CASE REPORT

A case report of congenital chylothorax

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SUMMARY

Pleural effusions in a neonate are generally congenital in about one third of the cases and acquired in the remaining two thirds. The incidence of congenital chylothorax is 1 in 12000 to 1 in 15000 making it rare. This case describes the complexity and challenges that are faced in diagnosing a case of congenital chylothorax in the newborn. The article also highlights the current management as well newer forms of management for congenital chylothorax.

INTRODUCTION

The thoracic duct function is to transport close to 70% of ingested fat at a concentration of 0.4–6 g/dl from the intestine to the circulatory system. A total of 2.4 L of chyle is transported through the lymphatic system.[1] Chyle is constituted by cholesterol, triglycerides, chylomicrons and fat-soluble vitamins. In addition to this, chyle is also made up of lymph, which consists of immunoglobulins, enzymes, digestive products and between 400 and 6800 white blood cells/ml.[1]

Chylothorax is characterised by the accumulation of chylous fluid in the pleural space. It is the most common cause of pleural effusion in the foetus and neonates. The causes of chylothorax can broadly be classified into traumatic as well as not traumatic.

The presentation can range from asymptomatic cases to non-immune hydrops fetalis. Congenital chylothorax (CC) can lead to poor lung development, which in turn leads to respiratory distress in the newborn period.[2] Occasionally CC has been associated with certain syndromes. In most cases, the outcome is a favourable prognosis, except in hydropic neonates.

The authors would like to present a case of CC that was managed medically and has been on regular follow-up with any complications.

CASE PRESENTATION

A 22-year-old Malay woman, gravida 2 with a maternal history of being overweight, and previous admission at 28 weeks for leaking liquor, underwent emergency caesarean section (EMLSCS) due to poor progress. The newborn delivered was a late premature at 36 weeks with a birth weight of 3380 g. APGAR score was 9 in 1 minute and 10 in 5 minutes with stable vitals. He was then admitted to the post-natal ward and started breastfeeding.

On day 2 of life, the patient developed physiological jaundice and was admitted to special nursery care (SCN) in Hospital Sultan Abdul Halim (HSAH). Phototherapy was initiated. On day 5 of life, it was noted that the patient was tachypnoeic (respiratory rate; RR > 70/minute) associated with subcostal recession. It was noted that the patient had been given his scheduled feeding an hour before. He was supported with nasal prong oxygen, and his saturation ranged between 90% and 91%. Prior to transferring him, he was kept nil-by-mouth and was presumptively covered for aspiration pneumoniae according to HSAH NICU protocol.

In NICU, the oxygen support was increased to nasal continuous positive airway pressure (nCPAP). A chest x-ray revealed left lower zone opacity/consolidation (Figure 1).

He was started on full intravenous maintenance fluid (IVD). Feeding was resumed the following day with close blood sugar monitoring. A trial of weaning the patient to nasal prong oxygen was unsuccessful as the patient developed respiratory distress in the form of subcostal recession and tachypnoea. He was then placed back on NcPAP for oxygen support.

On day 7 of life, patient had on-and-off desaturation associated with subcostal recession. Spo2 ranged between 82% and 83% under NcPAP support. He was thus intubated and was supported with synchronised intermittent positive-pressure ventilation (SIPPV). A repeated CXR post-intubation was done, demonstrating left pleural effusion and right loculated pneumothorax (Figure 2). Subsequently, an ultrasound (USG) thorax was done, showing a left huge simple pleural effusion.

Based on the USG findings, we revised our diagnosis to CC. We continued the feeding according to the age.

A left intercoastal chest drain (ICD) was inserted, and an initial drain of 70 ml of milky yellow was drained. Subsequent CXR showed there was marked resolution of the left-sided pleural effusion/collection after insertion of the left-sided chest drain (Figure 3).

The pleural fluid resulted in exudative pleural fluid. The pleural fluid was sent for further analysis which resulted in a yellowish coloured jelly like consistency with scanty amount of white blood cells. The labarotary analysis also revealed albumin, lactate dehydrogenase, protein as well as triglycerides.

