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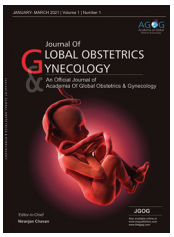
Case Report



Prenatal Diagnosis of Congenital Pulmonary Airway Malformation/Bronchopulmonary Sequestration Hybrid Lesion: A Case Report

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ABSTRACT

Congenital lung malformations are wide spectrum diseases that goes together with broncho-pulmonary dilatation. Most of the controversies are due to the lack of evidence on almost all prenatal and postnatal aspects of congenital pulmonary airway malformation (CPAM) and bronchopulmonary sequestration (BPS). CPAM is the most common congenital lung disease in the fetus. CPAM and BPS are the lung masses that do not function as normal lung tissue, CPAM has a vessel from the pulmonary artery and BPS has a vessel from the aorta that feeds them. Hybrid lesions defined as lung lesions can growth rapidly and otherwise look and behave similar to CPAM but the feeding vessel from the aorta. The prognosis is worse in hybrid lesions due to large-sized mass with feeding vessels from systemic circulation and increased risk of hydrops. We present a case of CPAM/BPS hybrid lung lesion that involved entire left lung.

Key words: Bronchopulmonary sequestration, Congenital pulmonary airway malformation, Feeding vessel, Prognosis

INTRODUCTION

Congenital pulmonary airway malformation (CPAM) is a rare fetal abnormality with a reported incidence of 1:25,000–1:35,000.[1] It is the most common congenital lung disease in the fetal period. The underlying pathophysiological mechanisms are not clear, but broncho-pulmonary developmental deficiencies probably occur at the early stages of embryogenesis, which is responsible for these lung masses.[2] No genetic cause is detected. It is a solid or cystic lung mass with arterial supply from the pulmonary artery. These lesions are often diagnosed during routine prenatal ultrasound. CPAM may spontaneously regress (disappearing CPAM), but large lesions can compromise alveolar growth and development by compressing adjacent normal lung tissue. Macro-cystic CPAM is characterized by 1 or more cysts >5 mm, often multiple cysts of varying sizes and may have single large cyst and the cyst's borders are poorly defined. Micro-cystic CPAM is characterized by cysts <5 mm, uniformly echogenic, and well-defined masses. It usually presents as incidental finding in second-trimester ultrasound. Most fetuses with CPAM are euploid. Because isolated forms are not associated with aneuploidy, there is no indication for fetal karyotyping in these cases. Majority remain stable or regress in utero and it has excellent prognosis without hydrops even if large at diagnosis. The important complications related to CPAM are polyhydramnios, compression of thoracic structures, mediastinal shift, and hydrops fetalis.[2] The presence of early-onset fetal hydrops has been associated with a poor prognosis.[3] Clinicians should consider these potential events and early detection to prevent unfavorable pregnancy outcomes. Most fetuses have good perinatal outcome. Delivery should be at the tertiary center due to risk for neonatal complications, including air-trapping and pneumothorax and large lesions may require extracorporeal membrane oxygenation. Post-natal work-up of all lesions is needed even if regressed in utero, for detection of small masses and definite

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diagnosis.[2] Post-natal complications include recurrent infection and small risk for developing malignancy.

Bronchopulmonary sequestration (BPS) also known as accessory lung is a bronchopulmonary foregut malformation with estimated incidence of 0.1–6.4%.[4] It does not connect to the trachea-bronchial tree and receives its arterial blood supply from the systemic circulation. The abnormal lung tissue is located within the visceral pleura of a pulmonary lobe in the intra-lobar variety, whereas the extra-lobar form has its own visceral pleura. No genetic cause is detected and the incidence of chromosomal abnormalities and genetic syndromes is not increased. It is usually hemogenously echogenic mass that located on the basis of the left lung. Spontaneous regression is common in BPS. BPS is an associated anomaly in 15–30% of patients with CDH.[5] Hybrid lesions consider when systemic vessel supplies cystic lung mass.

