Congenital Chylothorax

Phaniraj M Bangli[1], Ramesh Pol[1], Ramesh Hatti[2], Vikram Nagathan[2]
[1]Department of Pediatrics, [2]Department of Pediatric Surgery,
S. Nijalingappa medical college and research center, Bagalkot, Karnataka, India

Abstract

Congenital chylothorax is a rare condition with pulmonary and multiorgan system involvement with no standardized treatment. We describe a case of a 3-month infant who presented with a cough and hurried breathing for 2 months aggravated from 3 days requiring intubation and ventilator support and incidentally was found to have chylothorax which was managed by draining of chylous fluid by chest tube placement.

Keywords: Chylous fluid, chylothorax, infant.

Introduction:

Chylothorax is an accumulation of chyle in the pleural cavity. First described by Dr. Bartolet in the 17thcentury. 'Chyle' is derived from the Greek word "Chylos", meaning juice. Chyle is formed in the lacteal system of the intestines.

The small and the medium chained triglycerides in the diet gets broken in to free fatty acids by intestinal enzymes and absorbed into the portal circulation. However the large molecules of complex long-chain triglyceride unable to be broken combine with phospholipids, cholesterol, and cholesterol esters to form chylomicrons in the jejunum. These molecules get absorbed into the lymphatic system of the small intestine forming chyle. This lymphatic system joined with lymphatic of lower extremities form the thoracic duct later draining into the systemic circulation.[1]

Case report:

A 3-month-old male child with full-term normal vaginal delivery with no history of NICU admission and birth weight of 3.6 kg, presented with a history of cough and hurried breathing for 2 months on and off, aggravated since 3 days with the previous history of recurrent hospital visits for cough and cold treated on regular basis and no history of fever, weight loss, vomiting, poor feeding, trauma or any congenital deformities but with a history grandfather having pulmonary tuberculosis now treated.

On clinical examination, the child had tachypnea with decreased air entry in the left hemithorax and dull note on percussion with no signs of malnutrition or failure to thrive or evidence of trauma. Initially, the child was put on HHHFNC for increased work of breathing, was later in the view of increasing respiratory distress the child was intubated and was put on ventilator 3 for days. An intercostal drain was put on day 1 and a total of 1200ml of chylous fluid was drained over 10 days and drain was removed on the 15th day. The child was started with infusion aminoven along with IV antibiotics (vancomycin and piperacillin-tazobactam) after extubation was started on MCT (medium-chain triglyceride) supplement alternating with breast milk and was advised the same to continue at discharge.

On radiographic imaging showed left-sided effusion, A CT thorax done showed large left-sided pleural effusion.

Corresponding Author:
Dr. Phaniraj M. Bangli
Department of Pediatrics,
S. Nijalingappa medical college and research center, Bagalkot, Karnataka, India

INVESTIGATIONS	Day 1	Day 3	Day 5	Day 5	Day 11
Hemoglobin	10.6				9.5
Total count	13400				12900
Platelet	608000				724000
CRP	1				
Na		137			
K		4			
Cl		103			
Ca		8.8			
Creatinine	0.6	0.6			
Pleural fluid analysis Total count Lymphocytes Mesothelial cells Polymorphs	 15000 98% 2% 0%				
Pleural fluid Proteins Sugars Chloride ADA	 17.2 260 125 3				
Pleural fluid Triglycerides Cholesterol Chylomicrons			 284 45 present	 255 67	
Pleural fluid Z-N stain Leishman stain	 No acid-fast bacilli Smear shows predominantly lymphocytic, mesothelial cells and macrophages				
HIV Montoux	Negative Negative				
Pleural fluid culture	No growth				
Blood culture	No growth				
USG Thorax	Left-sided pleural effusion				
CT Thorax	Left side effusion				

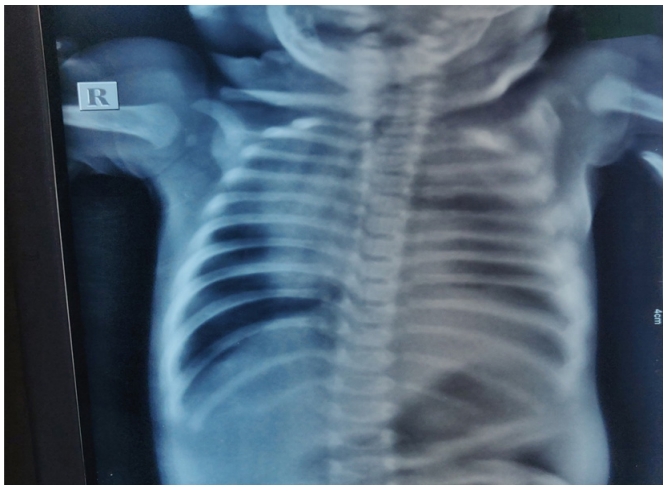

Fig 1: chest x ray showing collection in the left hemithorax.

