

Non-Chylous Congenital Pleural Effusion in New-Born

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Abstract

In neonates, congenital isolated pleural effusion is considered one of the rare causes of respiratory distress. Chylous effusion is the commonest presentation. We report a case of non-chylous congenital idiopathic pleural effusion, and we will discuss the background diagnosis and management.

Keywords: Congenital, Idiopathic, Pleural, Effusion, Non-chylous, New-born, Down syndrome, Trisomy 21, Respiratory distress, Hydrothorax

Introduction

Pleural effusion is defined as fluid accumulation in the pleural space. This potential space exists between the parietal pleura lining the chest wall and the visceral pleura around the lung. (1) The incidence ranging from 1/10,000 to 1/15,000. (2,3) There is a wide spectrum of aetiology ranging from idiopathic, chromosomal anomalies, inflammatory association, or structural malformations. (4) It is usually chylous. (5,6) Currently, there is no sufficient data regarding the incidence of isolated pleural effusion associated with inflammation or genetic syndromes. The aetiology can be classified in primary and secondary aetiologies. The primary causes could be hydrothorax or chylothorax. Secondary causes usually associated with hydrops fetalis either immune or non-immune. Non-immune causes usually associated with infection, bronchopulmonary sequestration, congenital heart disease, congenital lung lesion, chromosomal - genetic syndromes or congenital diaphragmatic hernia. (7)

Case report

A term baby girl 40+1 weeks of gestational age, weighing 3 kg, appropriate for gestation, was born by Kiwi assisted vaginal delivery done for foetal distress to a 25-year-old booked mother G2P1 with "O" positive blood group. The antenatal scan at 12 weeks of gestational age showed the nuchal translucency thickness at the upper normal limit (2.5 mm). The Foetal anomaly scan at 20 weeks of gestational age was reported normal. The repeated antenatal ultrasound at 31 weeks was normal but the antenatal scan at 39+5 weeks showed pleural effusion. Baby required resuscitation and intubation at birth and the Apgar scores were 5 and 7 at 1 and 5 minutes, respectively. She was in severe respiratory distress after resuscitation in the form of tachypnoea, with subcostal and intercostal recessions and breath sounds were diminished over the left hemithorax with right sided apex heartbeats auscultated for which she was admitted to the neonatal intensive care unit. She was observed with dysmorphic features in the form of upward slanting eyes, short neck, sandal gap deformity sign [Figure 1], but there was no signs of hydrops fetalis. Rest of the systemic examination was normal. The blood gas analysis revealed severe mixed acidosis with pH 6.8, PaO₂ 90 mmHg, and PaCO₂ 113 mmHg, HCO₃ and BE were unrecordable. The baby was connected to conventional mechanical ventilation; IV fluids and antibiotics were charted but there was progressive worsening with increasing Fio₂ requirements up to 100%. An urgent bedside chest X-ray was done revealed left side pleural effusion with mediastinal shift of the airway and heart to the right side [Figure 2]. Echocardiography was done and showed large sized patent ductus arteriosus with severe persistent pulmonary hypertension. Chest tube was inserted which drained straw-coloured fluid around 100 ml.

Following pleural drainage, the infant improved remarkably and was extubated successfully to nasal CMV after 2 hours. Repeat X-ray chest showed full expansion of lungs at 24 hours of age [Figure 3], and ultrasonography (USG) chest that was done showed no pleural effusion on the left side. Feeding was initiated on day 2. About 130 ml of fluid was drained over the next 2 days, which remained clear and non-chylous. On day 4 of life, the intercostal drain was removed.

Analysis of pleural fluid revealed transudate nature of the fluid containing sugar 118 mg/dl, protein 4 g/l, Cl- 99 meq/l, cholesterol 15 mg/dl, triglycerides 12 mg/dl, lactate dehydrogenase 100, white blood cells 4750 cells/ul, polymorph nuclear 5%, mononuclear cells 95%; no microorganisms were seen on gram staining and pleural fluid culture was sterile, indicative of non-chylous pleural effusion. The cytology slide reveals abundant lymphocytes admixed with mesothelial cells and cell debris. No malignant cells are seen, the cytomorphological features are consistent with chronic inflammation. The sepsis screen showed mildly elevated CRP, but the repeated CRP after 48 hours was normal. Baseline renal and liver function tests were within normal limits, the abdominal and skull were normal as well. The diagnosis of trisomy 21 was confirmed by karyotyping that was reported as 47-XX+21. A diagnosis of congenital idiopathic left side non-chylous pleural effusion was established. Feeding was started on day 3 and was upgraded gradually as tolerated to full feed. Full recovery was achieved with no recurrence of pleural effusion, and she was discharged on day 10 of life.



