Fetal primary hydrothorax with spontaneous resolution

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A B S T R A C T

Fetal primary hydrothorax is a rare congenital anomaly with an estimated incidence of 1:10,000–15,000 pregnancies, with an unpredictable clinical course, ranging from spontaneous resolution to fetal death. We present a case of a unilateral fetal pleural effusion identified at 35th gestational week. A 37 year-old woman (G2P1) presented to our routine term pregnancy evaluation. The pregnancy had been otherwise uneventful. At echographic evaluation a large anechogenic fluid collection was identified in the right fetal hemithorax, with atelectasis of right lung, displacing the heart and mediastinal structures to the contralateral hemithorax. Hydramnios was also identified. No other structural abnormalities were detected, as no signs of hydrops. Fetal biometry was compatible with gestational age. Fetal ecocardiogram was structurally and functionally normal. Doppler evaluation of the peak systolic velocity in the middle cerebral artery was normal. Screening for congenital infections was negative. Complete blood cell count, blood type and antibody screening ruled-out immune hydrops. Karyotype analysis was not performed as family decision. Serial ecographic re-evaluations showed a progressive volume decrease and at the 38th week there was total resolution of the effusion. A C-section was performed at the 39th week. A live female infant was born weighing 3205 g, with no need of ventilatory support. One year post-partum follow-up evaluation confirmed the child was healthy. Spontaneous regression has been reported to occur in 9–22% of primary fetal hydrothoraxes, but the features predicting a better prognosis remain difficult to define. Unilateral effusion, spontaneous resolution and absence of hydrops at the age of diagnosis seem to be indicators of better outcome.

1. Introduction

Fetal pleural effusions are rare congenital anomalies with an estimated incidence of 1:10,000–15,000 pregnancies [1]. In Portugal, a study of neonates admitted to six neonatal intensive care units from 1997 to 2004 reported 62 cases of pleural effusions (incidence of 0.06%) [2].

The pleural space is outlined by the visceral and parietal pleura that cover the lung surface and chest wall respectively. The volume of fluid in such, sometimes virtual space, results of the balance of fluid production by the visceral pleura and absorption by the lymphatics of the parietal pleura.

Given the etiology, pleural effusions can be classified in primary, correctly termed “hydrothorax” antenatally and “chylthorax” post-natally, and secondary, usually associated with immune or non-immune hydrops, as infections, congenital lung lesions (cystic adenomatoid malformations, bronchopulmonary sequestration or congenital diaphragmatic hernia), congenital heart disease or chromosomal/genetic syndromes [1].

Primary fetal hydrothorax is due to a lymphatic leakage into pleural cavity resulting from anomalies of development of lymphatics, and generally is a diagnosis of exclusion. The majority of cases are diagnosed in the third trimester [3].

Maternal assessment should focus on establish whether it is primary or a secondary in etiology. It should include a detailed medical and obstetrical history, maternal serology to exclude congenital infections (toxoplasmosis, rubella, cytomegalovirus, parvovirus B19, syphilis and herpes), blood type and antibody screening to rule-out immune hydrops and Kleihauer-Betke test if there is a concern about maternal-fetal transfusion. Detailed ultrasound and echocardiographic evaluation should exclude major congenital abnormalities. Doppler evaluation of the middle cerebral artery to rule out fetal anemia and fetal karyotype...
are also strongly recommended.

Irrespectively of underlying cause, these conditions have an unpredictable clinical course ranging from spontaneous resolution to fetal death. The development of pulmonary hypoplasia or hydrops due to mediastinal shift and caval vein obstruction are the main feared complications [1,3].

2. Case Report

We present a case of a unilateral fetal pleural effusion identified at 35th gestational week. A 37-year-old patient gravida 2 para 1 (1 caesarean section seven years before) presented to our routine term pregnancy evaluation. Personal and family histories were unremarkable. The pregnancy had been otherwise uneventful. At ecographic evaluation we identified a large anechogenic fluid collection in the right fetal hemithorax, with atelectasis of right lung, displacing the heart and mediastinal structures to the contralateral hemithorax (Fig. 1). Hydramnios was also identified. A detailed ultrasound examination was performed and no other structural abnormalities were detected, as no signs of hydrops. Fetal biometry was compatible with gestational age. Fetal ecocardiogram was structurally and functionally normal. Doppler evaluation of the peak systolic velocity in the middle cerebral artery was normal. Screening for congenital infections was negative. Complete blood cell count, blood type and antibody screening ruled-out immune hydrops. Karyotype analysis was not performed as family decision. Serial ecographic re-evaluations throughout the following weeks showed a progressive volume decrease and at the 38th week there was total resolution of the effusion. A C-section was performed at the 39th week. A live female infant was born weighing 3205 g, with no need of ventilatory support. One year postpartum follow-up evaluation confirmed the neonate was healthy.

3. Discussion

The overall mortality rate is about 22–55% and the most common cause of neonatal death in a fetus diagnosed with hydrothorax is respiratory insufficiency due to pulmonary hypoplasia [1,3,4]. There is no established consensus for the management of this condition, given the high heterogeneity of the disease. A conservative approach, antenatal thoracocentesis or pleuroamniotic shunting are the options for the management and the choice should be based on gestational age, severity of the effusion, evidence of progression, and the presence or absence of concomitant hydrops [6,7].

Spontaneous regression has been reported to occur in 9–22% of primary fetal hydrothoraces and has been associated with nearly 100% of survival. However, the features predicting a better prognosis remain difficult to define. Unilateral effusion and absence of hydrops or hydramnios at the age of diagnosis seem to be indicators of a better outcome [3,5,8]. The presence or progression of hydrops and rapid enlargement of a pleural effusion with mediastinal shift are indicators of a poor prognosis and for urgent fetal intervention [1].

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