Nonimmune hydrops is a common cause of hydrops fetalis. Here, we report a rare case of congenital chylothorax which presented with nonimmune hydrops and was effectively managed by conservative measures.

Keywords: Chylothorax, hydrops fetalis, neonate

INTRODUCTION

Congenital chylothorax is a rare but important cause of pleural effusion in early neonatal period. There is no definite consensus on the management of this condition at present. Most of the cases are managed by conservative means and surgical intervention is required in cases with persistent effusion. Here we report a case of congenital chylothorax managed effectively by conservative means.

CASE REPORT

A term male baby weighing 3.5 kg was delivered at 38 weeks of gestation to a primigravida mother. Antenatal scan revealed severe polyhydramnios with amniotic fluid index of 38 cm and bilateral pleural effusion in the fetus. The baby was delivered via lower segment cesarean section in view of fetal distress and was then referred to our center at 2 h of life for further neonatal care.

At admission, the neonate had severe respiratory distress with Downes score of 7/10 and was initiated on mechanical ventilation. On examination, there was generalized pitting edema without pallor or icterus. Vitals at admission were heart rate of 170/min with low-volume pulse, respiratory rate of 71/min, and saturation of 80%. No dysmorphic features were noted. Auscultation revealed severe reduction in bilateral breath sounds with normal heart sounds. An emergency chest X-ray and bedside ultrasound revealed bilateral pleural effusion without any pericardial effusion [Figure 1].

In view of severe hypoxemia and hemodynamic instability, emergency pleural tap was done which revealed straw-colored fluid with about 50 ml aspirated from the left side and 20 ml from the right side and a chest drain was inserted on the left side. A diagnosis of hydrops fetalis was made in view of antenatally detected polyhydramnios and pleural effusion. The neonate also had ventricular tachycardia (VT) at 27 h of life which responded to amiodarone and lidocaine and resolved by 75 h of life.

Initial blood investigations revealed blood group of mother and the baby to be O positive and direct Coombs test was negative. Hence, nonimmune hydrops was considered and workup was initiated accordingly.

Pleural fluid analysis showed cell count of 2000/mm³ with 80% lymphocytes and 20% neutrophils. Biochemical analysis revealed triglycerides of 548.8 mg/dL, protein of 2.5 g/dL, and glucose of 85 mg/dL. Rest of the investigations for evaluation nonimmune hydrops is depicted in Table 1.

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At this point, a differential diagnosis of nonimmune hydrops due to chylothorax was considered in view of lymphocytic pleocytosis, elevated protein, and triglyceride content in the pleural fluid. A possibility of hydrops due to VT was also considered; however, since VT resolved with medications and there was no history during antenatal evaluation, chylothorax was kept as the first differential diagnosis.

He was extubated on day 7 of life. On day 8, milky fluid was noted in chest drain which confirmed the diagnosis of chylothorax [Figure 2]. He was subsequently reintubated in view of worsening respiratory distress. Feeds were started from day 4 of life and gradually advanced till 100 mL/kg/day and maintained at this volume in view of edema. The baby was also started on subcutaneous octreotide at 5 µg/kg/day which was subsequently increased to a maximum of 30 µg/kg/day. Oral sildenafil was started in view of persistent chyle output.

In view of increase in chyle volume, the baby was kept nil per oral (NPO) for 10 days and started on total parenteral nutrition. The baby was extubated on day 17 of life. Chyle output stopped after 20 days and feeds were restarted on day 22 of life with special medium-chain triglyceride formula (PEPTAMEN®) and gradually hiked keeping a close watch on drainage from intercostal drainage tube. Octreotide and sildenafil were gradually tapered and stopped over a period of 2 weeks. The neonate was discharged on day 61 of life.

