Congenital Chylothorax: Case Report

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We present a case of fetal chylothorax while discussing about its management and outcomes. Fetal pleural effusion causes pulmonary hypoplasia of the lungs.

Patient with 31 weeks of gestation has detected to have a fetal pleural effusion by routine ultrasonographic imaging. Fetal echocardiography was unable to demonstrate a cardiac pathology. Fetal thoracentesis revealed lymphocyte predominance and chylothorax was diagnosed. Chromosome analysis, metabolic screening and TORCH screening were normal. After the application of antenatal steroids, a caesarean section was performed at 34th gestational week. The newborn was taken to neonatal intensive care unit. After treatment with medium-and short-chain fatty acids, octreotid, and pleural drainage newborn is discharged from the hospital well two months later.

Chylothorax is one of the rare causes of respiratory distress in term neonates and most commonly occurs due to hydrops fetalis and perinatal infections. Rarely, congenital malformations of the thoracic duct and variational changes of embryonic lymphatic network were reported after autopsy. Some cases of chylothorax are found to be associated with Turner, Down and Noonan syndromes. Genetic analysis and infection screening should be considered to reveal the etiology. Usually chylothorax has a good prognosis. However prematurity, hydrops fetalis and degree of pulmonary hypoplasia are the factors determining the mortality rate.

As a result, chylothorax should be included in the differential diagnosis of pleural effusion in the neonates, and thoracentesis should be made for early diagnosis. Appropriate and effective treatment of patients with chylothorax may contribute to the prognosis and neonatal survival.

Key Words: Fetal chylothorax, Fetal pleural effusion, Fetal hydrothorax, Fetal thoracentesis

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Introduction

Collection of fluid between the pleural membranes is called pleural effusion. With a very rare incidence in fetal period becomes about 1/10.000 to 15.000. The mortality rate is about 25% of cases.[1] The first case was identified in 1977.[2] Pleural effusion in fetal life is mostly caused by chylothorax. Chylothorax is unilateral in 90% of cases and are often on the right side.[4] Most serious complications occurs by the effect of pressure on the heart, lungs and lymphatic system.[5,6] It can also seen with combination of other malformations, or it can be found isolated. Chromosomal abnormalities such as trisomy 21 or monosomy X can also cause fetal chylothorax.[5,6,7] The differential diagnosis of hydrops must be done after detection of hydrothorax by ultrasound. Chylothorax detection is made by classic pleural fluid diffraction analysis and 1.1 mmol/L (110 mg/dL) or more triglyceride content and at least 80% of the cell count for lymphocytes, which is 1000 or more cells per ml. Prenatal treatment modalities are thoracentesis, plore-amniotic shunt and pleurodesis.

View and the amount of effusion, the severity of the pressure on the fetal lungs and the presence of hydrops determines the success of the initiatives to the prenatal period. Accompanying abnormalities, presence of bilateral pleural effusions, to be accompanied by fetal hydrops and term and preterm birth indicates in the neonatal period would be a poor prognosis.[5,6,7] In this article, a case of antenatally detected fetal chylothorax presented with literature.

Case Report

28 - year - old primiparous pregnant patient, all screening tests and ultrasonography was normal until the 31th gestational

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week. After this date, one-sided hydrothorax developed in fetus. In the family history there was no baby with anomaly. Wide pleural effusion and mediastinal shift to the right in the left hemithorax was identified in ultrasonography of the male fetus, biometric measurements compatible with 31th gestational week. There was no other accompanying morphological or cardiac anomaly observed in the detailed ultrasonography of fetus.

Laboratory studies determined rubella, toxoplasmosis, parvovirus B19, CMV Ig M and Ig G was negative. Indirect coombs and VDRL have negative results too.

Fetal echocardiography reported that the width of the heart chambers, heart rate and left ventricular contractility in the normal range, accompanied by minimal pericardial fluid despite massive pleural effusion in fetal echocardiography.

