A Case Report of Congenital Pulmonary Airway Malformation

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ABSTRACT

Introduction:. Congenital Pulmonary Airway Malformation (CPAM) , is a rare lower respiratory tract developmental disorder. Although rare , it is the most common congenital lung lesion. Histologically , there are 5 subtypes and the most common form is type 1.

Case Presentation: We present a case of Congenital Pulmonary Airway Malformation with 74*50*74 mm cystic mass with heterogeneous septations on the left side of the thorax , dextrocardia , amnion fluid index total 30 cm , ascites in the abdomen and hypoplasia of the right lung at 28 weeks 4 days of gestation.

A pregnant women who was 18 years old , gravida 2 abortion 1 , 28 weeks and 4 days according to her last menstrual date was referred to our center because of a mass in fetal thorax. There was no history of any disease or drug use in the pregnant womans's history. In the fetal ultrasound evaluation of the patient ; 74*50*74 mm cystic mass with heterogeneous septations on the left side of the thorax , dextrocardia , amnion fluid index total 30 cm , ascites in the abdomen and hypoplasia of the right lung were observed. Information about amniodrainage , shunt , cyst aspiration was given. Hospitalization was recommended and the patient was admitted. After betamethasone cure treatment , the cyst content was aspirated up to 90 cc and amniodrainage was performed. A consultation with the medical genetics department was made with the aminocentesis sample taken. A decrease was observed in the size of the mass in the fetal thorax and in the abdominal acidity. . In the controls , it was observed that the mass in the thorax regained its former dimensions. While pregnant 37weeks 5days ; the birth took place due to the onset of labor. After birth , thorax computed tomography and cardiac echocardiography were performed on the newborn. The pediatric surgery department planned cyst excision. Cyst excision pathology result revealed Type 1 Congenital Pulmonary Airway Malformation.

Discussion: Congenital Pulmonary Airway Malformation (CPAM) , previously known as congenital cystic adenomatoid malformation (CCAM) , is a developmental malformation of lower respiratory tract .With the widespread use of antenatal ultrasound ; prenatal diagnosis of CPAM is increasing.It may cause respiratory distress in the neonatal period or may remain asymptomatic.CPAM is pathologically divided into 5 groups from 0 to 4 ; it has different clinical presentations and prognoses.

Conclusion: Congenital Pulmonary Airway Malformation (CPAM) prenatally it is classified as microcystic and macrocystic ; although it can be seen in mixed lesions.Prenatal diagnosis is usually made for the first time according to the second trimester ultrason findings.Macrocystic CPAM is characterized by a >5mm diameter anechoic cyst , while microcystic CPAM is <5 mm solid and homogeneous.CPAM should include other congenital anomalies in the intial evaluation.Although there is no increase in the incidence of chromosomal anomalies in CPAM cases alone ; microarray research can be offered.

Keywords: Congenital Pulmonary Airway Malformation (CPAM), prenatal diagnosis and management

INTRODUCTION

Congenital Pulmonary Airway Malformation (CPAM) , is a rare lower respiratory tract developmental disorder. Although rare , it is the most common congenital lung lesion. Histologically , there are 5 subtypes and the most common form is type 1.

CPAM can affect the normal development of the airways , lung parenchyma and vasculature ; this spectrum is thought to be secondary to obstruction , and the resulting pathology depends of the level , degree and timing of the obtruction.Prenatal evaluation ; it depends on the size of the mass , the amount of the mediastinal shift , fetal hemodynamics and associated anomalies.In the absence of hydrops , the prognosis is good and the live birth rate is above the 95th percentile.Surgical intervention in the postnatal period is therapeutic and prognosis for survival is excellent.In the absence of surgery , recurrent pulmonary infections are the most common and even associated with malignancy.

CASE PRESENTATION

A pregnant women who was 18 years old , gravida 2 abortion 1 , 28 weeks and 4 days according to her last menstrual date was referred to our center because of a mass in fetal thorax.There was no history of any disease or drug use in the pregnant woman's history. In the fetal ultrasound evaluation of the patient ;74*50*74 mm cystic mass with heterogeneous septations on the left side of the thorax(figure1) , dextrocardia (figure 2) , amnion fluid index total 30 cm , ascites in the abdomen (figure 3) and hypoplasia of the right lung (figure 4) were observed.Information about amniodrainage ,shunt , cyst aspiration was given.Hospitalization was recommended and the patient was admitted. To the hospitalized pregnant ; a course of betamethasone , daily non-stress testing and biophysical test profile was performed. After betamethasone cure treatment , the patient ; she accepted amniodrainage and cyst aspiration treatment options. The cyst content was aspirated up to 90 cc and amniodrainage was performed. A consultation with the medical genetics department was made with the aminocentesis sample taken. A decrease was observed in the size of the mass in the fetal thorax and in the abdominal acidity. The patient , who was stable the next day , was discharged and a follow-up was recommended one week later. In the controls , it was observed that the mass in the thorax regained its former dimensions. While pregnant 37weeks 5days ; the birth took place due to the onset of labor. After birth , thorax computed tomography and cardiac echocardiography were performed on the newborn. The pediatric surgery department planned cyst excision. Cyst excision pathology result revealed Type 1 Congenital Pulmonary Airway Malformation. Congenital Pulmonary Airway Malformation (CPAM) , is a rare lower respiratory tract developmental disorder. Although rare , it is the most common congenital lung lesion. Histologically , there are 5 subtypes and the most common form is type 1. Here we present a case of type 1 CPAM .

