An Unusual Antenatal Presentation of a Mature Pericardial Teratoma Masquerading as Congenital Pulmonary Airway Malformation

Vivek Krishnan[1], Mohan K Abraham[2], Ajay Abraham[2], Indu Ramachandran Nair[3], Basil Mathews[1]*

[1]Division of Fetal Medicine and Perinatology, Amrita Institute of Medical Sciences, Kochi, Kerala, India, [2]Department of Pediatric Surgery, Amrita Institute of Medical Sciences, Kochi, Kerala, India, [3]Department of Pathology, Amrita Institute of Medical Sciences, Kochi, Kerala, India

Abstract

We report an antenatal presentation of a huge pericardial mature teratoma that was referred as congenital pulmonary airway malformation (CPAM) in the late third trimester of pregnancy. Initial ultrasound evaluation revealed a huge predominantly cystic lesion with mixed echogenicity in the left hemithorax. A provisional diagnosis of pleural tumor was considered in view of previous scans at 20–28 weeks being normal and associated pleural effusion. Magnetic resonance imaging of the fetus reported the lesion to be CPAM which was supported by postnatal computed tomographic imaging done on day 2 of life. However, intraoperatively, the lesion was found to be of pericardial origin which on subsequent histopathological examination was confirmed to be mature teratoma. We recommend considering potential differential diagnosis other than CPAM, especially when the lesion is found for the first time in the late third trimester.

Keywords: Cardiac tumor, congenital pulmonary airway malformation mature teratoma, pericardial teratoma

Introduction

Pericardial teratomas are benign, rare, primary pericardial tumors comprising ectoderm, mesoderm, and endoderm.[1,2] These are the second most common cardiac tumors, second only to rhabdomyomas.[3]

Case Report

A 30-year-old primigravida was referred at 37 gestational weeks with newly detected congenital pulmonary airway malformation (CPAM). Her midtrimester anomaly scan as well as a growth scan at 32 weeks were reported normal. Sonographic examination revealed a predominantly cystic lesion with mixed echogenicity in the left hemithorax, measuring 7.2 cm × 3.4 cm × 5.5 cm. The left lung was not seen separately. The largest cyst measured 8 mm in diameter, and the volume of the lesion was 70 cc. The mediastinum was shifted to the right with moderate pleural effusion on the left side [Figure 1a and b]. The left cardiac border was clearly made out and was seen separate from the lesion. No other abnormality was detected [Video 1]. A provisional diagnosis of a pleural tumor was made in view of previous normal studies and isolated pleural effusion on the left side. Fetal echocardiography showed a structurally normal heart with normal biventricular heart function. A fetal magnetic resonance imaging (MRI), however, reported the lesion as a Type II CPAM involving the left lung upper lobe [Figure 1]. CPAM volume ratio calculated at this point was 2.09 cm^2.

A full-term neonate of birth weight appropriate for gestational age was born by cesarean delivery in double setup for ex-utero intrapartum treatment (EXIT) procedure as respiratory distress and failure were anticipated in view of extrinsic lung compression. The neonate was stable at room air. Computed

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Address for correspondence: Dr. Basil Mathews, Division of Fetal Medicine and Perinatology, Amrita Institute of Medical Sciences, Ponekkara P.O., Kochi - 682 041, Kerala, India. E-mail: dr.basil_meck@hotmail.com

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tomography (CT) imaging on day 2 of life showed a multicystic lesion of 6.3 cm × 4 cm occupying the upper two-thirds of the left hemithorax causing a significant mediastinal shift to the right [Figure 2]. Systemic feeder arteries were seen to supply the lesion. A differential diagnosis of a hybrid CPAM involving the left upper lobe and pleuropulmonary blastoma was given. Complete resection of the tumor after vascular separation from the arch of the aorta was achieved on day 4 of life [Figure 3a]. Intraoperatively, the lesion was found to be of pericardial origin with left phrenic coursing over the pericardium covering the tumor. Postoperatively, the baby developed eventration of the left hemidiaphragm possibly due to the left phrenic nerve injury, which was subsequently repaired by thoracoscopic plication. Pediatric assessment at 3 months after birth was unremarkable.

