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Independent case reports

Fetal lung interstitial tumor: a cause of late gestation fetal hydrops

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Abstract Most fetal lung masses present by mid gestation, grow during the canalicular phase of lung development (18-26 weeks of gestation), and plateau in growth or shrink after 26 weeks of gestation. We describe the unique case of a fetal lung mass presenting at 37 weeks of gestation with hydrops and fetal heart failure. The late growth of this lesion and resultant hydrops prompted resection as part of the ex utero intrapartum treatment. Histopathology revealed a rare, recently described fetal lung interstitial tumor. This case demonstrates that a subset of fetal lung masses may continue to grow later in gestation and emphasizes the need for late gestation imaging and close follow-up in this patient cohort.

Fetal lung anomalies are increasingly identified during obstetric ultrasound evaluation. The differential diagnosis of a fetal lung mass commonly includes congenital cystic adenomatoid malformation (CCAM; or congenital pulmonary airway malformation), bronchopulmonary sequestration, bronchial atresia, bronchogenic cyst, and congenital lobar emphysema [1-4]. These lesions grow during the canalicular phase of lung development (18-26 weeks of gestation), and plateau in growth, shrink, or even involute after 26 weeks [5,6]. The vast majority of fetal lung masses are benign; however, some lesions are malignant pleuropulmonary blastoma [7]. We describe the rare case of a fetal lung interstitial tumor (FLIT) presenting in the third trimester. The unique late growth of this lesion and resultant hydrops prompted resection as part of the ex utero intrapartum treatment (EXIT) (EXIT-to-resection).

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1. Case report

A 37-year-old primigravida carrying a singleton fetus was referred to our fetal center at 36 weeks and 6 days of gestation with a large fetal lung malformation. The pregnancy had been otherwise uneventful. She had an ultrasound done at 22 weeks of gestation that was normal; however, at 36 weeks, a discrepancy between gestational age and fundal height prompted a second ultrasound. At that time, a solid, hyperechoic right-sided lung mass was identified and the patient was referred for further evaluation. Ultrafast fetal magnetic resonance imaging confirmed a 5.7 × 4.9 × 4.0-cm well-circumscribed, right-sided lesion exerting a marked mass effect with direct compression of the inferior vena cava (IVC), leftward displacement of the heart, eversion of the diaphragm into the abdomen, and compression on surrounding normal lung (Fig. 1). Fetal hydrops was present as defined by large ascites and pleural effusion, and the CCAM-volume ratio [8] was calculated to be 2.1. Fetal echocardiogram showed compression of the IVC and a large azygous vein venting venous return to the superior vena cava. The left atrium and ventricle (LV) were under filled, and there was depressed left ventricular systolic function. The LV myocardial performance index was quite elevated (0.93), indicating both diastolic and systolic LV dysfunction. Because of the presence of fetal heart failure and high likelihood of immediate respiratory insufficiency, an EXIT procedure was recommended to permit the safest delivery of the fetus. After a multidisciplinary discussion and nondirective counseling, the family elected for delivery by an EXIT procedure with resection of the thoracic mass. At 37 weeks and 1 day of gestation, an EXIT procedure was performed using the technique as previously described [9]. An endotracheal tube was placed, but ventilation was held initially to avoid high peak inspiratory pressures in the fetus' severely compressed lungs. Because of the severe compression of the IVC, a cutdown was performed to place a right internal jugular central venous catheter. Fluid was administered to optimize the fetus' volume status and avoid hypotension secondary to thoracic decompression after resection of the mass. After optimal ventricular filling was confirmed by continuous fetal echocardiography, a right posterolateral thoracotomy was performed through the fifth intercostal space. The large right lower lobe mass was partially delivered out of the chest, permitting ventilation of the compressed but otherwise normal remaining lungs with an appropriate rise in oxygen saturation. A decompressive paracentesis was performed to facilitate delivery of the fetus, and the umbilical cord was divided after 81 minutes on uteroplacental support. The child was then taken into an adjacent operating room to complete the right lower lobe resection while the mother's hysterotomy was closed. The child had an uneventful hospital course. His ventilator requirement was minimal as he was extubated on day of life (DOL) 5, and he was discharged home on DOL 21. Histopathologic examination revealed a 5.0 × 4.6 × 3.5-cm solid mass within the right lower lobe diagnosed as a FLIT, a rare, recently described lesion [10] (Fig. 2). After a multidisciplinary discussion of the case, it was decided that no adjuvant treatment was indicated. A chest x-ray and physical examination were performed for surveillance every 3 months for the first 2 years of life and once a year thereafter. At 5-year follow-up, the child had remained asymptomatic without respiratory tract infection requiring hospitalization, asthma, chest wall deformity, spinal abnormality or evidence of recurrent disease; and he had met or exceeded all developmental milestones.

2. Discussion

The natural history of fetal lung masses is variable. Most of these lesions are asymptomatic in the prenatal period; however, a small percentage of lung masses will grow at a rate faster than the fetal chest. In such cases, rapid growth of the mass leads to compression of thoracic structures, fetal

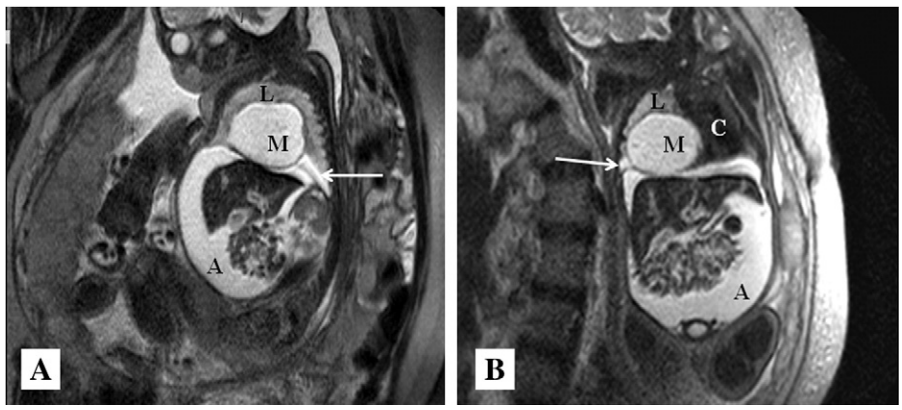

Fig. 1 Sagittal (A) and coronal (B) ultrafast magnetic resonance imaging images at 36 weeks of gestation showing a fetal lung mass (M) in the setting of cardiac compression (C), pleural effusion (arrow), and ascites (A) compressing the lung (L) and everting the diaphragm into the abdomen.