

Recurrent Congenital Chylothorax: A Case Report

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Abstract

We report a case of fetal chylothorax and hydrops. Chylothorax is a very rare condition, occurring in approximately 1 in 10000-15000 pregnancies, with an overall mortality rate of 25% to 50%. It is the most common form of pleural effusion in the prenatal period. Available treatments include thoracocentesis, pleuro-amniotic shunting and pleurodesis and the optimal antenatal management and timing are still controversial. We report a case diagnosed at 30 weeks' gestation that was submitted to bilateral pleural-amniotic shunt, but turned in to increased bilateral pleural effusion, generalized hydrops and polyhydramnios. Attempted intervention allowed to achieve good obstetric outcome. No etiology was found, revealing the diagnostic challenge this entity can be.

Keywords: Fetal chylothorax; Hydrops; Thoracocentesis; Pleuro-amniotic shunting; Pleurodesis; Noonan's syndrome

Introduction

Chylothorax is defined as the accumulation of lymph in the pleural cavity and it is the most common form of pleural effusion in the prenatal period [1]. It is a very rare condition, occurring in approximately 1 in 10000-15000 pregnancies [2], with an overall mortality rate of 25% to 50% [1,3]. It is more frequent in males, in a 2:1 proportion [4].

Pleural effusion can be isolated, associated with other anatomical malformations, or with a chromosomopathy, such as 45, 0X or trisomy 21 that is present in 5-17% of hydropic fetus [1,5]. It can also be a part of Noonan's syndrome or sialidosis [2].

After the diagnosis of pleural effusion, detailed investigation is essential to exclude other conditions and secondary pleural effusion such as congenital cystic adenomatoid malformation, bronchopulmonary sequestration, congenital diaphragmatic hernia or mediastinal tumors [2,6]. Primary fetal hydrothorax is a diagnosis of exclusion and should only be established after ruling out congenital infections (with maternal serology to toxoplasmosis, rubella, cytomegalovirus, syphilis, herpes and parvovirus B19), immune hydrops (blood type and antibody screen), fetal-maternal hemorrhage (Kleihauer-Betke test), fetal anemia (Doppler evaluation of the peak systolic velocity in the middle cerebral artery) and chromosomopathy (fetal karyotype) [2]. Diagnosis of chylothorax is made by cytological and biochemical analysis of the pleural fluid: yellow-colored fluid, a white cell count of greater than 1,000 cell/ μ L with a significant lymphocyte fraction (>70%) in the fluid, more than 1000 cells/ μ L, a triglyceride level of more than 110 mg/dL [1,4].

The natural history is variable. In 90% of cases pleural effusion is unilateral, usually located on the right side, and ranges from single effusion to important thoracic compression that can be fetal life-threatening [1,7]. The coexistence of hydrops, bilateral occurrence,

pulmonary hypoplasia, preterm labor or absence of pre-natal treatment is the main factors associated with poor neonatal outcome. The effusion may resolve spontaneously, remain stable or progress to hydrops with fetal or neonatal demise [1,2,7].

Available treatments include thoracocentesis, pleuro-amniotic shunting and pleurodesis [4,7,8]. The success of prenatal intervention through thoracocentesis depends on early diagnosis, the volume and reappearance of the effusion, the degree of pulmonary compression and the existence of hydrops, all of which are factors that worsen the prognosis [8]. However, there are no randomized trials comparing different interventions for fetal pleural effusions [2]. For hydropic fetuses with pleural effusions, retrospective outcomes from larger series support invasive therapy, and pleuroamniotic shunting appears superior to repeated thoracocentesis [6-8]. For non-hydropic fetuses, the benefit of shunting is less clear. Globally, the optimal antenatal management and timing are still controversial.

Case Report

We report the case of a 29 year old woman, gravida [1], followed on fertility clinic due to severe endometriosis, which had a spontaneous pregnancy. Her blood group was B Rh negative, with no other relevant history. She started pregnancy surveillance on our center and an increased nuchal translucency (NT=3.7 mm) was detected on 12 weeks' ultrasound (Figure 1), in association with reversed ductus venosus a-wave (Figure 2). Chorionic villous sampling was performed and the result of the karyotype was 46, XX.