In 32th week ultrasound-guided cordocentesis and thoracentesis applied to the patient. 72 mL yellow liquid was drained from the pleural cavity. There was no bacterial growth in the pleural fluid culture. Microscopic examination resulted as 1% eosinophils, 98% neutrophils and 1% lymphocytes . In biochemistrical analysis 1,5 g/dl protein and 656 mg/dl triglyceride were found.

Cordocentesis was reported normal (46XY). 24 hours later collection of fluid in the pleural cavity started again. following 72 hours, increase in the amount of pleural effusion and mediastinal shift occured again therefore another thoracentesis was performed. 56 mL yellow-colored liquid successfully evacuated. In the follow-up of the fetus no recurrence of chylothorax developed, patient was discharged and proposed to come twice a week. 15 days after the discharge ultrasonography showed, development of chylothorax and pericardial effusion, subcutaneous edema and hydrops again. Fetal thoracentesis was made for the third time and neonatal unit were interviewed. With an interval of 24 hours following the administration of two doses of betamethasone a caesarean section was proposed. Following the administration of doses of betamethasone, at the 34th gestational week the baby was delivered with caesarean section. 2750 gr baby boy was born with 1th minute Apgar score of 4, and 5th minute Apgar score of 7. In the examination of the newborn's respiratory system during the neonatal period, tachypnea and intercostal retractions, the middle and lower region decreased breath sounds over the left lung were found. The patient was intubated and it was under observation in the neonatal intensive care unit. Follow-up of arterial blood gas parameters was normal and then extubated. Chest radiography was performed. Determination of massive pleural effusion on chest radiograph, chest tube was applied. The white color of the pleural fluid pH and density in 1020 was 7.65. Leukocyte count, 6.200 cells / mm 3 (95% lymphocytes) were detected. Pleural fluid protein 2.2 g / dL, glucose 154 mg/dL, triglycerides 964 mg/dL measured. Octreotide and medium chain fatty acids rich formula added to the current treatment.With the educe of the amount of chest tube drainage on follow-up, chest tube taken at 17th day. At the end of 32-day follow-up baby was discharged with healing.

Discussion

Many primary and secondary reasons may cause hydrothorax. The primary causes of pleural effusion, are almost the same as that in hydrops. For differential diagnosis of congenital infections maternal serology (rubella, toxoplasmosis, syphilis, herpes, and parvovirus B 19), blood group and antibody screening should be performed. MCA Doppler peak flow rate should be examined for the evaluation of fetal anemia. Since they may be accompanied by 6-17% chromosomal abnormalities in cases of fetal hydrothorax, fetal karyotyping is recommended. Detailed ultrasound and echocardiography should be performed due to the fact that morphologic and cardiac abnormalities may be present 25% along with the pleural effusion. It should be noted that pleural effusion can develop with structural abnormalities in the lung, congenital cystic adenomatoid malformation, bronchopulmonary sequestration and congenital diaphragmatic hernia Congenital goiter, thyroid teratoma and mediastinal occupying lesions can also take place in the etiology.

If intrauterine treatment was not applied 59% survival rate is reported.[12] Appropriate treatment modality in patients with pleural effusion in the prenatal period is controversial, many studies are still being carried out.

Only observation may be sufficient alone because spontaneous regression can be seen in patients who have a small amount of fluid with no underlying cause of effusion.[13] If the amount of effusion increases or hydrops develops quickly, fetal intervention should be applied.[12]

Treatment options include thoracentesis, thoraco-amniotic shunt and pleurodesis. Although not shown superiority against each other between therapeutic modalities, there are various views that thoraco-amniotic shunt is more effective.[12-13] Pleurodesis using OK-432 is a new treatment and currently studies are in progress to assess the impact of it.[14]

Pleural effusion can disappear spontaneously after thoracentesis.[15-16] Thoracentesis may be considered to be repeated when pleural effusion occurs again. At this point, depending on the repeated thoracentesis hypoproteinemia can develop and this may cause the development of hydrops.[12] Therefore, caution should be taken. Thoracentesis done before the birth, lessened the burden of respiratory of fetus at neonatal period.[17]

In determination of the correct treatment modality, gesta-

tional age is also important. Aubard et al. propose thoraco-amniotic shunt placement before 32nd gestational weeks and the thoracentesis after 32nd gestational week.[18]

In our case, the patient's effusion was discovered at 32 week, and thoracentesis was performed first. Due to the preterm period, by applying the two-weeks follow-up after thoracentesis, neonatal risks was tried to be minimized.