CASE PRESENTATION

A 43-year-old pregnant woman, primigravid, IVF with egg donation, was referred to our center due to fetal lung lesion in anomaly ultrasound in gestational age of 19 weeks. She had no history of medical disease and drug usage during pregnancy. Her nuchal translucency ultrasound and first-trimester fetal aneuploidy screening were low risk for common trisomies (T13, 18, 21). In our ultrasound examination, we detect a lung mass (2.78 cm × 2.39 cm) in the entire left lung in four-chamber view that shifted the heart to the right [Figure 1]. It had microcystic changes in the superior lobe of the left lung [Figures 2 and 3] and had echogenic solid appearance in the inferior lobe of the left lung [Figures 4 and 5]. The involvement of the entire left lung was more pronounced in the coronal and parasagittal views [Figures 6 and 7]. We detected a large feeding vessel from the aorta to the lung mass in the parasagittal view [Figures 8 and 9]. This was also evident in the coronal view [Figure 10]. No feeding vessels were seen from the pulmonary artery to the mass [Figure 11]. In the examination of the heart, there was a shift of the heart to the right [Figure 12], but the ventricles and inferior vena cava (IVC) were not compressed [Figures 13 and 14]. The structure of the heart was normal and no obvious abnormalities were seen in the other organs. The diaphragm was intact, but the pressure effect on the left diaphragm was detected due to the left lung mass [Figures 15 and 16]. We detected the shifting of mediastinal vessels and trachea in the axial view [Figures 17 and 18]. The stomach was present with normal size and we had no evidence of esophageal dilatation and polyhydramnios. All of these indicate no esophageal obstruction following the secondary mediastinal shift to the lung mass. Middle cerebral artery peak systolic velocity showed no evidence of anemia and fluid accumulation was not seen in abdominal, pericardial, pleural, and subcutaneous compartments. The CPAM volume Ratio (CVR) was 0.62 (2.78 × 2.39 × 2.91 × 0.52/16.1), mass to thoracic ratio was 0.57 [Figure 19] and cardiomediastinal shift angle (CMSA) was 30.7 degree [Figure 20]. Because this finding was isolated, karyotype was not determined by amniocentesis and the patient underwent follow-up after consultation. Due to the absence of hydrops and CVR <1.6, treatment with betamethasone was not performed.

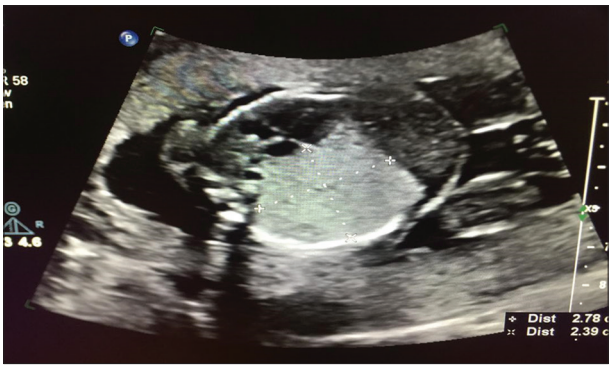

Figure 1: Echogenic lung lesion in left lung in four-chamber view that shifted the heart to the right

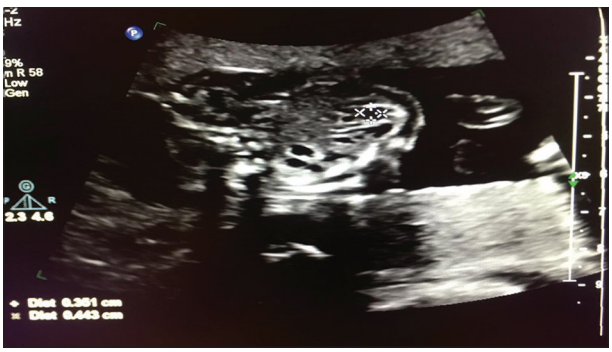

Figure 2: The lung mass shows microcystic changes in the apex of the superior lobe of the left lung in axial view

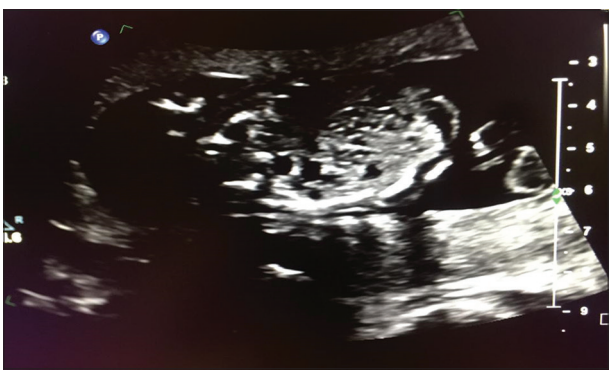

Figure 3: The lung mass shows microcystic changes in the middle part of the superior lobe of the left lung in axial view