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Table I: Summary of feeding and ventilation support.

Days	Feeding	Respiratory status
0–2	Breastfeeding on demand	Self-ventilating in air
2–4	Feeding according to age via cup	Self-ventilating in air
4–5	Nil by mouth	NcPAP Fio_2 30%, PEEP 5cmH_20
6–7	Started on half feeding and was increased to full feeds	NcPAP Fio_2 25%, PEEP 5cmH_20
7	Full feeding	SIPPV FiO_2 pressure 20/5
8–9	Nil by mouth	SIPPV FiO_2 pressure 20/5
9–2 months of life	MCT formula milk (basic F formula)	SIPPV FiO_2 pressure 18/5

Table II: Differences between congenital chylothorax, simple pleural effusion and pneumothorax

Characteristics	Congenital chylothorax	Pleural effusion	Pneumothorax
Definition	Accumulation of lymph in the pleural cavity	Accumulation of extra fluid around the lungs and the membranes around the lungs	Abnormal accumulation of air in the space between the thin layer of tissues that cover the lung and chest cavity
Clinical Findings	*(Depends upon the rate of chyle loss)* - hypovolaemia - respiratory difficulty secondary to pleural space fills with fluid - dyspnoea - malnutrition due to loss of proteins, fats, and vitamins	- respiratory distress - dyspnoea - dullness to percussion - decreased breath sounds	- reduced chest expansion - reduced air entry - hyper resonant on percussion
Radiology findings	Chest X-ray - homogeneous density obligating costophrenic angle and cardio phrenic angle Thoracic ultrasound - isodense echoic region without any septation or loculation Chest CT scan - low-attenuation tubular area in the posterior mediastinum MRI - shows cisterna chyli	Chest X-ray - blunting of costophrenic and cardio-phrenic angle - fluid within the horizontal or oblique fissure Thoracic ultrasound - Definite is by identifying the quad sign sinusoid sign Chest CT scan	Chest X-ray - hyperlucent hemithorax sign in case of anterior pneumothorax - medial stripe sign in case of medial pneumothorax

In view of the pleural fluid biochemistry confirming the diagnosis of CC, we kept the patient NBM with IVD maintenance according to age.

He was successfully extubated on day 4 of chest tube insertion and was weaned to room air support the following day.

He was kept NBM, till the availability of medium chain triglycerides (MCTs) based formula milk. On day 4 of chest tube insertion, we were able to restart the feeding for the patient with MCT-based formula.

Clinically the patient's condition showed improvement evident by the successful extubation of the patient; however, chest drainage was still approximately 30 ml/kg/day of chyle.
Hence, it was decided to start the patient on octreotide infusion at 1 mcg/kg/min and was titrated to a maximum dose of 2 mg/kg/h.

The addition of the octreotide infusion resulted in the reduction of the intercostal drainage to which allowed for the removal of the intercostal drain on day 17. The drain was removed after 17 days. He was continued on MCT-based formula for 6 weeks after which we introduced normal milk at 2 months of life (Table II). The introduction of formula milk was well tolerated by the patient, and he has been growing well. We are currently still following up in our outpatient paediatric clinic. At the first consultation in the paediatric clinic after discharge, a CXR was done, and it was noted there was no residual right pneumothorax present.

DISCUSSION

Congenital chylothorax is the most common form of pleural effusion during the new-born period. CC is defined as the accumulation of lymph in the plural cavity.[2] Criteria for the diagnosis of CC are the following: pleural fluid protein concentration >20g/L, triglyceride concentration > 100 mg/dl, number of cells per millilitre >100 with lymphocyte predominance and sterile culture.