Histopathological examination revealed a multicystic neoplasm, predominantly lined by tall columnar mucinous cells with interspersed goblet cells positive for cytokeratin 20 and negative for cytokeratin 7. Islands of mature cartilage, smooth muscle bundles, and seromucinous glands with few stratified squamous epithelium-lined cysts were also seen [Figure 3b]. Thus, it showed mature elements derived from endoderm, mesoderm, and ectoderm, with no immature components, favoring a pericardial mature teratoma.

Discussion

Pericardial teratoma frequently presents as a multicystic lesion on the right anterior border of the heart with associated pericardial effusion and a vascular pedicle from the root of the aorta.[2,3] Although excellent prognosis (92%) is documented for postnatal surgical resection, up to 77% of the prenatally detected cases end up with hydrops owing to the cardiac compression due to the location of the tumor, tamponade, or pericardial effusion requiring interventions in the form of pericardiocentesis, pericardial to amniotic cavity shunt, fetal surgery, early delivery, or EXIT.[1,3-5] Paralysis of the left diaphragm was reported previously in a 32 weeks preterm newborn after teratoma resection which was repaired on day 18 of life with no long-term respiratory problems.[6] In a series of six cases,[7] two fetuses with early presentation succumbed in the midtrimester due to rapid progression and cardiac tamponade, one had a successful fetal surgery, whereas three cases were closely monitored and delivered for tumor resection with good outcome in two. Furthermore, two cases were managed by EXIT procedure with tumor resection, one at 31 weeks and another at 36 weeks with good neonatal outcome.[5,8] Gestation at diagnosis and presence of nonimmune hydrops remain the two major prognostic markers.

Foregut malformations are by far the most common congenital thoracic lesions and are the first differential diagnosis for a macro or microcystic lesion unless proved otherwise. Pulmonary vasculature in CPAM differentiates it from bronchopulmonary sequestration (BPS) which is supplied by a systemic arterial feeder. BPS is further classified into intralobar, when the venous return to the pulmonary vasculature, and extralobar, when it is to the systemic system. It is difficult to differentiate these from the primary pulmonary neoplasms, which include pulmonary pleuroblastoma, fetal interstitial tumor, congenital myofibroblastic tumor (CMFT), and congenital or infantile fibrosarcoma. Since the congenital developmental lung lesions are identifiable from the early second trimester, if a lesion is not identified in the midtrimester scan, the lesion is more likely to be true neoplasm. This, however, does not hold good for CMFT which is even seen as early as the early second trimester. The rapid progression to hydrops and presence of pericardial effusion are the two distinct features that sometimes help in differentiating pericardial teratomas from other thoracic lesions.[9]

Our case was unique in the unusual location of the tumor, mimicking its origin from the upper lobe of the left lung, and

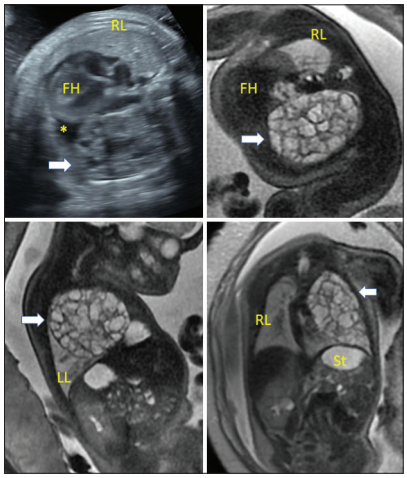

Figure 1: Prenatal ultrasound and fetal magnetic resonance images of the pericardial tumor (white arrow) showing the lesion to arise from the left lung (labeled as LL) occupying the left side of the thorax with pleural effusion (asterix). RL: Right lung, FH: Fetal heart, St: Stomach, LL: Left lung

