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RARE CASE OF CONGENITAL PULMONARY AIRWAY MALFORMATION – A CASE REPORT

Radiology

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ABSTRACT

Background: CPAM is a benign cystic or adenomatous terminal bronchiole disorder that is caused by a respiratory tract malformation.
Clinical description and Investigations: Our case is that of a 19-year-old pregnant female patient came for routine ANC at 20.4 weeks of gestation. The fetus was diagnosed with congenital pulmonary airway malformation. An enlarged right lung with a single large cyst of size 34x18x25 mm with multiple tiny cystic areas , a compressed left lung, and a compressed and displaced heart were observed. **Conclusion:** Early diagnosis of CPAM is based on an ultrasound examination of the pregnant woman. Most of CPAM lesions can be managed with the proper assessment, diagnosis, and surgical interventions.

KEYWORDS

Congenital Pulmonary Airway Malformation (CPAM),Broncho pulmonary sequestration , Cystic degeneration

INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM) is a fetal pulmonary development abnormality caused by airway dysgenesis. It is characterized by cystic or adenomatous lesions in terminal bronchioles. The severity of a case is proposed by the size of the mass, degree of mediastinal shift, and presence of hydrops and polyhydramnios. CPAM is a rare pathology, that is often accidentally diagnosed during routine antenatal ultrasounds. Congenital pulmonary airway malformation in the past was known as Congenital Cystic Adenomatoid Malformation (CCAM) as it is a congenital disorder of the lung just similar to bronchopulmonary sequestration. Usually, an entire lobe of the lung is replaced by a non-working cystic area of abnormal lung tissue in this condition and this abnormal tissue never functions as normal lung tissue. In this case report, we present a distinct case of CPAM, grayscale ultrasound findings are discussed here.

The cause of congenital pulmonary airway malformation is still unknown. Its incidence is approximately1 in every 30000 pregnancies. Malignant transformation can occur within a cyst with a small documented risk. Hence, early diagnosis and surgical resection are necessary to prevent grave complications. Congenital pulmonary airway malformation is also known as congenital cystic degeneration which is a rare abnormality of fetal lung development. In this disease, there is a benign cystic or adenomatous lung tumor that grows on terminal bronchioles and restricts their function. According to various research data, the incidence of CCAMs is reportedly between 1:8300 and 1:35000 live births.[1] This congenital pulmonary airway malformation mainly affects males but still, there is no evidence that sex has any effect on the probability of development of this disease in individuals.[2] Based on clinical and pathological features it is classified into 5 major types. Most CPAM lesions are manageable with the proper assessment, diagnosis, and surgical interventions.[3]

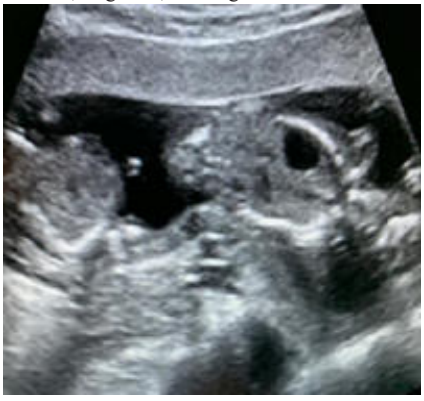

Figure 1. Coronal Section of Fetus showing enlarged right lung with a cyst and a compressed Contralateral Lung and Heart.

PRESENTATION OF CASE

A 19-year-old pregnant female patient came for routine ANC ultrasound at the Department of Radio diagnosis, PGIMS, Rohtak. The patient with her first pregnancy was seen for prenatal care and her first ultrasound at 20.4 weeks of gestation. The fetus was diagnosed with congenital pulmonary airway malformation. An enlarged right lung with a single large cyst of size 34x18x25 mm with multiple tiny cystic areas (Figure 1, 2, 3; Coronal, axial and sagittal section respectively), a compressed left lung, and a compressed and displaced heart were observed.

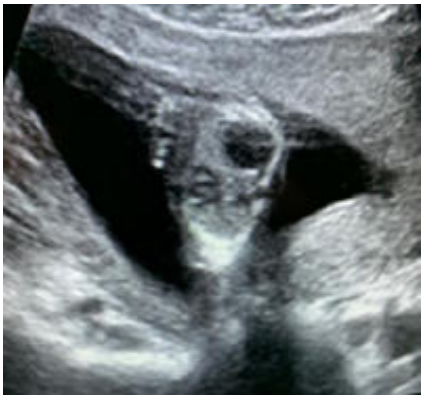

Figure 2. Axial Section of Fetus Showing the Cyst and Compressed Contralateral Lung and Heart.

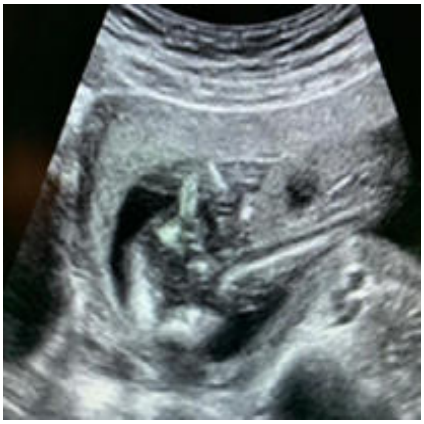

Figure 3. Sagittal Section of the Fetal Thorax Showing a Cystic Lesion.

