

Isolated congenital pleural effusion in two neonates

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ÖZET

İki yenidoğanda izole doğumsal pleural efüzyon

Doğumsal izole pleural efüzyon; yapısal malformasyonlar, inflamatuvar veya iatrojenik problemler, genetik sendromlar ve fetal hidrops ile ilişkilendirilen pleural aralıkta sıvı toplanmasıyla karakterize nadir bir anomalidir. Burada biri down sendromu diğeri de Burkholderia gladioli sepsisine bağlı empiyem ve kan yolu enfeksiyonuyla ilişkili izole konjenital pleural efüzyonu olan iki yenidoğan olgusu sunulmuştur. Bu nadir durum tanı ve tedavi yaklaşımı açısından tartışılmıştır.

Anahtar Kelimeler: İzole pleural efüzyon, down sendromu, Burkholderia gladioli sepsisi, torasentez.

SUMMARY

Isolated congenital pleural effusion in two neonates

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Congenital isolated pleural effusion, a non-specific accumulation of fluid in the pleural space, is an uncommon anomaly which can be associated with structural malformations, inflammatory or iatrogenic problems, genetic syndromes or fetal hydrops. Here, we present two neonates with isolated congenital pleural effusion, one of which was associated with Down syndrome and the other with empyema and bloodstream infection caused by Burkholderia gladioli septicemia. We wanted to discuss the diagnosis and management of this rare clinical entity.

Key Words: Isolated pleural effusion, down syndrome, Burkholderia gladioli septicemia, thoracentesis.

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Congenital pleural effusion is a rare clinical entity with an estimated incidence of 1/10.000-15.000 pregnancies (1). It has a wide spectrum of etiology from unknown etiology (idiopathic) to inflammatory association, chromosomal anomalies or structural malformations. The incidence of isolated pleural effusion in neonate and its association with inflammation or genetic syndromes have not been exactly known. Here, we report 2 neonates with isolated congenital pleural effusion, one of which was associated with down syndrome and the other with empyema and bloodstream infection caused by *Burkholderia gladioli* septicemia.

CASE REPORT

Case 1

A newborn, from 43-year-old gravida 4 para 3 woman, at 31 weeks gestational age was delivered by cesarean section without respiratory distress. He had been diagnosed as polyhydramnios and unilateral fetal pleural effusion on the right side by fetal ultrasonogram at 30 weeks of gestational age. The initial Apgar scores were 7 at 1 min and 8 at 5 min. After admission to neonatal intensive care unit (NICU), he had minimal tachypnea (66/min) and retractions without obvious respiratory insufficiency. Chest radiograph initially showed increased opacity of the right hemithorax with pleural effusion (Figure 1A). The baby had epicanthal folds, low-set ears, bilateral simian line and fifth-finger clinodactyly. There was no evidence of ascites, subcutaneous edema or pericardial effusion. Ultrasonogram confirmed anechoic fluid collection with 2 cm width in the right hemithorax. As there was no respiratory insufficiency, the infant was begun to follow up with conservative management. Repeated chest radiographs and weekly ultrasonogram were performed in order to detect the clinical course of the effusion. There was no rapid

enlargement of the effusion on serial ultrasound and the pleural fluid resolved spontaneously over a period of three weeks. The karyotype was 47, XY, +21, so he was diagnosed as down syndrome. Follow-up chest radiograph showed no recurrent pleural effusion over the next 6 months (Figure 1B).