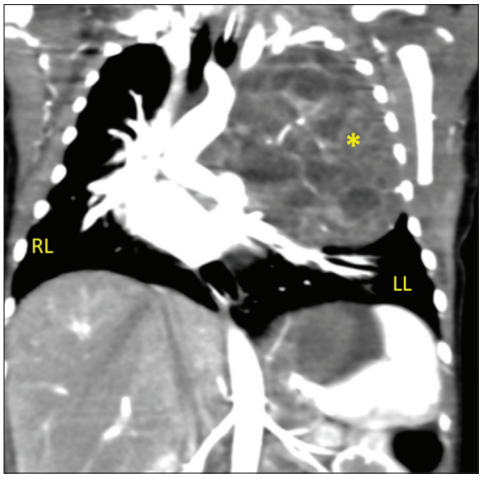

Figure 2: Postnatal computed tomographic imaging - Coronal view of the fetal thorax showing the multicystic lesion (asterix) occupying the left hemithorax with vascular supply from the arch of the aorta. RL: Right lung, LL: Left lung

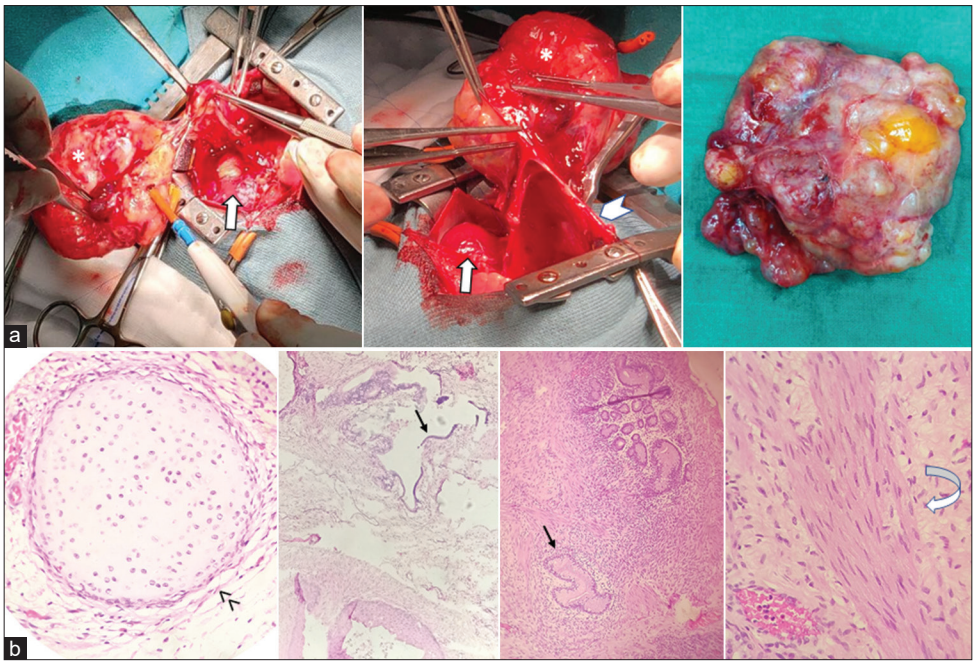

Figure 3: (a) Pericardial origin (arrowhead) of otherwise isolated tumor (asterix). FH (white arrow) is seen within the thoracic cavity free from tumor. Gross specimen showing a 7 cm × 6 cm × 3 cm nodular gray-white tissue, (b) histopathology: multicystic neoplasm with cysts lined by intestinal-type mucinous columnar and stratified squamous epithelium (black arrow). Intervening areas show cartilage (double arrowhead), smooth muscle (curved white arrow), lymphatic, and myxoid tissue with no immature neuroepithelial tissue. FH: Fetal heart

it is very late presentation, the latest reported teratoma hitherto being at 34 weeks. Neither prenatal fetal MRI nor postnatal CT imaging could correctly identify the lesion.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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