DISCUSSION

The development of the respiratory system begins at about 4 weeks of gestation. During fetal life, the lung develops as a liquid-filled organ. This liquid is produced by the fetal lung and leaves via the trachea from whether it is either swallowed or enters the

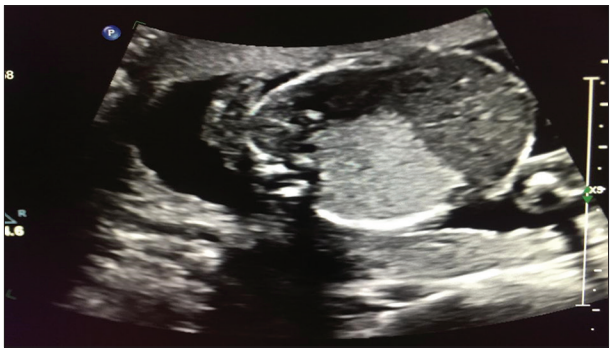

Figure 4: The lung mass shows echogenic solid appearance in the inferior of the lobe of the left lung in axial view

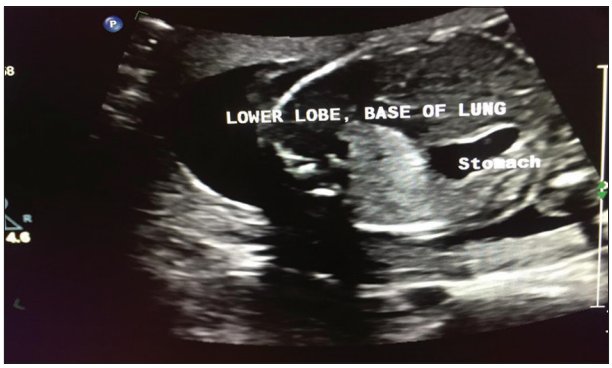

Figure 5: The mass in the inferior lobe of the left lung is extended to the base of the lung in axial view

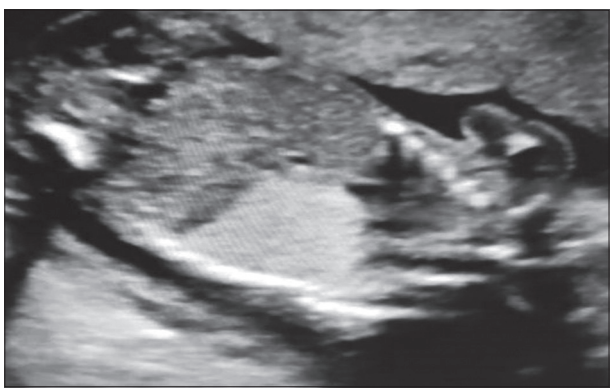

Figure 6: Coronal view shows involvement of the entire left lung

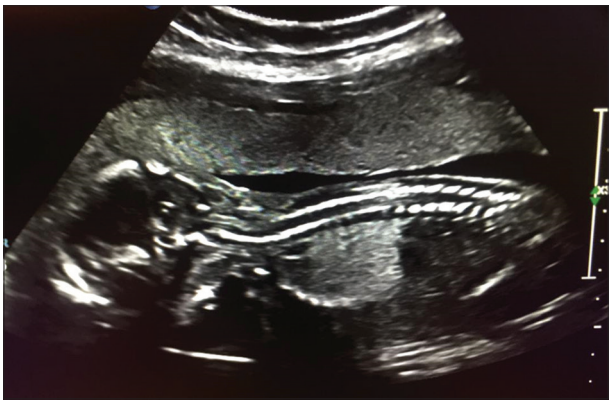

Figure 7: Parasagittal view shows a mass in entire left lung

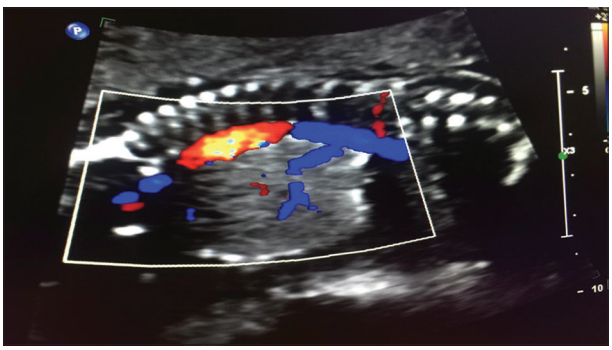

Figure 8: A large feeding vessel from the aorta to the lung mass in parasagittal view

