Isolated non-chylous congenital pleural effusion in a neonate – case report and review of literature on current prenatal and postnatal management

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Abstract

Respiratory distress is a leading cause of mortality and long term morbidity in neonates. Hyaline membrane disease, meconium aspiration, pneumonia, diaphragmatic hernia and cystic adenamatoid malformations are the commonest cause of respiratory distress in pre-term and term babies respectively. Pleural effusions are a rare cause of respiratory distress in new born babies. Most of these effusions are chylous. Chylous effusions are mostly seen in babies with Down's syndrome. Non-chylous effusions are rare and early recognition is crucial because it has an excellent prognosis on early diagnosis and treatment. A case of congenital non-chylous pleural effusion in a baby born in Brunei Darussalam is described and its clinical significance discussed.

Key Words: *chylothorax, pleural effusion, thoracentesis*

Introduction

Congenital pleural effusions are extremely rare in neonates. Most of the reported cases of congenital pleural effusions are chylous [1]. Antenatal diagnosis is important as early neonatal diagnosis and intervention could reduce mortality and morbidity. Adequate respiratory and hemodynamic support and drainage of the effusion gives excellent results in non-chylous effusions. Chylothorax is treated with thoracentesis and feeds with medium chain triglyceride containing formula. We report our experience with a case of congenital non-chylous pleural effusion in a baby that was diagnosed in an antenatal ultrasound scan.

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Case Report

A term female baby weighing 3100g was born to a gravida 2 mother by caesarean section. Mother had regular antenatal care and her serology was negative for HIV, Hepatitis B & C and syphilis and was immune to rubella. Her high vaginal swab culture was sterile. Her fetal ultrasound scans at 18 and 33 weeks were normal. A repeat fetal scan at 36 weeks showed a right congenital pleural effusion [Fig.1].

At birth, the baby cried spontaneously but became dusky one minute after birth. She required bag and mask ventilation followed by endotracheal intubation. The baby was transferred to the neonatal intensive care unit and mechanical ventilation was started. Umbilical catheters were inserted and the baby was stabilized with moderate conventional ventilator settings. A chest X-ray confirmed the prenatal ultrasound finding of a moderately large right sided pleural effusion [Fig.2]. An ultrasonogram of the chest showed the right lung surrounded by fluid with a shift of the mediastinum to the left. Two dimensional echocardiography, renal and head scans were normal. At 7 hours of life, a chest drain was inserted and 80 ml of fluid was drained [Fig.3].

Analysis of the pleural fluid showed an exudative effusion with a protein level of 35g/L, LDH 609 IU/L, triglycerides 0.31mmol/L, WBC 4640/mm3 with 72 % neutrophils and 28% lymphocytes. A repeat pleural fluid analysis on day 6 revealed protein 27g/L, LDH 2554 IU/L, triglycerides 0.59mmol/L, WBC 640/mm3 with 80% neutrophils and 20% lymphocytes. Culture and gram staining of the pleural fluid were negative for pathogens in both samples. Blood counts, C reactive protein, renal panel, liver function tests and coagulation profile were within normal limits. Blood and surface swab cultures were negative. Antibiotics were stopped on day 6. Karyotyping confirmed a 46XX normal chromosomal pattern. Chest X-ray on day 5 showed total clearance of pleural fluid.

By day 6, there was minimal amount of pleural fluid drainage and the chest drain was subsequently removed on day 6 with a total of 140ml of fluid drained. The baby was weaned off the ventilator and extubated at 52 hours of life. She was breastfed on day 4 and was on full feeds on discharge from the unit. A repeat chest X-ray on day 10 [Fig. 4] showed normal lung fields. She was discharged home. On review at 1 and 6 month of age, the baby remained well and asymptomatic with adequate weight gain.