Figure 1: Sandal gap sign

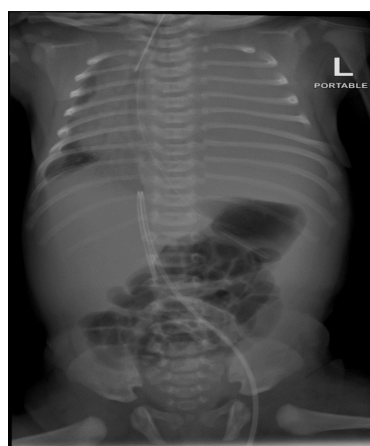


Figure 2: Chest X ray with left sided pleural effusion with mediastinal shift.



Figure 3: Chest X ray after drainage of pleural effusion.

Discussion

Congenital idiopathic pleural effusion is considered a rare cause of respiratory distress in neonates but still well described. It is defined as any effusion in a new-born (age less than 30 days), without any obvious explanation. (8, 9) In the retrospective study by Rocha et al, pleural effusions were congenital in 32% and acquired in 68%. (10)

The aetiology of pleural effusion in the new-born includes immune and non-immune hydrops, syndromes associated like Turner and Down syndromes, chylothorax, congenital heart disease, and congenital pneumonia. (11) Idiopathic chylous pleural effusion is frequently seen and usually on the right side. It is well known that after establishment of external feeds containing fat, simple effusions are known to turn chylous. (8, 12) However, in the current case the pleural effusion was antenatally diagnosed on the left side, postnatally after drainage it was straw coloured, and the fluid analysis showed its non-chylous nature.

Furthermore, the correlation between congenital pleural effusion and chromosomal abnormalities especially Down syndrome was discussed in many studies. According to these studies, the prognosis is highly variable, ranging from spontaneous resolution up to progression to foetal hydrops and eventual perinatal death. (13, 14, 15, 16) Compared to a previously published reports the current case developed the pleural effusion late in pregnancy and was not associated with pulmonary hypoplasia. At the time of birth developed severe respiratory distress and acute respiratory failure while on adequate invasive respiratory support however, a marked improvement was achieved only after chest tube drainage, and she was weaned from invasive ventilatory support within 2 hours.

Strategies for treatment of foetal pleural effusion are based on presentation and symptoms rather than the underlying causes. On one hand, conservative management and spontaneous resolution is expected with small volumes of pleural fluid without hydrops. Aubard et al. reported the survival rate for conservative treatment when hydrops present or not was 24% and 75% respectively. (17) Similar survival rates of 35% and 73%, respectively were reported by Rustico et al. (18) Indeed, Survival rates have improved significantly in recent years. Wada et al. reported the survival rates were 58% and 97.8%, respectively, that can be largely attributed to improved methods of neonatal treatment. (19)

On the other hand, foetal thoracentesis is sometimes needed to reduce respiratory distress and improve foetal pulmonary development, but in many patients, the procedure must be repeated after 24-48 hours, so thoracoamniotic shunting is usually recommended for fetuses with hydrops. (20) Thoracentesis is indicated when there is a progression of hydrops with rapid enlargement of a pleural effusion associated with mediastinal shift. Recently, around 60% survival rate for congenital hydrothorax with hydrops is reported for patients treated with thoracoamniotic shunting, from 35% to about 60% for those receiving conservative treatment and approximately 50% for those treated with thoracentesis. (18,19) In the current case, the thoracoamniotic shunting was not offered because of late diagnosis, no signs of hydrops, and the fluid volume was static.

Although in utero thoracentesis was not performed, but it was urgently needed after delivery to relieve the severity of respiratory distress and failure with an excellent prognosis after the procedure.

Conclusion

Although congenital pleural effusion is a rare cause of respiratory distress in neonates; still it should be kept in the differential diagnosis. There is a wide range of causes of congenital idiopathic pleural effusion and one of them is chromosomal anomalies associated plural effusion. Moreover, the management depends on the severity of symptoms rather than the aetiology. The prognosis depends on the associated hydrops, rapid enlargement of pleural effusion and presence of pulmonary hypoplasia.

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Statement of Ethics

Written consent was taken from the parents.

Conflict of Interest

The authors declare no conflict of interest.

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Author Contributions

Dr. Ahmed Elmelhat: conception, data acquisition, manuscript drafting, critical analysis, final approval. Dr. Maged Ahmed: conception, manuscript drafting, critical analysis, final approval. Dr. Mohammed Nabil: data acquisition, review, critical analysis, final approval. Dr. Hani Mohamed: analysis, review, design, critical analysis, final approval.

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