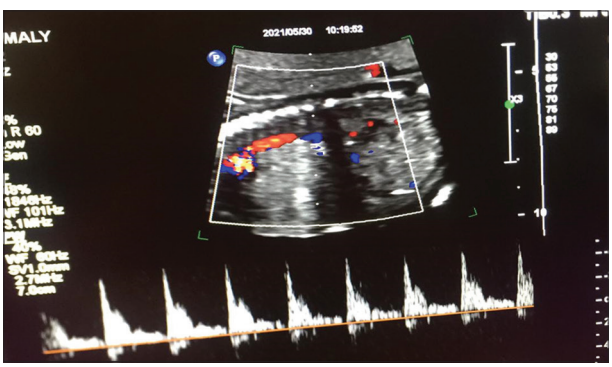

Figure 9: Peak systolic velocity of feeding vessel in Doppler ultrasound that resembles an aortic wave

amniotic sac. Fetal lung liquid plays a crucial role in the growth and development of lungs by maintaining them in distended state and stimulating their growth.[6]

The examination of the fetal chest in the first trimester includes the assessment of the right and left lung, the bony and cartilaginous thoracic cage, the diaphragm, and the heart and surrounding vasculature. In the normal fetus, the lungs appear slightly more echogenic than the liver and cardiac muscle. At the four-chamber view plane, the right and left lungs are seen and the rib cage assessed. Comprehensive evaluation of the lungs in axial views requires the assessment at the level of the four-chamber view and superiorly into the upper mediastinum at the three-vessel trachea view. Assessment of cardiac position and axis in the chest is helpful in the identification of lung abnormalities. The right and left parasagittal views of the fetal chest are important for the assessment of individual lung lobes, the diaphragm, and the rib cage. The transvaginal approach improves the visualization of all chest structures due to higher resolution. Clear visualization of the lungs in ultrasound can be achieved from about the 12th week of gestational onward.

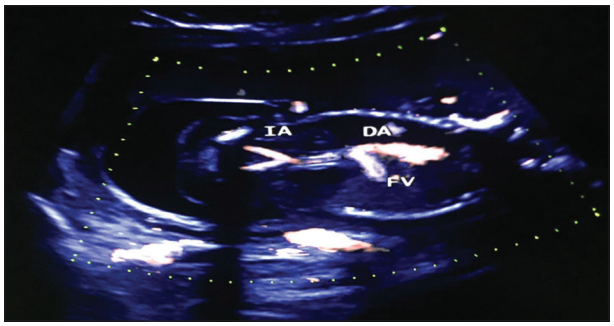

Figure 10: Feeding vessel from the aorta to the lung lesion in Doppler ultrasound in coronal view. (DA: Descending Aorta, FV: Feeding Vessel, IA: Iliac Arteries)

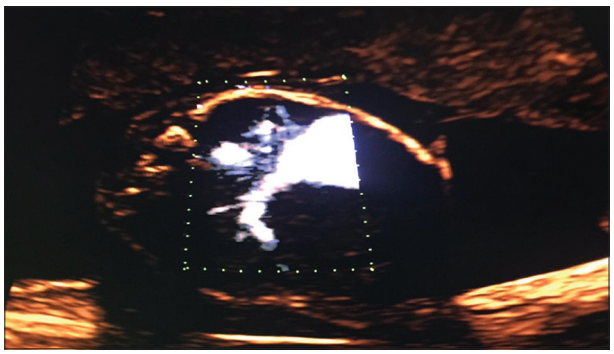

Figure 11: No feeding vessel is seen from the pulmonary artery to the lung mass in Doppler ultrasound in axial view

