Case Report

ISSN (O): 2347-338X; ISSN (P): 2347-3371

Antenatal Diagnosis of Pulmonary Sequestration: A Case Report

Gaurav Kaushik, Rohan Shelar, Pratik Bangde

Consultant Radiologist, Department of Radiodiagnosis, DY Patil School of Medicine, Navi Mumbai, Maharashtra, India.

Abstract

Pulmonary sequestration is an uncommon medical condition in which a lung tissue is formed, which does not perform any function and also is not either the other anatomical structures of the main lung tissue, not its blood supply. A 28 years old antenatal mother, visited our department for routine anomaly scan in her second trimester. Transabdominal ultrasound examination revealed a well-defined echogenic mass involving the left hemi-diaphragm extending upwards towards the lung. Colour Doppler study revealed feeding artery which originated from the descending thoracic aorta. This confirmed our diagnosis of pulmonary sequestration. A male baby, weighing 3150 grams was delivered vaginally. High Resolution Computed Tomography showed evidence of a large well defined soft tissue mass involving the left pleural cavity with pleural effusion, and shift of mediastinum and heart towards the right side. In addition, there was an evidence of a feeder artery which originated from the descending thoracic aorta.

Keywords: Extralobar pulmonary sequestration, antenatal diagnosis, ultrasonography, Computed Tomography.

Corresponding Author: Gaurav Kaushik, Consultant Radiologist, Department of Radiodiagnosis, DY Patil School of Medicine, Navi Mumbai, Maharashtra, India.
E-mail: gauravkaushik_7@yahoo.co.in

Received: 30 November 2021 **Revised: 08 January 2021** **Accepted: 15 January 2021** **Published: 19 June 2021**

Introduction

Pulmonary sequestration is an uncommon medical condition in which a lung tissue is formed, which does not perform any function and also is not either the other anatomical structures of the main lung tissue, not its blood supply. This medical condition was first observed and documented by Pryce in 1946 and defined as an abnormal artery from the aorta supplying a bronchopulmonary mass or cyst which is dissociated from the normally connected bronchial tree.[1] Pulmonary sequestration more commonly is observed in the lower left lung. It has been suggested that its origin occurs from a lung bud which is located caudally, which transversed caudally which happened at the time the lung was developing along with the esophagus. There are two forms of pulmonary sequestration: intralobar (ILS), which the cystic mass is surrounded by normal lung tissue & extralobar (ELS) which has a separate pleural layer. Thus ILS and ELS characterization depends on whether the formation of the cystic mass happens before or after the formation of pleural layer.[2] In this case report we describes case of ELS which was diagnosed on routine antenatal ultrasound examination in our department.

Case Report

A 28 years old antenatal mother, visited our department for routine anomaly scan in her second trimester. The pregnancy was unremarkable till date. Transabdominal ultrasound examination revealed a well-defined echogenic mass involving the left hemi-diaphragm extending upwards towards the lung. On further detailed examination of the mass, there was a solid well-defined echogenic mass with underlying collapse and associated pleural effusion with resultant shift of mediastinum and heart towards right side [Figure 1]. Based on the examination, a differential diagnosis of pulmonary sequestration and congenital cystic adenomatoid malformation was made. This was followed by colour Doppler study, which revealed feeding artery arising from the descending thoracic aorta [Figure 2]. This confirmed our diagnosis of pulmonary sequestration. A grave prognosis was discussed with the parents. The mother was followed closely during the remaining part of the pregnancy. Serial ultrasound examinations were performed and massive polyhydramnios developed. A male baby, weighing 3150 grams was delivered vaginally in our hospital. Severe respiratory distress was noted at birth due to which the baby was intubated and admitted to neonatal intensive care unit. APGAR score was 4 and 6 at 1 and 5 minutes respectively. Despite aggressive ventilatory support, chest x-

ray found small lung volumes. High Resolution Computed Tomography was done immediately. There was evidence of a large well defined soft tissue mass involving the left pleural cavity with pleural effusion, and shift of mediastinum and heart towards the right side. In addition, there was an evidence of a feeder artery originating from the descending thoracic aorta [Figure 3]. Mass resection was considered but not performed due to hypoplastic lungs. The neonate expired at 18 hours of age.