The presence of milky appearance of the fluid with positive Sudan III test results is diagnostic in orally fed infants.[3] CC

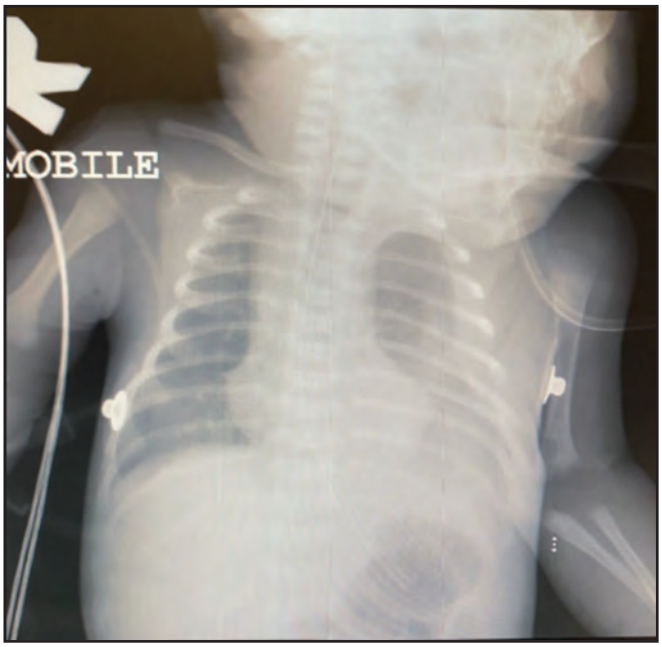

Fig. 1: A chest x-ray of the neonate given oxygen supplementation revealed left lower zone opacity/consolidation

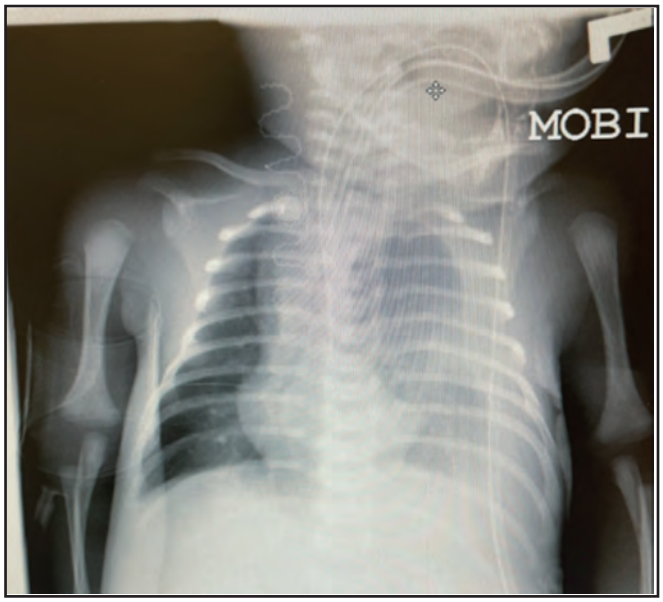

Fig. 2: Repeat CXR several days later (post intubation), revealed that there was a large left-sided pleural effusion, likely increasing in size from birth and a small loculated pneumothorax at the right lower zone. Thus, a provisional diagnosis of congenital chylothorax was made