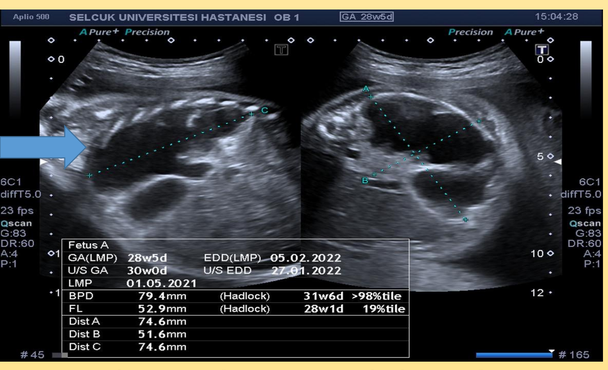

Figure 1: Blue arrow: pointing to thorax mass

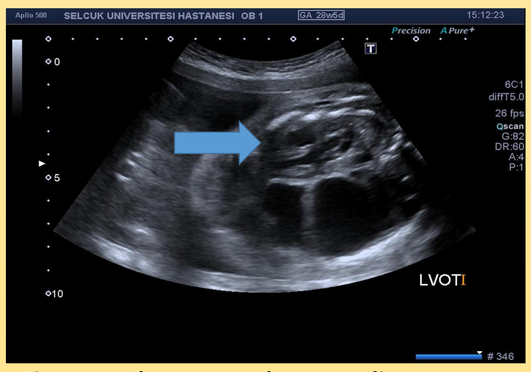

Figure 2: Blue arrow: dextrocardia

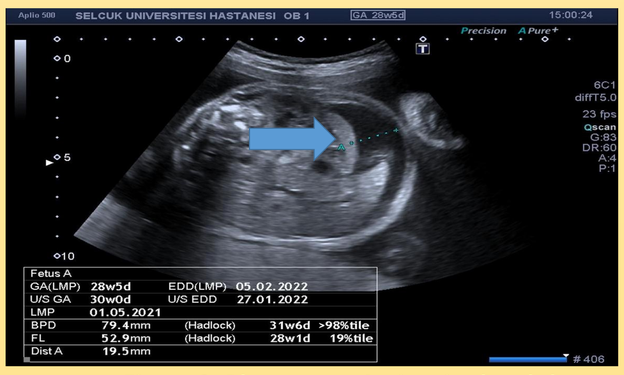

Figure 3: Blue arrow: Ascites in the abdomen

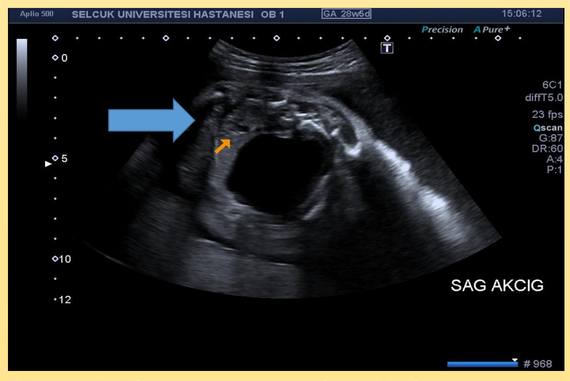

Figure 4 :Blue arrow:Hypoplasia Right Lung

DISCUSSION

Congenital Pulmonary Airway Malformation (CPAM) , previously known as congenital cystic adenomatoid malformation (CCAM) , is a developmental malformation of lower respiratory tract .With the widespread use of antenatal ultrasound ; prenatal diagnosis of CPAM is increasing.It may cause respiratory distress in the neonatal period or may remain asymptomatic.CPAM is pathologically divided into 5 groups from 0 to 4 ; it has different clinical presentations and prognoses.

CONCLUSION

Congenital Pulmonary Airway Malformation (CPAM) prenatally it is classified as microcystic and macrocystic ; although it can be seen in mixed lesions.Prenatal diagnosis is usually made for the first time according to the second trimester ultrason findings.Macrocystic CPAM is characterized by a >5mm diameter anechoic cyst , while microcystic CPAM is <5 mm solid and homogeneous.CPAM should include other congenital anomalies in the intial evaluation.Although there is no increase in the incidence of chromosomal anomalies in CPAM cases alone ; microarray research can be offered..