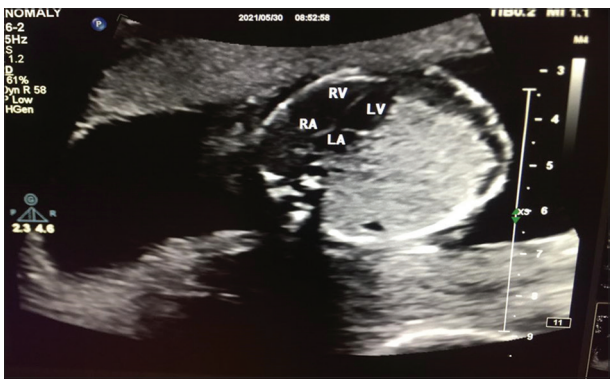

Figure 12: Dextroposition of the heart due to pressure effect of the left lung mass in axial view. (RV: Right Ventricle, LV: Left Ventricle, RA: Right Atrium, LA: Left Atrium)

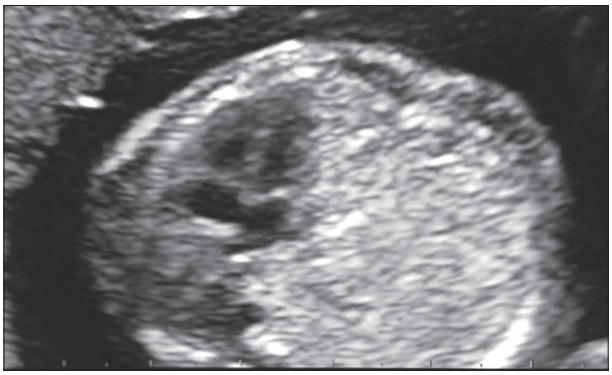

Figure 13: The heart cavity is not compressed in four-chamber view

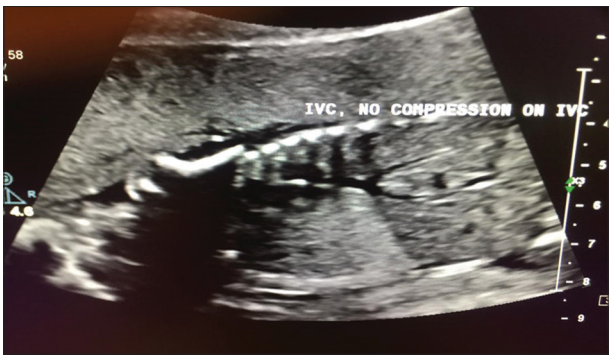

Figure 14: The IVC is not compressed in coronal view. (IVC: Inferior Vena Cava)

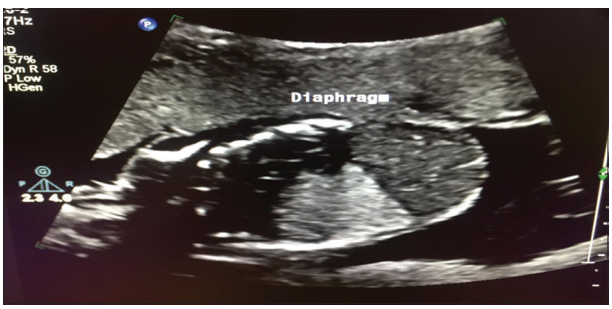

Figure 15: Diaphragm is intact. It is not accompanied by a diaphragmatic defec

CPAM

Size is variable, usually contained within a single lobe (our case: 2 lobes of left lung), can be massive (heart is often displaced to the opposite chest wall, as in our case). Morphology varies from solid-appearing (micro-cystic) (as in our case) to complex cystic mass (macro-cystic), 95% are unilateral (as in our case) and affect only one lobe, but can arise in all lobes, infrequently multiple lobes. No side predilection was seen in CPAM and it is more common at lung bases. Ultrasound classification includes microcystic and macrocystic. Our case had a microcystic type CPAM. The greatest growth occurred in microcystic type between 20 and 26 weeks.

