

Fetal Intralobar Pulmonary Sequestration: Very Unusual Two Cases


Aynur MUSTAFA,^a

Nazlı JALALZADA,^a

Mohamad S. MOUSTAFA,^a

Mete Gürol UĞUR,^a

 Hüseyin Çağlayan ÖZCAN,^a

 Neslihan BAYRAMOĞLU TEPE,^a

 Özge KÖMÜRÇÜ KARUSERCİ^a

^aDepartment of Obstetrics and Gynecology,
Gaziantep University Faculty of Medicine,
Gaziantep

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Correspondence:

Aynur MUSTAFA

Gaziantep University, Faculty of Medicine,

Department of Obstetrics and Gynecology,

Gaziantep, TURKEY

dr.aynur.moustafa@hotmail.com

ABSTRACT Intrapulmonary sequestration is a rare congenital abnormality occurs as a result of tracheobronchial tree malformation and can be associated with pulmonary hypoplasia and hydrops fetalis, both are considered as life threatening conditions. We report two cases with intralobar sequestration. First patient is a 30-year-old pregnant who visited our outpatient clinic for routine follow up. Ultrasonography showed a hyperechogenic lesion in the right lung of 25 weeks male fetus. In the second 25-year-old patient, we report a hyperechogenic lesion in left lower lobe in a 26 weeks fetus, who was also on a suspicion of ventricular septal defect that was later confirmed by fetal echocardiography. Our aim is to increase the awareness of pulmonary sequestration as a differential diagnosis of intrathoracic masses and to ensure the early diagnosis during pregnancy to avoid the hazardous complications.

Keywords: Bronchopulmonary sequestration; abnormalities

Pulmonary sequestration is a rare congenital malformation of the tracheobronchial tree. The affected part of the lung is nonfunctional and has no connection with the tracheobronchial tree. Blood supply of this segment of lung is provided by an anomalous systemic vessel commonly originating from descending aorta which distinguishes the pulmonary sequestration (PS) from other congenital pulmonary airway malformation (CPAM).¹ It has two subtypes: extrapulmonary and intrapulmonary. Latter is divided into intralobar and extralobar.¹ Intrapulmonary sequestrations can be further classified as extralobar, which means that they are enveloped in their own pleura, while intralobar are within the pleura of another lobe. Coincident malformations encompass situs inversus totalis, double aortic arch, and other abnormalities.^{2,3} Pulmonary sequestrations (PS) compose 0.15% to 6.45% of all pulmonary malformations.⁴ Extralobar pulmonary sequestration affects males more than females with a ratio of 4: 1. However, few reports suggest that there is no incidental difference between two genders.⁵ PS are usually followed up during pregnancy, and can be safely resected in postnatal period. Generally, the prognosis is excellent unless the lesion is complicated with pleural effusion or hydrops fetalis.

CASE REPORTS

We represent two cases of intralobar pulmonary sequestration:

1) a 30-year-old female (G2 A0 P1) presented to our outpatient clinic on November 24th, 2015 at her 25th week of gestation based on her last

menstrual period (LMP). She had no history of systemic disease, signs of infection, drug abuse or any known allergy. General and obstetric physical examination revealed no abnormalities. Her routine laboratory tests results were; Hg:11,5 g/dL, Hct: 36%. Obstetric USG showed a male fetus with a 21*12.5 mm hyperechogenic, wedge-shaped bright right lower pulmonary lesion (Figure 1). The diaphragm appeared intact with no pleural effusion, hydrothorax or polyhydramnios. Fetal biometric parameters were compatible with 25 weeks of gestational age. Fetal echocardiography demonstrated normal anatomic structures and vascular connections. **No other abnormalities have been detected.**

2) a 25-year-old female (G5 A0 P2) was referred to our department on a suspicion of fetal anomaly at her 26th gestational week based on her LMP. There was third degree consanguineous. Her previous medical and surgical history was unremarkable. Laboratory results were normal. An obstetric ultrasound revealed a male fetus with a 23*23 mm hyperechogenic left lower pulmonary mass, no pleural effusion, hydrothorax or polyhydramnios (Figure 2). This mass was consistent with intralobar pulmonary sequestration. Fetal biometric parameters were compatible with 26 weeks of gestational age. Fetal echocardiography suggested the possibility of ventricular septal defect (VSD) and postnatal reassessment was recommended. No other abnormalities have been detected.



FIGURE 1: A male fetus with a 21*12.5 mm hyperechogenic, wedge-shaped bright right lower pulmonary lesion.



FIGURE 2: A male fetus with a 23*23 mm hyperechogenic left lower pulmonary mass.

- Follow up USG for our first case showed complete regression of mass. On the other hand, we lost follow up second case, she was referred to our clinic from rural region and after diagnosis we couldn't reach her.

Our first pregnant delivered at 38w4-5d, and the neonate unit assessment revealed no complication in baby.