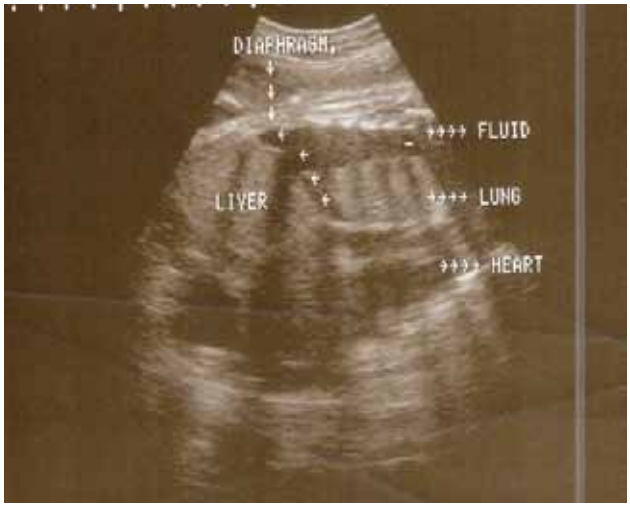

Figure 1. Antenatal scan showing pleural effusion

Figure 2. Xray at 2 hours of life with right sided pleural effusion

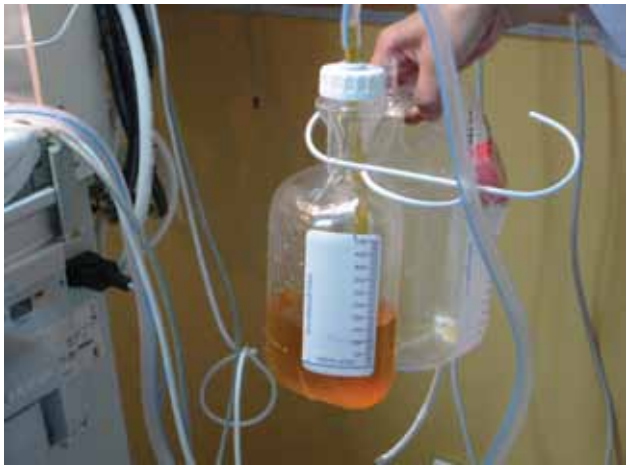

Figure 3. Drained pleural fluid from the right chest

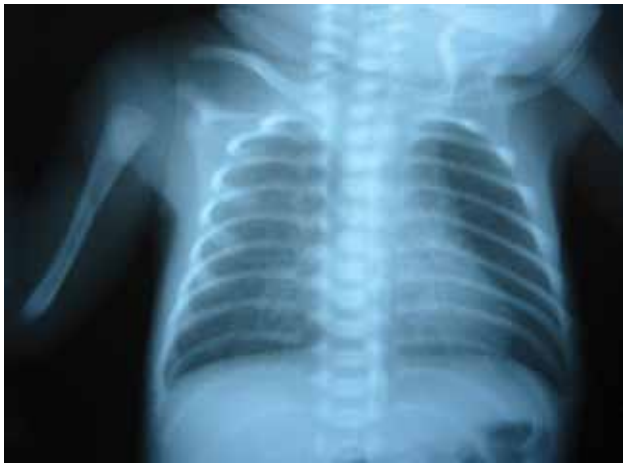

Figure 4. Xray chest on day 10 showing normal lung fields

Discussion

Congenital pleural effusions are rare in neonates and have been reported in 1:12000 to 15000 pregnancies [5]. Causes include hydrops fetalis, intra-uterine pneumonia, cardiac abnormalities, intra-thoracic or cardiac surgery, leakage from long lines and trauma [1, 2, 6].

Most of the congenital pleural effusion fluids are chylous in nature, resulting from a malformation or tear in the fetal thoracic duct and 60% of cases has been found in the right hemithorax [1]. Chyle is noninflammatory, lymphocyte predominant fluid having low LDH and reported as protein discordant exudates [10]. Chylous effusions may be initially serous and turns milky after feeding or initiation of total parenteral alimentation. Chromosomal anomalies like Down's syndrome and Turner's syndrome have been reported in association with neonatal chylothorax [1, 4].

Non-chylous pleural effusions however are rare and have been reported in babies with underlying conditions like primary lymphangiectasia, congenital cystic adenamatoid malformation, bronchopulmonary dysplasia, diaphragmatic hernia and pulmonary vein atresia and rarely Down's syndrome [9]. Non-chylous effusion seen in our case did not isolate any pathogens from blood or pleural fluid. Protein and cell count were high. There was no underlying lung pathology.

Fetal ultrasound scans has increased the number of antenatally detected cases with congenital pleural effusions as in this reported case. Chest radiograph in babies with significant pleural effusion usually demonstrates obliteration of the costophrenic angle and a homogenous haziness in the affected hemithorax with contralateral mediastinal shift [6]. Diaphragmatic hernia with fluid filled loops of gut, congenital cystic adenamatoid malformation or pulmonary sequestration and total atelectasis should be considered in the differential diagnosis. Ultrasonograpy of the chest is a useful non invasive tool for diagnosis of pleural effusion and underlying lung conditions [2, 6, 8].

Thoracentesis and fluid analysis are necessary for the diagnosis. Chylous effusion is milky-white after milk feeds or haemoserous in colour with an absolute cell count of greater than 1000/mm3, lymphocyte fraction greater than 80% and triglyceride level >1.1mmol/l [3]. Non chylous effusion could be a transudate or exudate. Transudates have total protein less than 30g/L and WBC fewer than 2000/mm3 with a predominance of mononuclear cells where as exudates have high protein levels and predominance of polymorphonuclear cells [9]. Exudative fluid may show the pathogen in gram stain or culture. The pleural fluid in our case was an exudate but sterile.

Fetal pleural effusions, if diagnosed antenatally, need to be followed up closely. These effusions may undergo spontaneous resolution or increase over time resulting in tension hydrothorax [4]. Tension hydrothorax will require intra-uterine thoraco-amniotic shunting in order to prevent pulmonary hypoplasia or even intra-uterine death. Neonates born with moderate to large effusions require early and active intervention. Intubation and mechanical ventilation will help to resolve early respiratory insufficiency. Thoracocentesis for decompression of the pleural effusion should be performed. Fluid reaccumulation following thoracocentesis should be treated with a chest drain. Antibiotics should be given to all neonates presenting with effusions at birth and continued until an infectious aetiology has been excluded [3].

For congenital chylothoraces, majority of the published literatures support a treatment regimen beginning with a period of parenteral nutrition followed by medium chain triglyceride containing formula. Somatostatin or its analogue octreotide has been reported with success in chylothorax not responding to chest tube drainage and bowel rest [7].

In conclusion, early recognition of neonatal pleural effusion is crucial because of the excellent prognosis following timely diagnosis and treatment.

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