**DISCUSSION**

Congenital chylothorax is an accumulation of chyle in the plural cavity during fetal or neonatal period.[1] The incidence is approximately 1 in 10,000 live births and is one among the common causes of pleural effusion in neonatal period.[1] Congenital chylothorax may be due to abnormalities of lymphatic system or due to congenital lung malformations which can even lead to hydrops fetalis.[2] The initial appearance of chyle in the index case was clear straw-colored fluid and changed to milky appearance after starting feeds. This is due to progressive increase in the concentration of triglycerides in chyle in neonates who are fed.[1,3] Respiratory distress in these neonates after birth is a result of pleural effusion with or without some degree of pulmonary hypoplasia. In our case, it was mainly due to the pleural effusion as antenatal scans, and postnatal imaging did not show evidence of pulmonary hypoplasia. Since chyle contains proteins including albumin, immunoglobulin, fibrinogen, triglycerides, and a large amount of lymphocytes, loss of chyle leads to reduced immune function making the baby susceptible to infections. This was also noted in our case where we encountered culture positive sepsis due to loss of immunoglobulin and lymphocytes from chyle.[4]

**Table 1: Investigations**

| Parameter                        | Value                  |
|----------------------------------|------------------------|
| Hemoglobin (g/dL)                | 15                     |
| Total lymphocyte count (per mm³) | 12,200                 |
| Total serum bilirubin (mg/dL):   | 4.73: 4.55/0.18        |
| Indirect/direct                  |                        |
| Urea (mg/dL)                     | 41.6                   |
| Creatinine (mg/dL)               | 0.38                   |
| Tandem mass spectrometry         | Normal                 |
| CT scan (contrast)               | Patchy ground-glass    |
| Karyotyping                      | 44+ XY                 |
| Echocardiography                 | Normal                 |
| ECG                              | Normal                 |
| TORCH                            | Normal                 |
| Peripheral smear                 | Normocytic normochromic|
|                                  | blood picture          |

CT: Computed tomography, ECG: Electrocardiography, TORCH: Toxoplasma, other agents, rubella, cytomegalovirus, and herpes simplex
Treatment options in these patients include conservative approach, medications, and surgery. Initial management in these cases includes keeping the baby NPO along with initiation of total parenteral nutrition and monitoring of chyle output from chest drain. Keeping the patient NPO decreases the chyle formation and also the lymphatic flow. Octreotide, a somatostatin analog, has been used in the management of these patients. It acts by reducing the blood flow in splanchnic circulation including lymph flow, decreases the secretion of gastric and intestinal secretions, and decreases intestinal absorption ultimately, leading to reduced chyle flow. Even though most of the studies have used intravenous octreotide, subcutaneous octreotide has also been used in many cases up to 70 µg/kg/day.\(^5\) In our case, we used up to 30 µg/kg/day to achieve therapeutic benefit. Another drug used for this baby was sildenafil. Sildenafil prevents the degradation of cyclic guanosine monophosphate by selective inhibition of phosphodiesterase and could thereby facilitate lymphatic growth and/or remodeling.\(^6\) Furthermore, an important step in management is elimination of long-chain fatty acids by administering total parenteral nutrition till volume of chyle reduces followed by slow introduction of diet containing medium-chain triglycerides (MCTs) and protein to supplement calories. MCTs are directly absorbed into portal venous system bypassing the lymphatic drainage.\(^6\)

**Conclusion**

Congenital chylothorax is an important cause of pleural effusion in a neonate and should be considered irrespective of the color of pleural aspirates. It can be effectively managed conservatively without the need of surgical intervention.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initial(s) will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**References**

1. Attar MA, Donn SM. Congenital chylothorax. Semin Fetal Neonatal Med 2017;22:234‑9.
2. Bengtsson BO. Neonatal lymphatic (chylous) disorders. Neoreviews 2013;14:e600‑12.
3. Ball PL, Nethercott S, Beardsall K. Rare case of congenital chylothorax and challenges in its management. BMJ Case Rep 2019;12:e228023.
4. Vaghela P, Mangukiya H. Congenital chylothorax in a late preterm neonate associated with hydrops fetalis and successful treatment with octreotide and pleurodesis with betadine. J Clin Neonatal 2017;6:208.
5. Bellini C, Cabano R, de Angelis LC, Bellini T, Calevo MG, Gandullia P, et al. Octreotide for congenital and acquired chylothorax in newborns: A systematic review. J Paediatr Child Health 2018;54:840‑7.
6. Bagur Krishnamurthy M, Malhotra A. Congenital chylothorax: Current perspectives and trends. Res Rep Neonatol 2017;7:53-63.