Congenital Chylothorax: Diagnosis Problems

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ABSTRACT

Fetal pleural effusion is most frequently a marker of chylothorax. It is usually unilateral and frequently located on the right side, due to the anatomical position of the thoracic duct. Fetal hydrothorax can be either primary or secondary to chromosomal, cardiovascular, hematologic, liver or metabolic diseases, congenital infections and skeletal dysplasia, or placenta or cord abnormalities.

Congenital chylothorax in the newborn can be associated with symptoms of severe respiratory distress, requiring rapid evacuation and respiratory support. In chylothorax, the appearance of the fluid is milky, yellowish, with a high cell count and increased level of triglycerides.

The treatment of neonatal chylothorax involves removal of substantial effusion, pleural drainage and initiation of a special diet to limit chylothorax fluid restoration.

This paper presents the case of a premature newborn at a gestational age of 36 weeks, with fetal hydrothorax, which was diagnosed in utero at 28 weeks of pregnancy. After the initial resorption of the pleural effusion, it restored in parallel with the increasing quantity of ingested breast milk.

Keywords: Chylothorax, Newborn congenital infection, Chromosomal disorders.


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INTRODUCTION

Chylothorax is the most frequent form of fetal pleural effusion. The location is usually unilateral and frequently on the right side due to the anatomical position of the thoracic duct. Fetal hydrothorax can be primary (chylothorax or congenital lymphangiectasia) or secondary to pulmonary pathology such as chromosomal, cardiovascular, hematologic, liver or metabolic disease, congenital infections, skeletal dysplasia or placenta or cord abnormalities.

Intrauterine intervention to evacuate the chylothorax is recommended if it is associated with hydrops or if the chylothorax is large. Thoracentesis is always performed with an ultrasound follow-up for monitoring of the effusion recurrence.

Congenital chylothorax in the newborns can be associated with symptoms of severe respiratory distress, requiring rapid evacuation and respiratory support. Chylothorax is characterized by a milky, yellow appearance, rich cellularity, an increased number of lymphocytes, and increased levels of chylomicrons and triglycerides after the initiation of enteral feeding.

The treatment of neonatal chylothorax involves evacuation of the substantial effusion, pleural drainage and initiation of a special diet to prevent chylothorax recurrence. Special products containing medium-chain triglycerides are used, as they are absorbed directly from the bowel, without going through the chylomicrons.

CASE REPORT

Here, we present the case of a newborn of a 23-year-old mother from the rural area who was under observation in the dispensary during the pregnancy. This was her second pregnancy. Her first child, a healthy baby, was delivered by cesarean section after a pregnancy with oligohydramnios. The current pregnancy was a physiological one until week 28 of gestation, when a bilateral hydrothorax was observed. The paraclinical and immunological investigations performed in the last trimester of pregnancy revealed high anti-toxoplasma IgG and anti-cytomegalovirus IgG titers. The levels of IgG antibodies against the Herpes simplex virus 1 and 2 were elevated, but with negative IgM antibodies against Herpes simplex virus 1 and an equivocal result at testing for IgM antibodies against Herpes simplex virus 2. The anti-parvovirus B19 antibodies (IgG and IgM) were negative. The results from the mother’s paraclinical investigations (blood count, biochemistry, glucose tolerance test and hemoglobin electrophoresis) were within normal range.

Given the condition of the fetus, a C-section was scheduled at 35 weeks of gestation. The mother received two doses of dexamethasone antepartum. At birth, the newborn had a weight of 2400 gm, an Apgar score of 9 at one minute, and 7 at five minutes, with ineffective breathing.
decreased vesicular murmur in the right hemi-thorax and hemodynamic impairment. Postnatal chest X-ray showed right pleural effusion (Fig. 2). Eleven millimeter of sero-citrine sterile fluid, with the biochemical characteristics of a transudate, were removed during the pleural puncture. Because of respiratory distress secondary to the presence of pleural fluid, respiratory support was necessary for two days. Additionally, the newborn was administered diuretics, analgesics, prophylactic antibiotic therapy and total parenteral nutrition (TPN). The evolution was slow but favorable, enteral feeding with breast milk was gradually initiated and parenteral nutrition was withdrawn. On the 14th day of life, symptoms of respiratory distress (tachypnea, breathing effort, intercostal and subcostal retractions and decrease in breath sounds) reappeared. The chest X-ray revealed pleural effusion and blunting of the right lung (Fig. 3).

Ultrasound examination did not reveal any heart abnormality. The fluid removed during the pleural puncture had characteristics similar to the one removed during the first pleural puncture. Based on this evolution and the fluid’s characteristics, congenital chylothorax was suspected. The contrast CT angiography (Fig. 4) showed moderate right pleural effusion, reduced left pleural effusion with
pulmonary condensation adjacent to the scizural line and some areas with exudative alveolitis. The thoracic duct and the lymphatic circulation could not be evaluated.