Case 2

A 33-year-old multipar woman, gravida 4 para 2, was referred to obstetrics and gynecology department of our hospital at third-trimester for evaluation of a unilateral pleural effusion in her fetus. Follow-up prenatal ultrasounds demonstrated persistence of the pleural effusion and premature prelabor rupture of the membranes (PPROM) occurred. A 3900 g infant girl was born at 38 weeks of gestation by emergency cesarean delivery for fetal distress. On admission to NICU, respiratory insufficiency required intubation and positive pressure ventilation. The initial chest radiograph showed unilateral pleural effusion on the right hemithorax with the collapsed right lung (Figure 2A). Ultrasonogram confirmed massive pleural effusion with 3 cm width in the right hemithorax but collapsed or agenetic right lung could not be evaluated. Therefore, thoracic computerized tomography revealed intensive content of the pleural fluid with collapsed right lung and there was neither congenital mass nor structural or vascular malformation in the thorax of the baby. Complete blood count with differential demonstrated a white blood cell count of $20 \times 10^3/\mu\text{L}$ (78% neutrophils, 4% bands, 14% lymphocytes, 4% monocytes); hemoglobin 16.4 g/dL; hematocrit, 49%; and platelets, $285 \times 10^3/\mu\text{L}$ with a C-reactive protein value of 3.02 (N: 0-8) mg/L. Thoracentesis yielded 25 mL of turbid fluid with pH 7.30; lactate dehydrogenase 118 IU/L; glucose 53 mg/dL; protein 2.8 g/dL and triglycerides 25 mg/dL with innume-

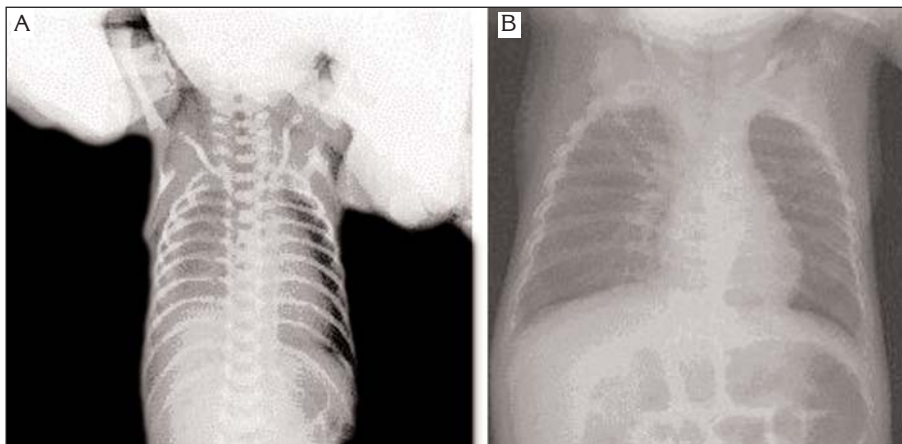


Figure 1. Chest radiograph view of the case 1 (A) showing increased opacity of the right hemithorax with pleural effusion on admission to NICU and (B) after 6 months on follow-up.

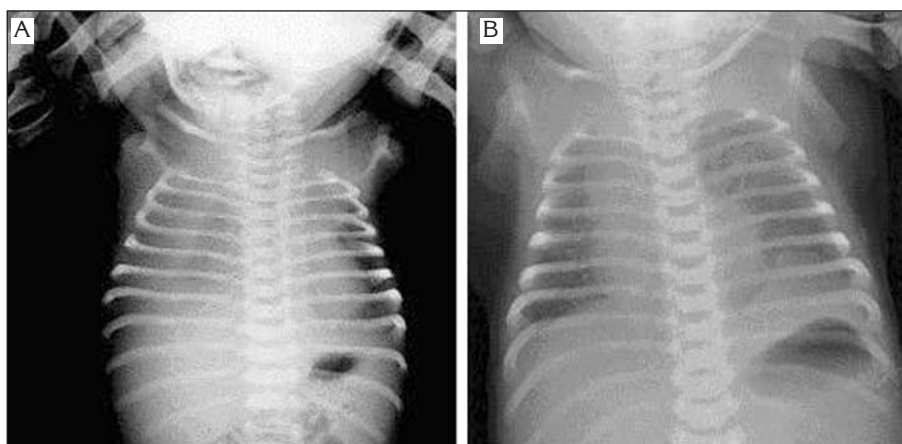


Figure 2. Chest radiograph of the case 2 showed (A) unilateral pleural effusion on the right hemithorax with the collapsed right lung and (B) clear resorption of pleural effusion after treatment.

rable white blood cell count. A chest tube was inserted for continuous drainage of the effusion and the pleural fluid was hazy as empyema. Bacteriologic examination demonstrated *B. gladioli* in the pleural fluid and admission blood culture of the baby. Antibiotic therapy with cephalexin and vancomycin was initiated. The pleural fluid resolved over a period of two weeks, allowing discontinuation of chest tube drainage. Follow-up chest radiograph showed clear resorption of pleural effusion (Figure 2B).