For determination of intrauterine and postnatal fetal outcomes, calculate CVR: CVR >1.6 indicates increased risk of developing hydrops and fetal demise. A novel measurement of mediastinal shift in cases of CPAM and its relation with adverse perinatal outcomes and hydrops is CMSA.[7] CMSA was not correlated with lesion laterality or gestational age or CPAM subtypes. Median CMSA is 13.8 in microcystic, 26.5 in mixed microcystic, and 30.2 in macrocystic types. CVR and CMSA were found to have a strong, positive correlation with one another. The ability of CMSA to predict adverse perinatal outcome is optimized at a cutoff of 34.3

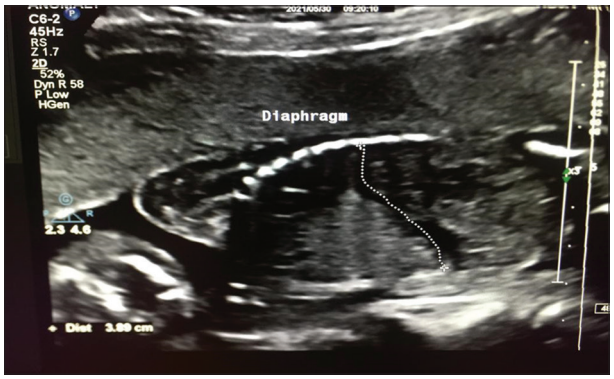

Figure 16: This image shows the compressive effect of the left lung mass on the left side of diaphragm

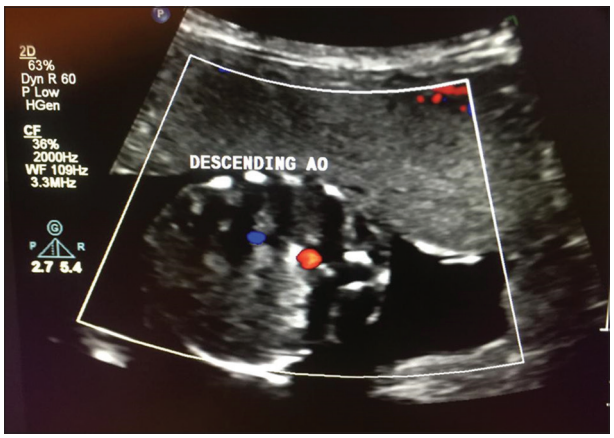

Figure 17: Left descending aorta shift due to mass pressure, indicating mediastinal shift in the axial view. (Ao: Aorta)

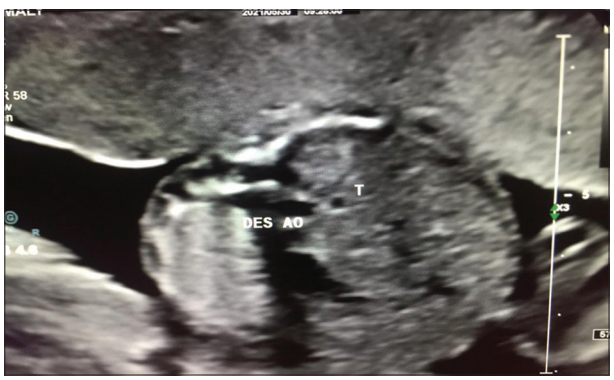

Figure 18: Tracheal and aortic shifts due to mass compression in the axial view. (DES AO: Descending Aorta, T: Trachea)

degrees with a sensitivity of 72% and specificity of 85%.[7] In our case, CVR was 0.62 and CMSA was 30.7 degrees. Furthermore, the ratio between the transverse diameter of the mass and the transverse diameter of the thorax, measured on the axial view in the four chamber view defined as mass/thoracic ration, >0.51 is associated with adverse outcomes. This ratio was 0.57 in our case. If hydrops has not developed by approximately 28 weeks, it is unlikely to develop since CPAM growth plateaus by the end of the second trimester and the fetal thorax continues to increase.

