

Case Report

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Congenital chylothorax: a rare entity

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ABSTRACT

Pleural effusions in a neonate are generally congenital in about one third of the cases and acquired in the remaining two thirds. Congenital isolated pleural effusion is rare. It has an incidence of approximately 1 in 12000 to 1 in 15000 pregnancies. Chylothorax is the most common cause of neonatal congenital pleural effusion. Incidence of congenital chylothorax is 1 in 8600 to 1 in 10000 deliveries with a male to female ratio of 2:1. It poses both a diagnostic as well as therapeutic challenge to the neonatologist. Authors hereby present a rare case of congenital chylothorax which was medically managed and discharged. The neonate responded well to octreotide and medium chain triglyceride (MCT)-diet and was discharged without any complications.

Keywords: Chylomicron, Congenital chylothorax, Congenital pleural effusion, Octreotide

INTRODUCTION

Pleural effusion is a rare condition in neonates. Pleural effusion (accumulation of fluid in the pleural space) may be due to an increase in the rate of filtration or a reduction in the clearance by the lymphatics or even both.¹⁻³ It can be either congenital or acquired. The causes for congenital pleural effusion include fetal hydrops, isolated congenital chylothorax, congestive failure, coagulopathy, intrathoracic neoplasm or it may be even idiopathic in up to 50 percent of the cases.

Acquired pleural effusion may be either due to postoperative damage to the thoracic duct, leakage of total parenteral nutrition, pneumonia, congestive heart failure; nephrotic syndrome or at times the reason may be unknown. Pleural effusion can be diagnosed in the antenatal or in the neonatal period. It can be asymptomatic or may present with respiratory distress. Chylothorax, which results from accumulation of chyle in the pleural cavity may be due to either damage or obstruction of the thoracic duct. It has a favorable prognosis generally, except in hydropic neonates.⁴

Authors present a rare case of congenital chylothorax which was medically managed and is being regularly followed up without any complications.

CASE REPORT

Seven days old, 35weeker late preterm male child with a birth weight of 2.9 kg was born of non-consanguineous marriage by caesarean section to a 38-year old multigravida. Her antenatal scan at 32 weeks of gestational age was suggestive of polyhydramnios and left sided pleural effusion. He was brought by parents to another institute with the chief complaint of rapid breathing since birth. He was investigated for this respiratory distress and the antenatal finding of left sided pleural effusion was confirmed by a chest x ray chest and ultrasound of the thorax. Therefore, an intercostal chest drain (ICD) was inserted. He improved over a period of two days and hence the drain was removed. However, he soon developed respiratory distress and thus was referred to our institute. On arrival to our institute; he had severe respiratory distress that needed mechanical ventilation (Figure 1). He was not hydropic and had no dysmorphic

features. Also, rest of his examination including anthropometry was within normal limits.



Figure 1: Clinical picture on admission.



Figure 2: Chest X-ray left sided pleural effusion.

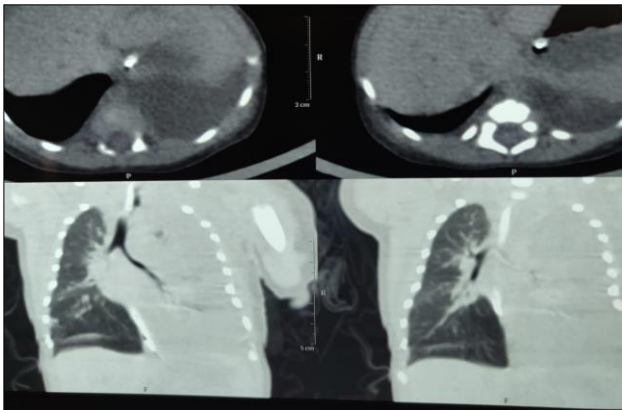


Figure 3: CT-Scan left sided gross pleural effusion with a mediastinal shift.

A chest X-Ray and subsequently a contrast-enhanced computed tomography (CECT) of the chest (Figure 2 and 3) were done, which were suggestive of re-accumulation of left sided pleural effusion resulting in a mediastinal shift. Thus, an ICD was reinserted and the collected fluid (Figure 4) (that was yellowish in colour) was sent for investigation. The pleural fluid was reported to have raised triglyceride levels and chylomicrons, clinching the diagnosis of congenital chylothorax. He was also

evaluated for congenital tuberculosis (pleural fluid gene expert for Tuberculosis and Adenosine deaminase (ADA) levels were normal). An echocardiography and ultrasound of the other body cavities were also normal, thus excluding hydrops. There were no malignant cells in the pleural fluid. The karyotyping for our neonate was normal; although quadruple marker screening test in the antenatal period was positive for Trisomy 21.

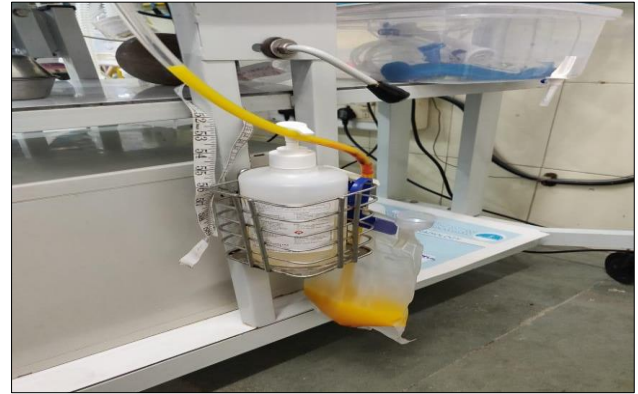


Figure 4: Pleural fluid on admission.