DISCUSSION

Pleural effusion in the fetus may cause pulmonary hypoplasia and is a risk factor for severe respiratory insufficiency after birth. It may also lead to the development of generalized hydrops and polyhydramnios, which are associated with preterm delivery, neonatal asphyxia and perinatal death (2). The natural history of fetal pleural effusion is not well understood and it can regress, remain stable or worsen in prognosis. Aubert et al. reported spontaneous regression in 22% of 204 cases of primary fetal pleural effusion (3). Spontaneous regression can even occur with large effusions and, very rarely, in the presence of hydrops (1,4,5); however, it is more likely when the diagnosis is made early in the second trimester, if the effusion is unilateral and in the absence of hydrops or polyhydramnios (3). Premature delivery and the presence of hydrops have been reported as poor indicators while gestation at diagnosis and polyhydramnios had no impact on outcome. When effusions were bilateral, outcome has been worse in some studies but not in others (1,3,6,7).

The content of the isolated pleural effusion is mostly chylous, resulting from a malformation or leakage in the fetal thoracic duct. However, in a minority of the ca-

ses, the content of the effusion is serous. Some authors reported that serous congenital pleural effusion may be associated with underlying thoracic cause such as primary lymphangiectasia, congenital cystic adenomatoid malformation, bronchopulmonary dysplasia, diaphragmatic hernia, chest wall hamartoma and pulmonary vein atresia (8-10). Several cases have been reported about congenital or fetal pleural effusion with chromosomal anomaly such as Down syndrome and Turner syndrome (11-13). Most of these pleural effusions were chylothorax or associated with hydrops (14,15). However, like one of our cases, isolated non-chylous pleural effusion has been rarely reported (16,17). Hence, karyotyping is indicated in a fetus or newborn with isolated pleural effusion for the evaluation of associated chromosomal anomaly (11).

Pleural fluid analysis is helpful in determining the etiology of effusion, either infectious or non-infectious. Pleural pH is a useful marker of differentiation between transudative and exudative effusions. Pleural fluid with a pH less than 7.3 is classified as exudative and is most likely infectious, whereas a pH greater than 7.45 categorizes the fluid as transudative. Additionally, high white blood cell counts with neutrophil predominance and fluid culture are other supportive findings for infectious pleural fluid. Although *B. gladioli* is mainly known as a plant pathogen, the spectrum of infections caused by *B. gladioli* includes respiratory tract infections and septicemia (18). The origin of this microorganism in our second case is not clear. Congenital infectious pleural effusion with PPRM history and identification of the microorganism from both thoracentesis and blood culture yielded us that this gram-negative microorganism was responsible from all profiles.

For a long time, the rarity of this condition and its unpredictable clinical course prevented a uniform approach to management, and indeed, gave rise to a great deal of uncertainty about the usefulness of any prenatal intervention for fetal pleural effusion. The meta-analysis by Weber and Philipson was the first to lend support to the idea of prenatal intervention for fetuses with pleural effusion (6). The prenatal treatment to the fetus include thoracentesis, pleuro-amniotic shunting and pleurodesis (1,19,20). Despite these in utero interventions in selected cases, management of pleural effusions in neonates should be also conservative depending on the clinical course of the baby, if possible.

In conclusion, variable etiologic factors should be investigated in cases with fetal pleural effusions and necessary interventions should be performed in selected cases both in utero and ex utero.

CONFLICT of INTEREST

None declared.

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