A fetal echocardiography was performed at 18 weeks', which was normal. However, morphologic exam at 22 weeks' revealed a ventricular septal defect (VSD; Figure 3) confirmed by posterior echocardiography-without other malformations.

Immunoglobulin anti-D was administered at 12th and 24th week, after negative Coombs test. The ultrasound performed at 28 weeks' revealed a fetus in 80th percentile and a polyhydramnios with amniotic fluid index (AFI) of 22 cm. Two weeks later (30 weeks+1 day), the

control ultrasound revealed polyhydramnios, persistent VSD and unilateral pleural effusion (Figures 4 and 5). Fetal lung maturation was accelerated with 12 mg of betamethasone daily, two doses.

Serologic tests investigation was negative for TORCH group infection, parvovirus B19 and VDRL. Also, negative indirect Coombs was confirmed.

At 30 weeks+4 days ultrasound showed aggravation, with increasing of the polyhydramnios, and the pleural effusion, which became bilateral (Figure 6).



Figure 1: Ultrasoundin 12th week with increased nuchal translucency

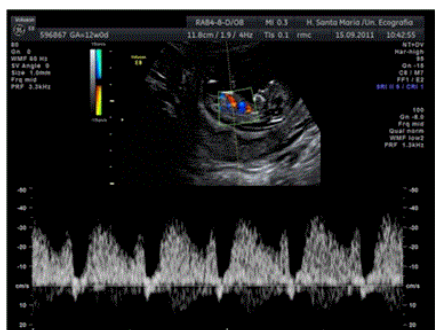


Figure 2: Reversed ductus venosus a-wave in 12th weeks

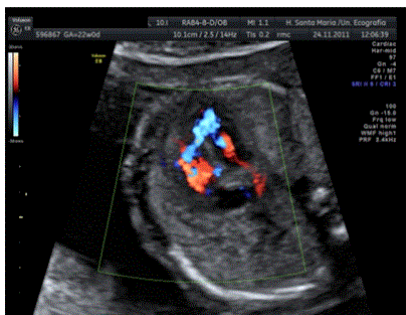


Figure 3: Morphologic exam with ventricular septal defect

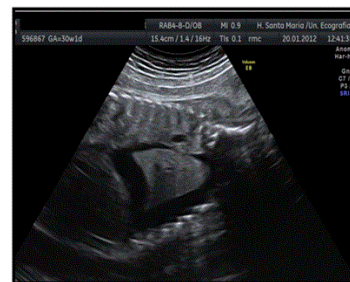


Figure 4: Unilateral right pleural effusion



Figure 5: Unilateral right pleural effusion



Figure 6: Bilateral pleural effusion

The patient was submitted to a diagnostic fetal thoracocentesis, in Maternidade Bissaya Barreto, Coimbra, Portugal. The presence of 98% lymphocytes suggested the diagnosis of chylothorax. Amniotic serologies for CMV, Hepatitis C, HIV, toxoplasmosis and syphilis were confirmed to be negative. Fetal RhD blood group positive was confirmed by PCR-based amplification assays and Coombs test was negative.

A double-pigtailed pleuro-amniotic shunt (Rocket Medical, Watford, UK) was inserted at right side, under sonographic guidance. Tocolysis and prophylactic antibiotic was given. Two days later, left pleuro-amniotic shunt was inserted and a total of 35 mL of amniotic

fluid was removed. Control ultrasound performed the day after the procedure reveal thin layer of bilateral pleural effusion. Conservative management with serial scans was suggested.

On the 32nd week, increased bilateral pleural effusion (Figure 7), generalized hydrops, Fetal Growth Restriction (FGR) and abnormal dopplerfluxometry were present, with cerebro-placental ratio above 5th percentile. Expectant management with fetal surveillance was decided. One week later ultrasound revealed bilateral pleural effusion aggravation, polyhydramnios with Doppler evidence of cerebro-placental redistribution.



Figure 7: Thirty-third week ultrasound revealing bilateral pleural effusion aggravation and polyhidramnios. In this scan it was also noted Doppler evidence of redistribution cerebro-placental.