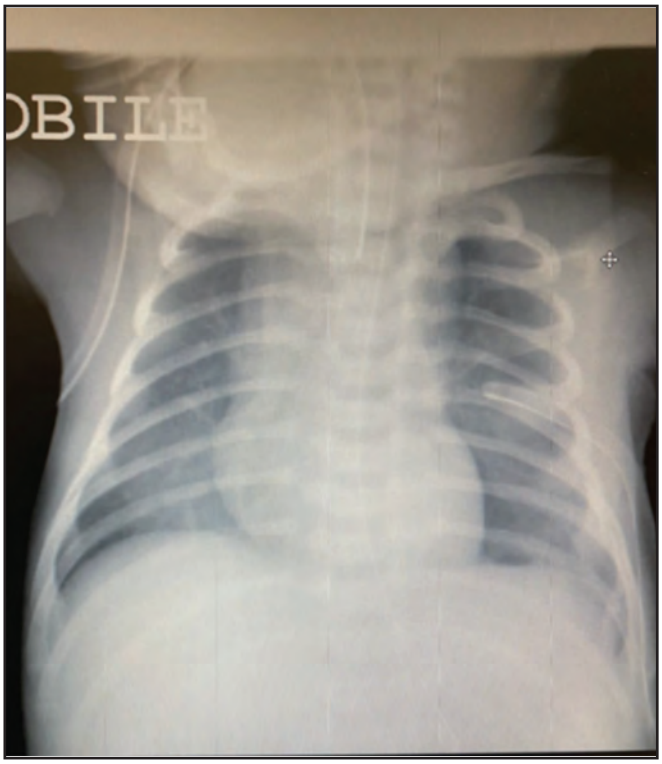

Fig. 3: CXR showed there was marked resolution of the left sided pleural effusion/ collection after insertion of the left-sided chest drain

can be idiopathic due to congenital lymphatic malformations, atresia or hypoplasia of the thoracic duct. It can also associate with syndromes such Down's, Turners, and Noonan.

The main goal of the treatment for CC is removing the chyle, preventing re-accumulation, managing the complications, and looking for the underlying aetiology. If not treated appropriately, it is a potentially a life-threatening disorder that can lead to serious respiratory distress, metabolic disorder, immunodeficiency, and nutritional complications.[4] The percentage of mortality increases depending on associated findings, gestational age and the duration and severity of chylothorax.

One of the most challenging aspects is the initial diagnosis of CC. The diagnosis requires a high degree of suspicion as radiographically the appearance may mimic pleural effusion or pneumothorax. The main diagnostic tool is thoracic ultrasound. Mainly in CC, the USG thorax will appear as an isodense echoic region without any septation or loculation. The differences between CC, pleural effusion and pneumothorax are summarised in the table below (Table II).

The management of CC can be conservative or surgical. The conservative approach involves replacing lost nutrients, draining the accumulated chyle and administering low-fat medium triglycerides (MCTs) orally.[5] By introducing MCT formulation, we bypass the intestinal lymph system, and in return, this reduces the flow of chyle into the thoracic duct, which in turn allows it to heal. The success rate of the MCT diet in CC is up to 75%.[2] However, failure of the MCT

formulation to plug this gap may result in complete nil-by-mouth and full total parental nutrition being initiated.[5] In addition to the MCT formulation, the infusion of octreotide has been proven helpful as a tool in the conservative approach. The octreotide can be administered either via venous infusion or subcutaneously. Being a somatisation analogue, the splanchnic blood flow is reduced by mild vasoconstriction, leading to less intestinal secretion and absorption. Hence, this decreases the thoracic duct flow.[2]

Surgical intervention is often required if the drainage from the ICD is more than 100ml/body weight/day or if the chyle flow is present for more than 2 weeks. Another indication for surgical intervention is the rapid decline in nutritional status despite conservative management.

Another new approach to the management of CC cases is by administrating Picibanil (OK-342) during the in-utero period. Tanemura et al. reported the usage of Picibanil (OK-342) in a patient who was scanned at the 20th week gestation period and showed severe pleural effusion, ascites, skin oedema and polyhydramnios. The Picabanil (OK-342) was administered in utero at 23, 24 and 25 weeks of gestation. The pleural effusion started to subside by 28th week of gestation and completely disappeared by the 34th week of gestation [6]. This resulted in the delivery of a healthy neonate.

Conversely, CC cases that persist for more than 2 weeks or have a high-volume leak of more than 1000–1500 ml/day, are usually managed surgically. The surgical techniques include thoracic duct ligation, mass ligation of the tissue, pleurodesis and pleuroperitoneal shunting. The surgical method that is most preferred is the ligation of the thoracic duct, as it has a higher rate of success and a lower rate of failure.

CONCLUSION

CC is a diagnostic challenge for the neonatologist. The presentation may closely resemble other common lung pathology during the neonatal period. However, an early diagnosis with prompt intervention can determine the course of management and improve outcomes. In most reported cases, a favourable outcome can be achieved with a conservative approach. We report one such case.

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