Due to the development of the hydrops after 34th gestation week, thoracentesis was performed just before birth and a healthy baby was born by caesarean section.

Although the fetal hydrothorax is rare, it is an important clinical condition which may cause fetal death. If the chylothorx development take place in the early stages of pregnancy, it can cause fetal pulmonary hypoplasia. Therefore appropriate procedures should be applied without delay. During periods near term, after performing a thoracentesis to reduce the risk of neonatal respiratory distress, birth can be made.

Konjenital Şilotoraks: Olgu Sunumu

Şilöz karakterli plevral efüzyon (şilotoraks) term yenidoğanın solunum sıkıntısının nadir nedenleri arasında yer almaktadır. En sık hidrops fetalis ve perinatal enfeksiyonlara bağlı olarak oluşmaktadır. İntrauterin dönemde plevral efüzyon, akciğerlerin gelişimini engellemekte ve akciğer hipoplazisine neden olmaktadır. Bu yazıda antenatal dönemde plevral effüzyonla takip edilen ve postnatal dönemde term bir yenidoğanda saptanan konjenital şilotoraks olgusu sunulmuştur.

Yirmisekiz yaşında gravida 1, parite 0 olan 31. gebelik haftasındaki hastada saptanan plevral efüzyon olgusu sunulmuştur.

Gebelik takiplerinde sorun olmayan hastada 31. gebelik haftasında plevral efüzyon saptanmıştır. Yapılan fetal ekokardiografide bu efüzyonun kardiyak kökenli olmadığı saptanmıştır. Yapılan fetal torasentez sonucunda lenfosit hakimiyeti saptanmış ve şilotoraks tanısı konulmuştur. Kromozom analizi, metabolik taramalar ve TORCH taramasında patoloji saptanmamıştır. Hastaya antenatal steroid uygulandıktan sonra 34. gebelik haftasında sezeryan ile doğum gerçekleşmiştir. Yenidoğan yoğun bakım ünitesinde 2 aylık tedavi döneminde, orta ve kısa zincirli yağ asitlerinden zengin nutrisyon, okreotid tedavisi ve plevral drenaj uygulanmış olup, yenidoğan taburcu edilmiştir.

Konjenital şilotoraks etyolojisi oldukça karmaşıktır. Genellikle doğum sırasında artan venöz basıncın torasik duktusu rüptüre etmesi neticesinde oluşmaktadır. Nadir olarak otopsilerde bildirilen vakalarda, konjenital torasik duktus malformasyonu görülmekte ve embriyonik lenfatik ağın varyasyonel değişiklikleri bildirilmektedir. Bazı şilotoraks olguları ise Turner, Down, Noonan sendromları ve hidrops fötalisle ilişkilidir. Şilotoraks etyolojisini saptamada genetik inceleme ve enfeksiyon taraması akılda tutulmalıdır. Şilotoraksın prognozu genellikle iyidir. Ancak prematürite, eşlik eden pulmoner hipoplazinin derecesi ve hidropsun varlığı mortaliteyi arttıran nedenler olarak belirtilmektedir. Sonuç olarak yenidoğan döneminde saptanan plevral efüzyonun ayırıcı tanısında şilotoraks yer almalı ve torasentez yapılarak erken dönemde tanı konulmalıdır. Şilotoraks olgularında yapılacak uygun ve etkin tedavi prognozu olumlu yönde etkileyecek ve yenidoğan sağkalımına katkıda bulunabilecektir.

Anahtar Kelimeler: Fetal şilotoraks, Fetal plevral efüzyon, Fetal hidrotoraks, Fetal torasentez

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