The patient was admitted to the hospital the same day, starting spontaneous labor a few hours later. A cesarean section was performed for suspected fetal distress and a female newborn weighing 1700 g was delivered. Both thoracic drains were immediately clamped (Figure 8). Apgar scores were 7 and 9 at 1st and 5th min, respectively, but requiring endotracheal intubation and prolonged ventilation (until 62nd day of life).



Figure 8: Right thoracic drain being immediately clamped

On the 1st day of postnatal life, bilateral thorax shunts drained approximately 100-250 mL/day of pleural fluid, showing that they were functional. Cytochemical exam was always consistent with chylothorax.

Some dysmorphism was noted in the newborn: hypertelorism, low-set ears and downslanting palpebral fissures. Noonan syndrome was investigated, with inconclusive results.

A thoracic duct ligation was performed at 27th day: thoracic duct was found to be anomalous, with a large diameter. A bilateral

pleurodesis with talc was performed, which solved the pleural effusion. Alfa 1 antitrypsin was normal, CT scanning of the thorax was not typical of pulmonary lymphangiomatosis. The newborn was transferred to the pneumology unit for pulmonary function control. On the 6th month postnatal, cardiologic investigation showed multiple VSD without surgical indication. Conservative management with diuretics and oxygen was performed to optimized cardiac function.

Currently, aged 12 months (corrected age), the child has normal psychomotor, height-weight is below the age standards, and maintenances specific surveillance at the hospital.

Discussion

Hydrothorax may be associated with several possible diagnoses including mediastinal tumor, pulmonary sequestration, infection or chromosomal abnormalities [2], Noonan syndrome was suggested because of the dysmorphism observed. This is the most common genetic syndrome found in fetuses with an increased nuchal translucency and a normal karyotype [9], and must be considered in every fetus with these characteristics associated to pleural effusions [10]. Heart defects are found in about 60% of the cases [9] and FGR may also be part of the clinical presentation [10]. However, DNA studies for the syndrome were inconclusive.

FGR is not usually directly associated with fetal pleural effusions or chylothorax. Concomitant presence of FGR, cardiac defects and pleural effusion in our case may suggest a multifactorial cause instead of anatomical defect of the thoracic duct and lymphatic system.

In the present case, chylothorax was suggested because the effusion was initially unilateral and it was confirmed by the lymphocyte count of over 70% in the pleural fluid. Prognostic factors, such as early appearance of the effusion, the volume and speed of its replacement, influence the treatment to be undertaken and the associated morbidity/mortality [7]. Approach varies from expectant management through to monitoring by ultrasound scans to therapeutic intervention [7].

We performed a thoraco-amniotic shunt to treat chylothorax, due to its large volume. Thoracoamniotic shunting is recognized in several reports as the gold standard of treating fetal chylothorax, with success rate about 33-66% in hydropic fetuses and may approach 100% in non-hydropic fetuses [7,8]. The outcome after shunting is predominantly influenced by the presence of hydrops, with perinatal survival of 70-100% in its absence compared to 45-65% survival in hydropic fetuses [8]. In the present case we document a successful intervention with reduction of the pleural effusion, despite the recurrence observed two weeks later. There were no technical complications in our case, in a procedure with a mortality rate of 35% described [11].

The first signal in the present case was polyhydramnios at the 28th weeks' ultrasound. The presence of polyhydramnios frequently observed with pleural effusion is probably due to mediastinal and esophageal compression, with the obstruction of the physiological fetal swelling [7]. Polyhydramnios, with a consequent increase in intra-amniotic pressure, may reduce the drainage of the pleural effusion and may lead to preterm labor [2,4,7] as seen in the present case.

In this case the chylothorax did not overcome spontaneously in the neonatal period, despite the conservative treatment with assisted ventilation, pleural drainage and nutritional measures. The etiology remains unclear.

An overall mortality up to 50% has been reported, with poor prognosis in cases with gestational age less than 32 weeks at diagnosis, delivery prior to 35 weeks gestation or hydrops are associated [3], as we report on the present case. Despite all this factors, obstetric outcome and newborn development was favorable. We believe that early diagnosis and in utero intervention was crucial to prevent pulmonary hypoplasia and congestive cardiac failure described in similar cases. This was especially important to minimize fetal complications, and maximize the potential successful evolution of the children.

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