Our patient was meanwhile kept nil per oral and was tried on conservative management. However, as there was no clinical response in the form of reduced drain output for more than a week, he was started on intravenous continuous infusion of injection octreotide at a dose of 1 µg/kg/hour with strict blood sugar monitoring in order to avoid hyperglycaemia. Following this, there was a gradual decrease in the ICD output within ten days. Feeds were introduced once the output had significantly reduced within ten days. Medium chain triglyceride (MCT) oil was added to the feeds and it was decided to be continued for at least four to six weeks. Although, a lymphangiogram was planned, it was not required later as there was no drain output following octreotide infusion. Since he tolerated feeding and was growing well, he was discharged on 58th day of life. He has been following with us in the outpatient department after discharge and has had a satisfactory weight gain.

DISCUSSION

Congenital chylothorax (CC) is the most common form of pleural effusion in a newborn. CC is defined as accumulation of lymph in the pleural cavity. Its incidence is about 1 in 10,000 live births. The mortality in CC may vary between 20 to 50 percent.⁵⁻⁸ Chylothorax that occurs in the foetus can lead to pulmonary hypoplasia (pressure effect), congestive heart failure and even hydrops (hypoproteinaemia and impaired venous return). When chylothorax is associated with hydrops, mortality can be as high as 98%.^{5,6,9} Its a etiology may be idiopathic or may be due to congenital malformation of the lymphatic system such as lymphangiomatosis, lymphangiectasia or cystic hygroma. There are various genetic conditions such as Down's syndrome, Turner syndrome and Noonan

syndrome associated with fetal chylothorax, thus affecting their prognosis.^{8,10,11}

Table 1: Criteria for diagnosis of chylothorax.¹²

Characteristic	Description
Appearance	Clear yellow (milky with fat-containing feeds)
Cell count	>1000cells/mm ³
Lymphocyte proportion	>80%
pH	7.4-7.8
Triglycerides	>1.1mmol/L
Cholesterol	65-220mg/dl
Albumin	1.2-4.1g/dl
Total protein	2.2-5.9g/dl

Thus, it is seen that lipids (60% to 70% of ingested fat absorbed by intestinal lymphatics) form the major component of chyle. Other components are protein, fat soluble vitamins, antibodies, urea nitrogen and enzymes. The effusion may be either unilateral (right or left sided) or bilateral but unilateral effusion is more common than bilateral effusion and is dependent on the location of the trunk. The level at which thoracic duct gets damaged decides the side of effusion. Lesions above the 5th thoracic vertebra generally lead to left sided effusion whereas damage below that result in right sided effusion. Our neonate had left sided effusion. As CC is a rare condition, its management is also challenging.

Most of the cases of chylothorax respond well to conservative management using parenteral nutrition and medium chain triglyceride (MCT) rich formula feeds. MCT bypasses the lymphatic drainage and gets directly absorbed in the portal system without processing to chylomicrons. The success rate of conservative treatment (MCT-diet) in CC is up to 75%.¹³ However, our case responded well to octreotide with MCT-diet. Octreotide is a somatostatin analogue that reduces the splanchnic blood flow by mild vasoconstriction. This causes less intestinal secretion and absorption) which in turn may decrease the thoracic duct flow. It can be administered subcutaneously in a dose of 10-70 µg/kg/day given 6-24 hourly, or as an infusion at 1 to 10 µg/kg/hour.¹⁴ Authors had given inj octreotide infusion at the rate of 1 µg/kg/hour for 10 days. Once the chest drainage is controlled, octreotide infusion is gradually weaned off over several days. This newborn responded over 10 days. While on therapy, serum glucose, thyroid function test and liver enzymes need continuous monitoring. Apart from these, supportive care such as protein replacement, electrolyte supplementation and adequate calories also plays an important role. Moreover, care needs to be taken to avoid infections as they are vulnerable to secondary immunodeficiency caused by loss of immunoglobulin in the pleural fluid.

Complications as discussed above may include pulmonary hypoplasia (in cases of foetal chylothorax),

malnutrition, hypoproteinaemia, fluid electrolyte imbalances and secondary immunodeficiency. Our case had Klebsiella pneumonia sepsis needing antibiotics for 14 days. Nevertheless; he had an adequate weight gain with good nutrition support while in the hospital and so could be successfully discharged by about two months of life.

In some cases if the conservative therapy fails to decrease the drainage to <10ml/kg/d till 4 weeks, it is advisable to undergo surgical intervention.¹² These include pleurodesis (chemical using povidone-iodine or rarely surgical), thoracic duct ligation or pleuroperitoneal shunts. Thoracic duct embolization though challenging, may be helpful in thoracic duct blockage with extravasation. In such cases a lymphangiogram may be performed prior to the procedure. Parenteral nutrition needs to be supported throughout this prolonged course. At times while dealing with congenital malformations, even pleurodesis may fail to give a result. A lymphoscintigraphy and magnetic resonance imaging may be useful to identify anatomical abnormalities as well as thoracic duct injuries.

CONCLUSION

Congenital pleural effusion usually occurs as hydrops or congenital chylothorax. It is a diagnostic challenge for the neonatologist. Nonetheless, prompt diagnosis and adequate management of congenital chylothorax are crucial. Clinical outcome is generally good with timely management, except in hydropic neonates. This was a typical case of congenital chylothorax that responded well to medical management. Since there are no universally accepted guidelines for this uncommon condition, more evidence-based studies are needed to formulate the management of such cases.

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