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CASE REPORT

PRENATAL DIAGNOSIS OF EXTRAPULMONARY SEQUESTRATION BY ULTRASOUND - A CASE REPORT

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ABSTRACT

A 24 years old primigravida underwent a routine ultrasound scan at 26 weeks of gestation; which showed a single normal growing foetus with intrathoracic mass on the left side of thoracic cage along with left displacement of cardiac apex. The mass was closely associated with lower portion of left lung. The extralobar sequestration was diagnosed which was further confirmed by colour Doppler during foetal period and by computed tomography and x-ray chest postnatally. Surgical excision was done.

INTRODUCTION

Extra pulmonary sequestration [EPS] is a rare congenital malformation (Chih-Hung Lin *et al.,* 2013). It is the cystic mass of non-functioning primitive lung parenchyma that does not communicate with the trachea-branchial tree and pulmonary vessels (Shah *et al.,* 2003). It is separated from normal lung and receives its blood supply from systemic circulation and has its own pleura (Shah *et al.,* 2003; Debra *et al.,* 1992) and has anomalous venous return (Kyriakos *et al.,* 2010). The prevalence of extra pulmonary sequestration is about 0.15 -6.4% of all pulmonary malformations (Chih-Hung Lin *et al.,* 2013; Debra *et al.,* 1992) and the prevalence is greater in males than females with a ratio about; M: F is 4:1 (Shah *et al.,* 2003; Debra *et al.,* 1992). Among this group of anomalies more than 90% are found in thorax and less than 10% under the diaphragm (Chih-Hung Lin *et al.,* 2013). Hence early diagnosis is essential to prevent hydrops foetalis and pulmonary hypoplasia (Debra *et al.,* 1992). Prenatal sonographic demonstration of the bronchopulmonary sequestrations have been reported sporadically in the radiological literature (Alan *et al.,* 1994; McClelland *et al.,* 1977; Mariona *et al.,* 1986). Thus pulmonary sequestration should be included in the differential diagnosis of solid masses found either within the normal lung (intralobar) or outside the normal lung (extralobar). Extralobar sequestrations can also be found below the diaphragm where they may be mistaken for intrabdominal malignancies such as neuroblastoma (Darlene Moore Bak, 2001). The disparity in occurrence between the intralobar and extralobar sequestrations may be due to the fact that the extralobar sequestrations are nor susceptible to the infective complications as compared to intralobar sequestrations and therefore less frequently detected (Olufemi Willams and Enumah, 1968).

Among the detected pulmonary sequestrations 75% of cases are intralobar located within the pleural investment of the normal lung parenchyma and mainly affects the lower lobe of the lung. Approximately 25% are extralobar usually situated at the base of left chest and diaphragm. The arterial blood supply of extralobar sequestration arises from either thoracic or abdominal aorta in 80% of cases and from spleenic, gastric artery, subclavian artery or intercostal arteries in 15% cases and pulmonary artery in 5% cases. The venous drainage of extra pulmonary sequestration is usually through the systemic circulation into hemiazygous vein, azygous vein, inferior vena cava or portal vein (Herman and Siegel, 2009).

Approximately 65% of extralobar sequestrations are located on the left side and 70% of these are located between the left lower lobe and the diaphragm and 10% are located below the diaphragm (Herman and Siegel, 2009; Laje *et al.,* 2006).

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CASE REPORT

A 24 years old woman with 6 months (26 weeks of gestation) came to the hospital for antenatal Ultrasonography for the first time. She was primigravida. A routine ultrasound was performed which revealed a solid intrathoracic mass. The mass was situated along the medial paraspinal part of left lower lobe. A cystic adenomatoid lung malformation and pulmonary sequestration was suspected. The size of the mass was approximately (6x5x4) cm and was above the left dome of diaphragm (Fig. 1). This mass slightly displaced the aorta to the right. Sonography showed an anomalous vessel branching from aorta which was entering into the echogenic mass.