DISCUSSION

Pulmonary sequestration (also known as bronchopulmonary sequestration) is a rare malformation, The affected part of the lung is nonfunctional and has no connection with the tracheobronchial tree which accounts for approximately 0.15% to 6.45% of all pulmonary malformations.⁴ its blood supply is provided by an anomalous systemic vessel commonly originating from descending aorta which distinguishes the PS from other congenital pulmonary airway malformation (CPAM).¹

These lesions usually appear next to the lung (extrapulmonary) or within one part of the lung (intrapulmonary). Intrapulmonary sequestrations can be further classified as extralobar, which means that they are enveloped in their own pleura, while intralobar are within the pleura of another lobe. Fortunately, intralobar sequestrations (ILS) are more common and compose 75%-85% of all intrapulmonary sequestrations, and they have a lower coincidence of other malformation compared to ELS (extralobular sequestrations).⁶ ILS frequently

affect left lower lobe and posterior basal segments of the lung.⁷ Moreover, extralobar pulmonary sequestration affects males more than females with a ratio of 4:1. However few reports suggest that there is no incidental difference between two genders.⁵

One of our cases was diagnosed as intralobar sequestration of the right lung, which is very rare. Most likely the underlying pathology is the bronchial atresia or bronchial obstruction leading to these malformations.⁸

The diagnosis established by ultrasonography in prenatal period by detecting a homogeneous, hyperechogenic mass in the fetal thorax. As neonates and infants are usually asymptomatic, more than half of cases are diagnosed later in childhood or in adulthood.⁸

Even though, the blood supply of the pulmonary sequestration is from the aorta, untreated cases can be associated with a large ipsilateral pleural effusion in approximately 5 to 10 percent of cases, and some times with pulmonary hypoplasia and hydrops fetalis.¹ Coincident malformations encompass situs inversus totalis, double aortic arch, and other abnormalities.^{2,3} As in our second case we reported PS associated with VSD.

Furthermore, frequent pneumonia, recurrent hospital admissions and increased morbidity and mortality rates are considered as late complications of PS if undiagnosed in prenatal period.⁷

During pregnancy many of these lesions regress and 40% of them show complete resolution, especially in the third trimester as in our first case.¹ Prenatal conservative management is the most appropriate method if sequestration is not accompanied with any other malformation or complication. Persistent conditions can be managed safely either by postnatal lobectomy or postnatal segmental resection.⁹

Some prenatal disorders, such as congenital pulmonary airway malformation, bronchogenic cyst, pulmonary arteriovenous malformation, scimitar syndrome have similar imaging findings and should be considered in the differential diagnosis of pulmonary sequestration.

Prognosis of the bronchopulmonary sequestrations largely depends on the concomitant anomalies, cardiovascular malformation and presence of hydrops fetalis. However, the reported overall survival exceeds 95% for bronchopulmonary sequestrations.⁹

There are many strategies to manage this condition especially if it was complicate with hydrops fetalis. These methods include the following procedures: direct fetal furosemide and digoxin therapy also combined absolute alcohol injection into the feeding vessel and placement of a thoraco-amniotic shunt. Last intervention can be used to decrease intra uterine fetal demise rate resulting from mediastinal compression the can lead to polyhydramnios and fetal hydrops thus prenatal death.¹⁰

One study reported that using polidocanol to sclerose feeding artery of these lesions is effective and save.

Percutaneous ultrasound guided fetal sclerotherapy leads to relief fetal hydrops, polyhydramnios and mediastinal compression due to discontinuation of blood supply to the feeding artery of sequestered mass.¹⁰ Although, open fetal surgery with lobectomy can be considered in microcytic disease it is high invasive intervention to the mother.¹¹ On the other hand, in postnatal period a Doppler or magnetic resonance imaging scanning of systemic blood supply malformations is recommended to neonates with a history of antenatal lung mass to detect the presence of cystic adenomatous malformation that can be associated with of pulmonary sequestration.¹²

Also, post natal imaging and respiratory complications indicate patients who are candidate for postnatal interventions.¹³

CONCLUSION

We presented these two cases to increase the awareness about the pulmonary sequestrations in the differential diagnosis of fetal intrathoracic masses. When suspected, patients with pulmonary sequestration should be referred to tertiary centers

for appropriate follow up. Although, it is a rare condition which regresses spontaneously in most cases, it should not be ignored. Early diagnosis in prenatal period helps avoiding the possible complications such as pulmonary collapse and hydrops fetalis, recurrent pneumonia and developmental delay in the childhood.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: Aynur Mustafa; **Design:** Mohamad S Moustafa; **Control/Supervision:** Mete Gürol Uğur; **Data Collection and/or Processing:** Hüseyin Çağlayan Özcan, Özge Kömürcü Karuserci; **Analysis and/or Interpretation:** Nazlı Jalalzada; **Literature Review:** Neslihan Bayramoğlu Tepe; **Writing the Article:** Aynur Mustafa; **Critical Review:** Nazlı Jalalzada; **References and Fundings:** Hüseyin Çağlayan Özcan; **Materials:** Özge Kömürcü Karuserci.

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