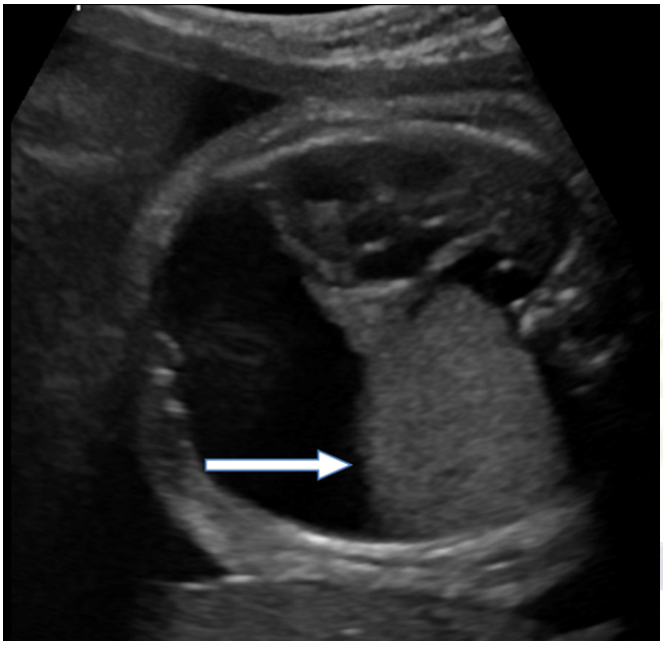

Figure 1: Ultrasound showing a solid well-defined echogenic mass with underlying collapse and associated pleural effusion with resultant shift of mediastinum and heart towards right side.

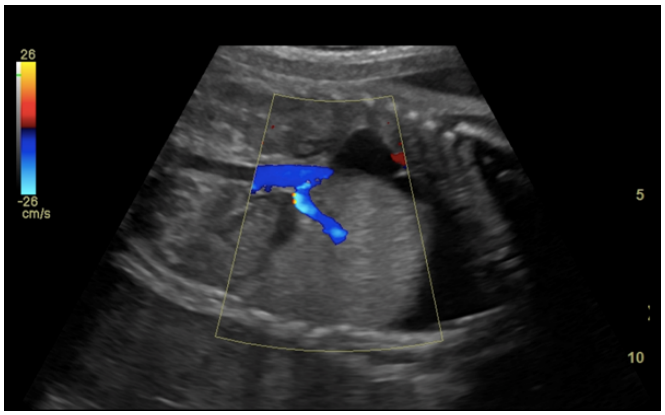

Figure 2: Colour Doppler study revealing feeding artery arising from the descending thoracic aorta.

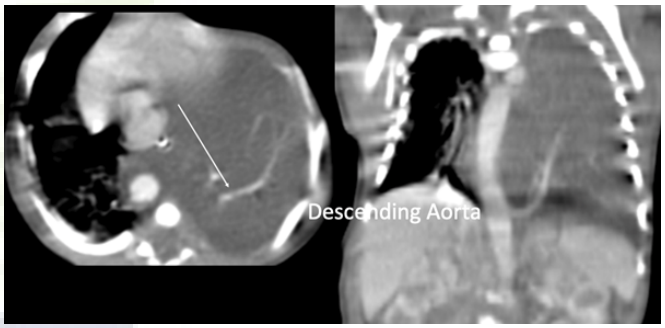

Figure 3: High resolution CT showing a feeder artery arising from the descending thoracic aorta.