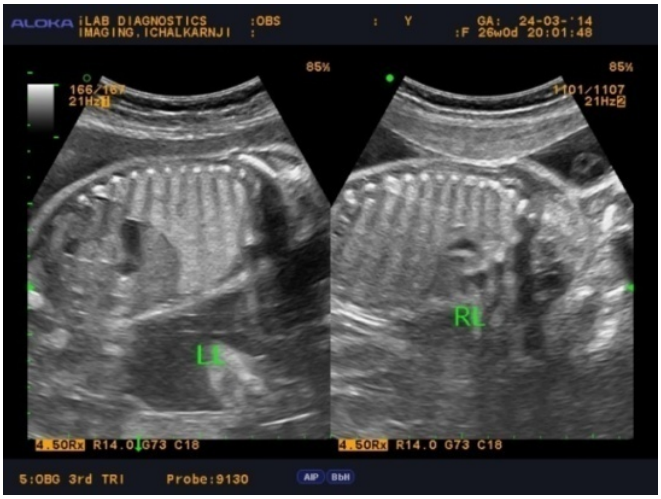
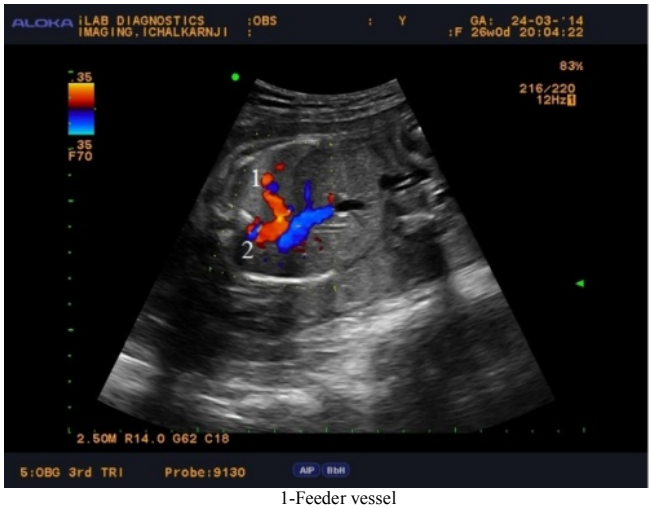


Fig. 1. The Ultrasonography of 26wks foetus showing the left and right lungs respectively

To enter into the perfection of diagnosis further colour Doppler Ultrasonography was done (Fig. 2) which visualized a feeder vessel arising from the aorta showing the flow of blood towards the mass. Repeated ultrasounds were performed on weekly basis. Once the patient reached 29 weeks of gestation investigations i.e. all biophysical exams were normal and there was no evidence of foetal compromise. The size of the mass remained consistence and there was no evidence of aortic compression.