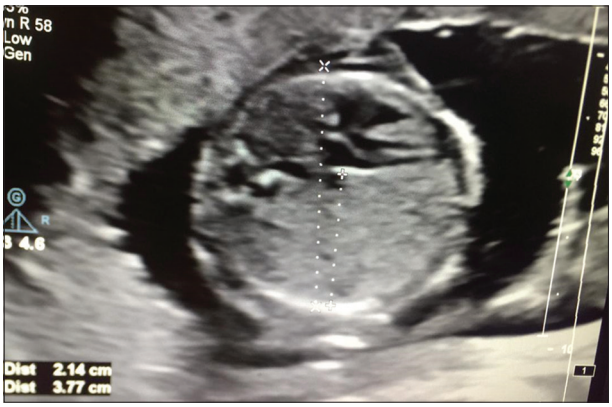

Figure 19: Mass to thoracic Ratio is 0.57

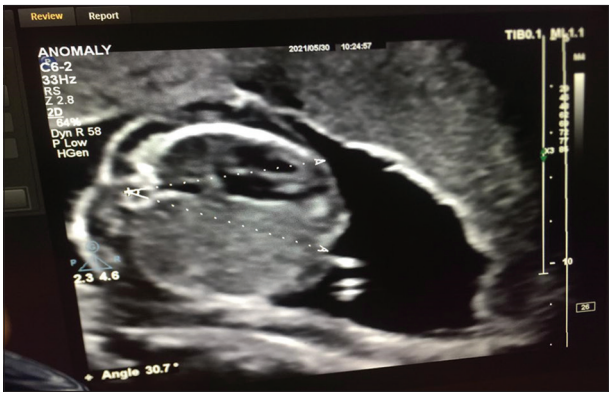

Figure 20: Cardiomediastinal shift angle is 30.7 degree

BPS

Extra-lobar BPS is often detected in prenatal ultrasound and becomes symptomatic early in life. Fetal intra-lobar sequestration is extremely rare, more commonly identified later in life secondary to recurrent infection. Best diagnostic clue in ultrasound is solid lung mass (our case: microcystic) with arterial supply from the aorta (as in our case). It is generally small to moderate size, rarely can fill entire chest. Location in 85–90% of cases is supra-diaphragmatic, 10–15% sub-diaphragmatic, and 90% left-sided (as in our case). In morphology, pleural investment results in well-marginated mass, triangular or lobar shape. Unilateral pleural effusion presents in a number of cases and may cause tension hydrothorax. Prognosis is excellent when it is an isolated finding. Severe BPS is associated with swallowing impairment, poly-hydramnious, cardiac, and mediastinal shift causing cardiac failure and non-immune hydrops. The prognosis of untreated severe BPS is highly unfavorable, with a 95% risk of intraiuterine fetal death.[8]

Lung masses can cause mediastinal shift. Hence, Rotation of the heart, compression of IVC, DV, and heart can lead to hydrops. The esophagus can become compressed and obstructed, resulting in dilation of proximal of the esophagus, a small stomach,

polyhydramnious and the ipsilateral diaphragm may be flattened. Causes of hydrops in lung lesions are: Impaired venous return, heart failure due to cardiac compression, compression of lymphatic vessels (Cisterna chili, thoracic duct), and high cardiac output heart failure in BPS. Hydrops has not been described after mass growth plateaus.

Prenatal treatment options such as maternal administration of steroids, minimally invasive procedures, or open fetal surgery can decrease the mass effect and prevent the progression of complications and fetal outcomes.[9] In macrocystic lesions, a thoracoamniotic shunt under guide of ultrasound may be helpful in decompressing the cyst.[10] Maternal betamethasone treatment has been suggested to have beneficial effects on large microcystic CPAMs.[11-13] It is the only medical option in the management of CPAM in hydropic fetuses or in cases determined to be at risk for developing hydrops because of CVR >1.6. Betamethasone is administered in two doses of 12 mg intramuscularly in mother 24 h apart. The fetus is followed weekly and depending on the circumstances, this betamethasone course can be repeated up to three times. In cases of BPS, treatment by laser ablation of the feeding vessel may be needed in complicated fetuses with hydrops or anemia in the prenatal period. There are a few indications for intervention before birth. The indications depend on size, solid or cystic features, and the development of hydrops.[14]

A lung lesion that has features of both CPAM and a BPS is called a hybrid lesion. In these cases, imaging studies will show an abnormal arterial blood vessel feeding the mass as well as cysts within the lesion. This type of lung lesion usually has a poorer prognosis due to large-sized mass and blood supply from the systemic circulation. Therefore, their early detection and accurately follow-up in terms of progression to anemia and hydrops are crucial. Due to the effectiveness of treatments such as betamethasone, early treatment can prevent adverse fetal outcomes.

CONCLUSION

With the advancement of ultrasound, lung lesions have become easier to diagnosis prenatally. Early prenatal and postnatal diagnosis, accurate follow-up, and treatment if indicated will help improve outcomes. Due to rarity of these lung lesions, little is known about how to manage it. Therefore, cases need to be reported. We suggest setting up a patient registration system to find out more about this fetal lung lesions.

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