Discussion

Pulmonary sequestration, also known as bronchopulmonary sequestration (BPS), is a cystic mass of non-functioning pulmonary tissue that does not communicate with the tracheobronchial tree. In addition, the blood supply is not from pulmonary artery but from a feeding artery which comes from the descending aorta. Two forms of pulmonary sequestration are typically seen: ILS which exists within the normal lung parenchyma, and ELS which has separate pleural layer and is thus completely separated from the normal lung parenchyma. In a few cases, this cystic mass may exist outside the thoracic cage as well. BPS is a common congenital abnormality of the lung tissue, which is only second to congenital cystic adenomatoid malformation (CCAM). It has been observed that the among the cases of BPS diagnosed, 35% to 50% are comprised of ELS.[3] This variation in the prevalence of ELS in the prenatal and postnatal period could be due to the fact that ILS could happen postnatally.[4] ELS was observed to be four times more common among males, but recent data suggest a similar prevalence in either gender.[5]

ELS can be diagnosed in the prenatal period as a mass in the intrathoracic or abdominal mass, with typical appearance on ultrasonography and color Doppler. The dimensions of the cystic mass and the presence of hydrothorax affect the overall prognosis of the patients. In many cases, BPS can resolve spontaneously, thus resulting in favorable prognosis. This was observed when consecutive ultrasound images were obtained.[6] It has been suggested that thrombosed feeding artery may be responsible for the spontaneous resolution. On ultrasound examination, BPS appears as a solid or cystic mass with increased echogenicity. In majority of cases, BPS is detected during the second trimester of pregnancy, after which it can increase in size and regress during the later stages of pregnancy. In cases who have BPS mass below the diaphragm,

it may be mistaken for a kidney mass. A feeding artery helps in the diagnosis. It originates from the descending aorta and enters the mass, as it does not receive any supply from the pulmonary artery.

One important differential diagnosis is that of CCAM. It may appear similar to BPS on ultrasound examination; however, absence of a feeding artery helps to differentiate it. Interestingly, in approximately half of all cases, CCAM and BPS may exist together. In these cases, feeding artery may be present.[7] Other differentials will include bronchial atresia, which involve the all of the lung tissue, appears hyperinflated, has high echogenicity and an everted diaphragm. In Scimitar syndrome the lung is hypoplastic, of some part of the right lung with venous drainage commencing in the inferior vena cava. The ELS which is present below the diagphragm may be confused with a kidney mass like a blastoma or neuroblastoma. Presence of a feeding artery helps in differentiating. Moreover, BPS may appear as a highly echogenic mass on ultrasound.

It has been observed that majority of the cases diagnosed with BPS resolve in the antenatal period of pregnancy. Thus experts advise to wait for it resolve on its own, if BPS is not compromising the fetus. It can be done by regular follow up of the antenatal mother and looking for ultrasonographic signs of fetal complications. If the fetus develops hydrops fetalis during the initial part of the third trimester, an intervention is indicated. Resection of the BPS (lobectomy) in the fetus is indicated, in which approximately half of all cases can survive.[8] But the risks involved and the expertise required to perform an intrauterine surgery makes it a rarely performed procedure. The survival of the neonates born with BPS depend on the size of the cystic mass and the location of it. When the mass is too large, it can compress on the main lung tissue and result in hypoplasia and hydrops fetalis. These cases very rarely survive and intrauterine death is the expected outcome. In a few cases, some neonates may only show signs of distress, lung infection, failure to feed or circulatory failure.

Conclusion

Anomaly scan is an important component of antenatal care. We were able to detect this rare condition antenatally using advanced imaging modalities. Although the baby did not survive, we were able to counsel the parents regarding the grave prognosis of the baby.

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How to cite this article: Kaushik G, Shelar R, Bangde P. Antenatal Diagnosis of Pulmonary Sequestration: A Case Report. Asian J. Med. Radiol. Res. 2021;9(1):14-16.

DOI: dx.doi.org/10.47009/ajmrr.2021.9.1.3

Source of Support: Nil, **Conflict of Interest:** None declared.