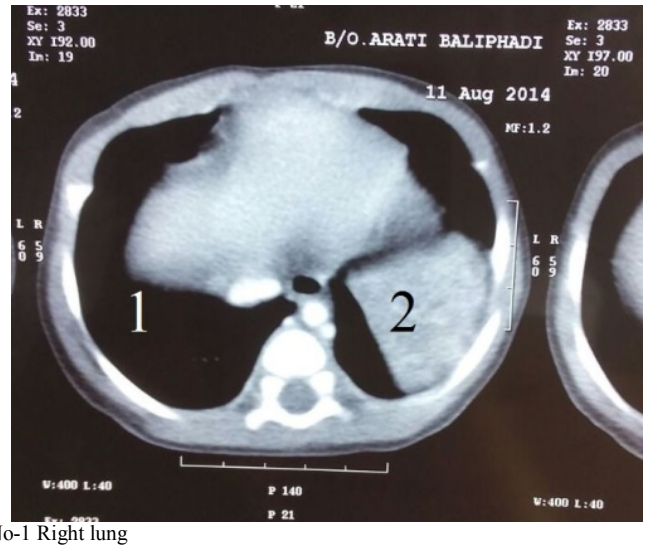


1-Feeder vessel
2-Foetal aorta

Fig.2. The colour Doppler study

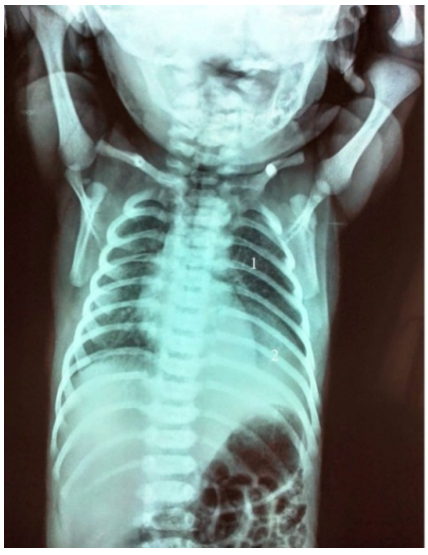
At delivery the baby was stable and was not in respiratory distress. The pulmonary function of new born was checked and revealed oxygen saturation of 98% at room temperature. There were no signs of pulmonary symptoms. The diagnosis was confirmed with postnatal ultrasound of chest and CE Computed Tomography chest (Fig.3) and x-ray chest (Fig. 4) of new born which showed the density along the basal portion of left lung which was consistent with sequestration.

Since there was no evidence of neonatal complications the sequestration was monitored by ultrasound and was awaited for surgical resection. One month after the delivery the baby was operated and the sequestrated mass was removed surgically (Fig.5). The specimen was sent for histopathological study which reported that the mass showed the dilated bronchioles, alveolar ducts and alveoli. This confirmed the extralobar sequestration. Six months after birth the x-ray chest of baby (Fig.6) showed normal lung on the left side and baby is completely asymptomatic.



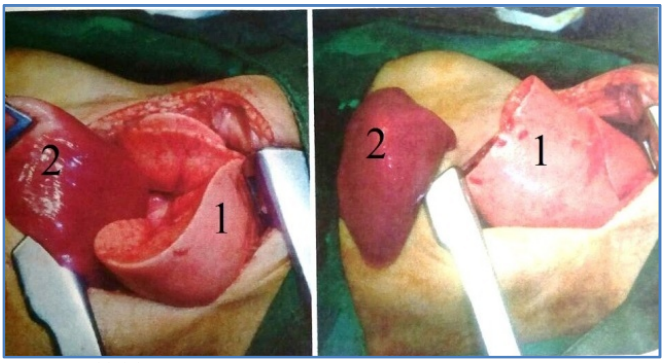
No-1 Right lung
No-2 Extralobar sequestrated part of left lung.

Fig. 3. The CE Computed Tomography of the new born baby



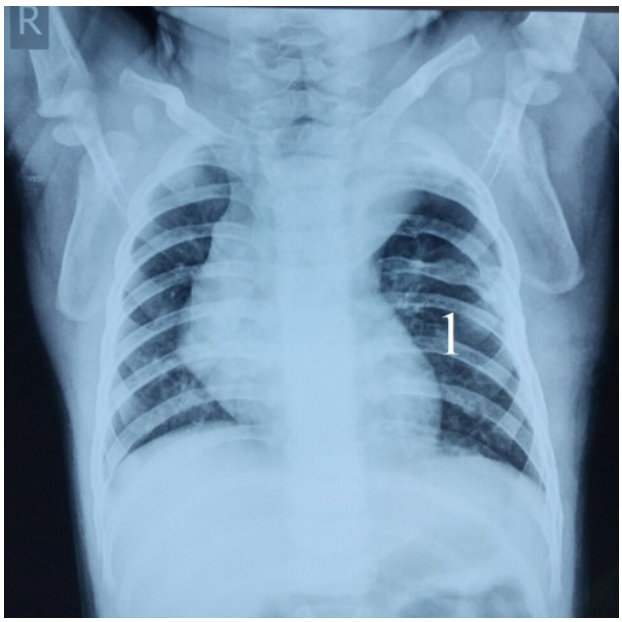
1-Left lung
2-Sequestrated lung

Fig. 4. The postnatal foetal x-ray chest



No-1 Normal left lung
No-2 Extralobar sequestrated portion of left lung

Fig.5. The surgical removed extralobar sequestration of left lung



No 1- Normal left lung

Fig.6. The X-ray of baby one month after surgery

Embryological Basis

The respiratory tract develops from the primitive gut; specifically the foregut. The lower respiratory tract begins to develop around the middle of fourth week of gestation from an out pouching of laryngotracheal groove (Shah *et al.,* 2003). Most frequently supported theory of how a sequestration arises is that an accessory lung bud develops from the ventral aspect of the primitive gut. The pleuroperitoneal tissue from this additional lung bud migrates in a caudal direction with the normally developing lung (Debra *et al.,* 1992). This accessory lung does not have any connection with the rest of the lung. The time of separation is important.

