and non-conventional views. The mass also showed venous drainage into the azygous vein. Anterior displacement of the stomach was noted, but there were no other masses or cysts within the fetal abdominal cavity.

The fetal diaphragm appeared intact. The heart was evaluated and appeared unremarkable. The echogenicity of the lungs appeared normal, and there were no other masses seen within them. The ipsilateral kidney and adrenal gland, as well as the spine, showed no additional abnormalities.

This was a female (or male) fetus with an estimated fetal weight of XXX grams, plotted on the XXX centile. No abnormal fluid collections, such as hydrothorax, pericardial effusion, or ascites, were present. The amniotic fluid volume was normal.

We explained to the parents that, although the differential diagnosis is broad, including neuroblastoma, adrenal haemorrhage, mesoblastic nephroma, retroperitoneal teratoma, and lymphangioma, the findings were consistent with the diagnosis of an isolated intra-abdominal pulmonary sequestration.

The risk of chromosomal abnormalities in such cases is not increased above the baseline. However, if additional abnormalities are detected, an amniocentesis with microarray is advised, as the risk of underlying chromosomal abberations may rise in these cases.

Serial ultrasounds have been scheduled to monitor fetal growth and well-being, as well as the size of the mass and to screen for fetal hydrops.

We have explained to the parents that hydrops is a sign of a poor prognosis, and if it develops, we will need to reformulate the plan. In complicated cases with hydrothorax or hydrops earlier than 32-34 weeks of gestation fetal intervention with percutaneous laser ablation of the feeding vessel, thoracocentesis, or thoraco-amniotic shunt placement can be considered, but this mostly applies to thoracic lesions. These interventions are rarely - if ever – needed with subdiaphragmatic pulmonary sequestration. In such complicated cases, if the gestational age is at least 32-34 weeks, delivery after corticosteroid-induced lung maturation can be considered.

Delivery planning will be in conjunction with the neonatologists and pediatric surgeons. Large lesions merit delivery at a tertiary center with neonatal intensive care facilities. Intra-abdominal BPS itself is not an indication for early delivery, and C-section should be performed only for the usual obstetric indications.

Most infants with small lesions are asymptomatic at birth. However, all cases should be evaluated by postnatal imaging, including cases that appeared to have resolved during the antenatal period, as postnatal CT scan/MRI are more sensitive than prenatal ultrasound for detecting small lesions.

The prognosis is generally favorable, with a survival rate of more than 95%. Cases with associated congenital anomalies and hydrops may have worse outcomes.

Management after birth can either be conservative therapy or surgical resection, which remains the treatment of choice, because complete excision eliminates the potential risks of complications, such as infection, torsion, or malignant degeneration.

We conveyed to the couple that intra-abdominal BPS is thought to be sporadic, with a very low risk of recurrence in subsequent pregnancies.