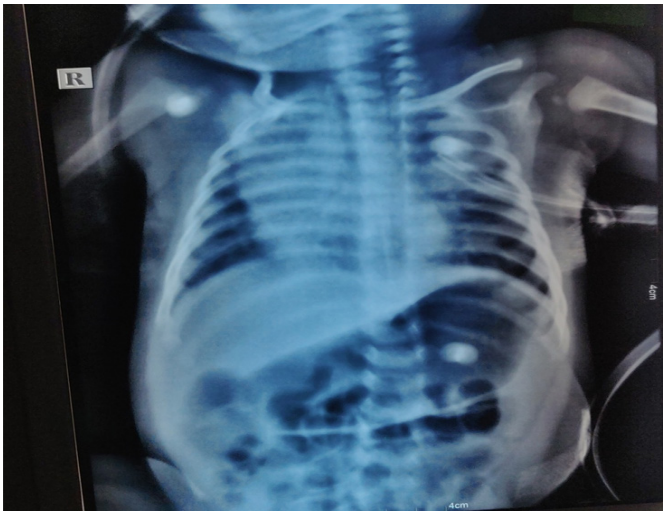

Fig 2: post intercostal drainage insertion.

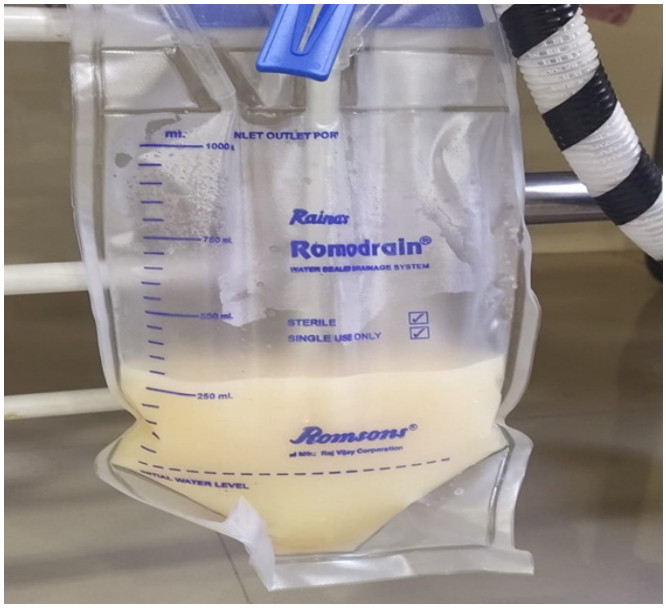

Fig 3: chylous fluid collection in the drain bag.

Discussion:

Cisterna chyli which transports the chyle lies anterior to 2 lumbar vertebras. This duct passes through the esophageal hiatus into the thoracic cavity ascending through the posterior mediastinum right to the vertebral column crossing to the left side at 4-6th thoracic vertebrae. Later, entering the superior mediastinum between the aortic arch and subclavian artery and the left side of the esophagus terminating into the left jugular veins.[2]

Idiopathic chylothorax is the most common type of chylothorax present in infancy due to the weak thoracic duct or abnormalities of the lymphatic system. Therefore, any increase in the venous pressure causes the break in the congenitally weakened thoracic duct. Right-side chylothorax is more common than the left side (55% Vs. 35%).[2] The common mechanism of formation of chylothorax in babies is due to trauma, rupture of thoracic duct by hyperextension of the spinal column, or secondary to increased systemic venous pressure during birth.

CECT being the choice of investigation helps in finding the anatomical cause of chylothorax. To locate the site of leak lymphangiography and lymphangio scintigraphy are the best choice, these investigations were not performed due to feasibility in our case as the baby was ventilated and difficulty in transport along with the cost of the investigation. In general, the idiopathic congenital chylothorax resolves by conservative management andelimination of long-chain fatty acids or replacement with TPN[4], With mortality ranging from 20% to 60% depending on associated findings, duration, and severity. In our case, it was a left-sided chylothorax of unknown cause which resolved with partial parenteral nutrition along with proper respiratory support and placement of chest drain.

Conclusion:

Congenital chylothorax is a very rare cause of respiratory distress in neonates. Diagnosis of this condition is based upon clinical, radiological, and pleural fluid biochemical findings. Medical management with medium-chain triglycerides and partial or total parenteral nutrition and surgery only in resistant cases can be thought of.

References:

1. *Healy H, Gipson K, Hay S, Bates S, Kinane TB. Management and outcomes of congenital chylothorax in the neonatal intensive care unit: A case series. Pediatr Invest. 2017 Dec;1(1):21-5.*
2. *Singh S, Rawat J, Singh G, Chaubey D. Congenital idiopathic chylothorax: A very rare case. J ClinNeonatol. 2017;6(3):205.*
3. *Kirkland I. Chylothorax in Infancy and Childhood: A Method of Treatment. Archives of Disease in Childhood. 1965 Apr 1;40(210):186-91.*
4. *Hermon M, Tenner E, Burda G, Strohmaier W, Schlager G, Golej J. Chylothorax, and Chylous-Like Diseases in Children: Clinical Management. Front Pediatr. 2019 Jun 27;7:*

Conflict of interest: Nil

Source of funding: Nil

Date of submission: December 9th 2020

Date of acceptance: December 28th 2020