The accessory lobe that arises before the formation of pleura is surrounded by same pleura as normal lung and is called as intralobar while the one that arises after the formation of pleura has its own pleura and is known as extralobar (Debra *et al.,* 1992). Both the types have arterial supply from the thoracic or abdominal aorta and venous drainage via the systemic venous system (Shah *et al.,* 2003; Debra *et al.,* 1992).

DISCUSSION

The first description of an aberrant systemic artery to the lung was by Huber in 1777 (Debra *et al.,* 1992), the term sequestration was first introduced by Pryce in 1956 after his description established the lesion as a distinct clinical entity (Chih-Hung Lin *et al.,* 2013; Debra *et al.,* 1992). The first antenatal diagnosis of extralobar pulmonary sequestration was in 1986; by Weiner et al (Debra *et al.,* 1992). Numerous theories about the etiology of pulmonary sequestrations have proposed. For extralobar sequestration the most widely accepted theory is that; it results from the persistence of systemic arteries causing traction over the lung such that a portion of lung separates from the main lung mass (Chih-Hung Lin *et al.,* 2013; Debra *et al.,* 1992; Darlene Moore Bak, 2001; Herman, 2009).

Extralobar sequestrations are located outside the normal lung parenchyma and are enclosed in its own visceral pleura (Shah *et al.,* 2003; Debra *et al.,* 1992; Darlene Moore Bak2001). In our case also such intrathoracic mass which was diagnosed as extralobar sequestration was enclosed in its own visceral pleural sac. Extralobar sequestrations are predominantly found on left side between the lower lobe of left lung and diaphragm (Debra *et al.,* 1992; Alan *et al.,* 1994; Darlene Moore Bak, 2001; Olufemi Willams and Enumah, 1968; Herman and Siegel 2009). In our case the Ultrasonography examination revealed the same position of the intrathoracic mass. This was also confirmed after birth of the baby by computed tomography and x- ray chest.

According to the various studies; the blood supply of extralobar sequestration arises from thoracic or abdominal aorta in 80% of cases (Chih-Hung Lin *et al.,* 2013; Debra *et al.,* 1992; Darlene Moore Bak, 2008; Herman, 2009). Such feeder vessel was identified in our case also, the vessel was arising from aorta and the venous drainage was in the systemic azygous vein. The Colour Doppler ultrasound in our case confirmed the presence of such feeder vessel along with the blood flow. The anomalous blood supply is the hallmark of diagnosis of sequestration and it was confirmed. The presence of feeder blood vessel is very important because it distinguishes a pulmonary sequestration from lobar emphysema or congenital diaphragmatic hernia.

Increasingly extralobar sequestrations are discovered by Ultrasonography and about 35% to 70% regress during pregnancy or postnatally (Herman, 2009). Extralobar pulmonary sequestrations are usually asymptomatic and in some cases reversible without any treatment (4). In our case surgical intervention was necessary as the sequestration may lead to infection. Post surgery foetal X ray showed normal lung on left side with all pulmonary functions.

In conclusion a prenatal diagnosis of intrathoracic mass enables its early postnatal evaluation. Extralobar pulmonary sequestration should be considered in the differential diagnosis of mediastinal malformation in children. The prognosis of extralobar pulmonary sequestration depends on the presence of associated anomalies and is good in foetus with an isolated finding; same thing was observed with our case, there were no

associated anomalies. The study of this case was to raise the awareness of radiologist, pediatric surgeons in the diagnostic process of mediastinal lesions in children as well as for the undergraduate medical students as a part of applied anatomy to be kept in mind

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