Based on the clinical and paraclinical data, congenital chylothorax was diagnosed. Respiratory support (continuous positive airway pressure) was ensured for 24 hours, enteral feeding was withdrawn and parenteral nutrition was initiated in association with Monogen orally. The evolution was favorable with a complete bilateral remission of the effusion. Consequently, parenteral feeding was withdrawn and Monogen was exclusively used for 14 days. Then, the breast milk was gradually reintroduced and the baby was discharged at the age of 39 days with a good general condition, no signs of respiratory failure, with weight gain according to his age and fed on breast milk and Monogen. After discharge, the evolution was favorable, without any effusion and Monogen therapy was ceased at the age of 2 months.

**DISCUSSION**

Congenital chylothorax is a relatively rare disease with an incidence of 1/10000 live births. A number of specific investigations must be performed to exclude any congenital infection of the TORCH group, infections with parvovirus B19, adenoviruses or syphilis. Hemoglobinopathy must also be excluded by performing hemoglobin electrophoresis in the pregnant woman. Because in 6 to 10% of cases, hydrothorax is associated to chromosomal abnormalities, more commonly Down or Turner syndromes,1,2 performing the fetal karyotype is also recommended, to exclude any chromosomopathy but in the presented case was not performed.1,2,11 Detailed antenatal ultrasound assessment is extremely important, because 25% of cases are associated with different malformations. The ultrasound signs suggesting a primary hydrothorax are: unilateral or bilateral asymmetric effusion. Unilateral effusion is usually not associated with other malformations; if other effusions are also present, the pleural effusion is disproportionately high compared to effusion in other sites.

Irrespective of the cause, fetal pleural effusion may be responsible for fetal or neonatal death by pulmonary hypoplasia due to chronic intrathoracic compression, cardiac compression and superior vena cava obstruction, with reduction in the venous return and decrease in the ejection volume or compression of the esophagus associated with an excess of amniotic fluid and premature birth. Neonatal mortality in cases with antepartum onset of pleural effusion ranges from 15 to 95%,1,2,8,10

There are cases of chylothorax associated with maternal morbidities, such as the mirror syndrome, in which the mother presents generalized edema due to a hydric placenta that produces vasoactive substances. In the case presented in this paper, the etiology of the hydrothorax could not be established before birth. Congenital infection was not confirmed, the maternal hemoglobin electrophoresis was normal and the mother did not present any pathology during pregnancy. The karyotype was not performed antepartum, and no abnormalities suggestive of chromosomopathy were found in the newborn.

The newborn’s evolution depends on the amount of effusion, and on whether hydrops is present or not. When fetal hydrothorax is diagnosed before the gestational age of 32 weeks, initiation of the antenatal treatment is recommended, consisting in single or serial thoracentesis and pleural amniotic drainage, especially in cases with significant effusion and associated hydrops.2,10,11,14 Recently, fetal intrapleural injection with OK-432 has been recommended for fetal hydrothorax. Until April 2012, 16 fetal chylothorax cases treated with this sclerose-inducing agent were reported worldwide. Thirteen of them had a favorable evolution until their discharge from the hospital.5-7 Cases treated with fetal intrapleural injection of maternal blood were also reported.9

In the case presented in this paper, thoracentesis was not performed during the fetal period. When the effusion became significant and influenced the fetus’s condition, C-section was performed, considering the proper gestational age in terms of lung and brain development.

During the neonatal period, the pleural effusion may be present since birth or may occur secondary to heart surgery, insertion of a central venous catheter, or surgery for diaphragmatic hernia or esophageal atresia. Pleural effusion may occur in some neonatal pathology, such as heart abnormalities, thrombosis of the superior vena cava or of the subclavian vein, heart failure, tumors or neonatal sepsis. The pleural effusion fluid can be transudate or chylous. The characteristic features of the lymphatic fluid are the macroscopic milky appearance; triglycerides >100 mg/dl; lipid electrophoresis reveals the presence of chylomicrons; lymphocytes are predominant (60-90%); it is sterile; the amount of proteins and other biochemical components are generally similar to those in the blood and pH is 7.4. For differential diagnosis, imaging investigations, such as lymphoscintigraphy and lymphogram may be performed.2,17,18

In terms of therapy, numerous nonsurgical and surgical procedures are described in the literature. The nonsurgical therapeutic methods, also important for diagnosis are: withdrawal of lipids administration, total parenteral nutrition and the initiation of enteral feeding with the special formula containing medium-chain triglycerides. These medium-chain triglycerides do not need chylomicrons for intestinal absorption and do not promote the recurrence of lymphatic effusion. Cases with favorable evolution after Octreotide, a Somato-statin analog, administration were also reported.13,15,16 The case presented in this paper had an oscillatory evolution.
After the initial resorption, the pleural effusion reappeared consequently to an increase in the lipids intake. Afterward, the special formula diet with high content (90%) of medium-chain triglycerides (Monogen) was initiated. This facilitated the initial effusion resorption and supported the favorable evolution of the newborn. The effusion did not recur and the withdrawal of the special formula was possible, being followed by transition to enteral feeding on breast milk.

Surgical interventions, such as the control at the fistula site, pleurodesis, duct ligation, pleuropertitoneal shunting, diaphragmatic fenestration are therapeutic options and are recommended for chylothorax cases with a large or persistent leak.12,17,18

**CONCLUSION**

Chylothorax is a rare condition in neonatology. The analysis of pleural fluid can help to identify the etiology of this condition. Conservative management is recommended for most of the cases.

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**REFERENCES**