DISCUSSION

CPAM is a rare condition distinguished by immature lung which is malformed and has a cystic appearance. New-borns and stillborn infants are affected by CPAM, and unusually children beyond infancy are known to be affected. It is a hamartomatous lesion that is usually symptomatic in the first few days of life. The babies with CPAM present as neonates with severe, progressive respiratory distress due to cyst expansions. Hydrops may be present.[4] The exact etiology of CPAM is not known, although it is considered to be hamartomatous malformation and abnormal proliferation of pulmonary tissue at different sites. Sometimes CPAM may communicate proximally with airways, although this form is abnormal. Most CPAMs are supplied by the pulmonary artery and their drainage is by the pulmonary veins, except hybrid lesions which have a systemic blood supply. Fetal ultrasound in large cyst CPAM will show multiple variable-sized anechoic spaces with echogenic soft tissue. On fetal MRI (Magnetic Resonance imaging), large cyst CPAM manifests as hyper intense unilocular or multilocular cysts with well-defined walls on T2-weighted images. On the postnatal chest, radiography variable density in the region of the mass with mediastinal shift is seen. In the neonatal period, a large cyst CPAM is seen as a round soft-tissue mass gradually filled with air, with delayed clearance of fetal lung fluid from those cysts through the abnormal airway. Air-fluid levels may be visualized. There may be a solitary well-defined air-filled cyst with imperceptible walls or multiple cysts of varying size. On gross pathological examination of an excised large cyst CPAM, we can appreciate a large (> 2cm) cyst and occasionally multiple or multiloculated cysts, often with small cysts that will be merging with the adjacent lung. Microscopically, the cysts have ciliated epithelium as their lining and may contain mucin forming epithelium as well. This highlights the embryologic origin of the lung-foregut derivative.[5] On prenatal ultrasound, a small cyst CPAM is an echogenic mass with multiple small cysts whereas a microcystic (predominantly solid-appearing) CPAM is homogeneously echogenic. There may be a mediastinal shift and consequent polyhydramnios and hydrops in fast-growing CPAM. The indicators of poor prognosis are large lesions, bilateral lung involvement, and hydrops. It has been proposed that the designation of this lesion "CCAM" be changed to "CPAM" to reflect the fact that lesions described as cystic are present in only 3 of the 5 types and "Adenomatoid" only in one type (Type I). CPAM is classified as follows:[6]

CPAM Type 0 - Acinar dysplasia/agenesis is a rare malformation largely incompatible with life. Lungs are small and firm with a diffusely granular surface.

CPAM Type I - It accounts for nearly 65 % of cases. It is operable with a good prognosis. Grossly, the lesion is predominantly cystic type (measuring 3 - 10 cm in diameter) surrounded by smaller cysts.

CPAM Type II - accounts for 10 % - 15 % of cases and is mainly seen in the first year of life. It has a poor prognosis because it is frequently associated with other congenital anomalies. Grossly lesion is composed of medium-sized cysts measuring 0.5 to 2.0 cm in diameter that are evenly distributed and blend with the adjacent normal parenchyma. CPAM Type II has been noted in nearly 50 % of cases of extra-lobar sequestration.

CPAM Type III - It is infrequent and accounts only for about 5 % of cases. It consists of a small cystic or solid type exclusively seen in the first few days to months of life with characteristic male preponderance. It is seen to be associated with maternal polyhydramnios, and fetal anasarca commonly. So it has a high mortality rate. Also, there is increased alpha-fetoprotein levels in the second trimester. Grossly cysts are small measuring less than 0.2 cm in diameter, affecting large bulky mass involving an entire lobe or even an entire lung.

CPAM Type IV - It is a hamartomatous malformation of the distal acinus and accounts for 10 % - 15 % of cases with an age range of newborn to 4 years. This lesion involves a single lobe.[5]

It's peculiar to note that the infants who are diagnosed to have CPAM lesions on antenatal ultrasonography are later found to be asymptomatic at birth. Most of this CPAM is associated with other congenital anomalies.[7] Congenital lobar emphysema can be differentiated from CPAM as it contains Broncho-vascular marking which extends to the periphery. In certain cases, when a macro cystic form of CPAM appears, there is a risk of pulmonary hypoplasia and the development of fetal hydrops due to compression of developing lungs by large cysts.[8] Fetuses suffering from CPAM can have various survivability prognoses and it depends on the following factors: the size of the growth, degree of mediastinal displacement, hydrops, and polyhydramnios. Good, moderate, and poor prognosis groups can be identified based on these factors. However, as the number of studies examining CPAM is growing around the world, it is being discovered that many cystic growths regress on their own in the third trimester of pregnancy.[8] Expectant management is useful for the non-hydropic fetus, although the survival of fetuses with hydrops may improve only with fetal intervention. If the mass is predominantly a single cyst, thoraco-amniotic shunting may be an option. After 32 weeks of gestation, surgical resection of the mass may be possible while the fetus is maintained on placental circulation, using an ex-Utero Intrapartum Treatment ("EXIT") procedure. If the condition is not recognized antenatally on ultrasound, it is often detected when a baby of fewer than 2 years of age presents with respiratory difficulty or subsequent pulmonary infection. Symptomatic infants are treated with surgical resection, where lobectomy or segmental resection is done.[5]

CONCLUSION

CPAM is a benign cystic or adenomatous terminal bronchiole disorder that is caused by a respiratory tract malformation. Its early diagnosis is based on an ultrasound examination of the pregnant woman. Most of CPAM lesions can be managed with the proper assessment, diagnosis, and surgical interventions.

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