

A Case of Idiopathic Congenital Chylothorax in A Premature Newborn

Samet Benli^{1*}, Atika Çağlar¹, Zeynep Hüz², Mustafa Aydın¹

¹Fırat University Hospital, Neonatology Clinic, Elazığ/TURKEY

²Fırat University Hospital, Clinic of Pediatrics, Elazığ/TURKEY

Article Info

Received: September 29, 2022

Accepted: July 15, 2023

Published: July 25, 2023

***Corresponding author:** Samet Benli, Fırat University Hospital, Clinic of Neonatology, Elazığ/TURKEY.

Citation: Samet Benli, Atika Çağlar, Zeynep Hüz, Mustafa Aydın (2023). "A Case of Idiopathic Congenital Chylothorax in A Premature Newborn", *J Pediatrics and Child Health Issues*, 4(1); DOI: <http://doi.org/07.2022/1.1054>.

Copyright: © 2023 Samet Benli. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly Cited.

Abstract

Chylothorax is defined as the accumulation of lymphatic fluid in the pleural space and occurs due to congenital and acquired reasons. Although the etiology of congenital chylothorax is not fully understood, it is thought to be the result of a developmental disorder of the lymphatic system. Some patients are asymptomatic or clinically present with mild respiratory distress, while most cases present with potentially life-threatening respiratory distress if left untreated. Conservative and surgical methods are used in the treatment of chylothorax. Conservative approach includes treatment of the underlying disease, repeated thoracentesis or thoracic tube drainage, total parenteral nutrition with interruption of enteral nutrition, and diet containing medium chain triglycerides. The prognosis in chylothorax varies according to the underlying etiology. With appropriate treatment, the prognosis of congenital chylothorax is generally good. However, the degree of accompanying pulmonary hypoplasia, prematurity and presence of hydrops have been reported as causes that increase mortality.

Keywords: chylothorax; newborn; pleural effusion; respiratory distress

Introduction

Congenital chylothorax in newborns is a serious problem. Congenital chylothorax is defined by the accumulation of abnormal chylous fluid in the pleural cavity and is considered a rare cause of respiratory distress (1). Its incidence is twice as high in males and is seen in 1/10000 infants (2). It is associated with syndromes such as Down syndrome, Noonan syndrome, Turner. However, mostly the underlying pathology cannot be detected and it is defined as idiopathic congenital chylothorax. In the etiology of acquired chylothorax, it usually develops secondary to thoracic duct injury after cardiac (congenital heart diseases) or thoracic surgery (congenital diaphragmatic hernia) in newborns (3). Diagnosis is based on the evaluation of pleural fluid, but is first diagnosed by postnatal direct X-ray or prenatal ultrasonography. Thoracentesis is required for definitive diagnosis. While the fluid taken by thoracentesis in the first postnatal days contains clear and abundant lymphocytes; After feeding, the liquid becomes cloudy and takes on a chylous character. The protein content of the pleural fluid is equal to or less than the plasma protein and tends to collect again after the first thoracentesis. In the treatment, surgical intervention is performed in cases unresponsive to conservative treatment (4). In our article, we wanted to discuss a postnatal idiopathic congenital chylothorax case in a patient followed up for prematurity in the light of the literature.

Case Report

The patient, who was born 750 grams at 27 weeks of gestation from the second pregnancy of a 27-year-old mother, was hospitalized in the neonatal intensive care unit due to prematurity and respiratory distress. The patient was intubated and surfactant was administered by endotracheal route when the direct chest X-ray showed signs of respiratory distress syndrome. On the postnatal third day, due to decreased aeration of the right lung by listening and a hyperdense image on direct graphy, thoracic ultrasonography was performed and pleural effusion was detected (Figure 1). After thoracentesis performed for differential diagnosis, it was observed that the patient's pleural fluid was white and clear (Figure 2). The density of the pleural fluid was 1015 and the pH was 7.8. In the cell count, 4250 leukocytes/mm³ (92% lymphocytes) were detected. In the pleural fluid, protein was found to be 4.2 g/dl, glucose 165 mg/dl, and



triglyceride 1896 mg/dl. Gram stain was unremarkable. There was no growth in pleural fluid culture. His echocardiography did not reveal any pathological findings other than minimal peripheral pulmonary stenosis. Complete blood count examination revealed hemoglobin 14g/dl, hematocrit 52%, leukocyte 17,420/mm³, 54% neutrophils and 46% lymphocytes in peripheral formula. Complete urinalysis was normal. Plasma urea, uric acid, creatinine, electrolyte values, cholesterol, triglyceride, blood sugar values are within normal limits; serum albumin was 3.2 g/dl. No pathology was detected in chromosome analysis, metabolic scans and TORCH scan. In our patient; Congenital chylothorax was diagnosed because clinical and laboratory findings were compatible with chylothorax and there was no risk factor in the postnatal period in the differential diagnosis. Our patient died on the postnatal seventh day due to respiratory distress.

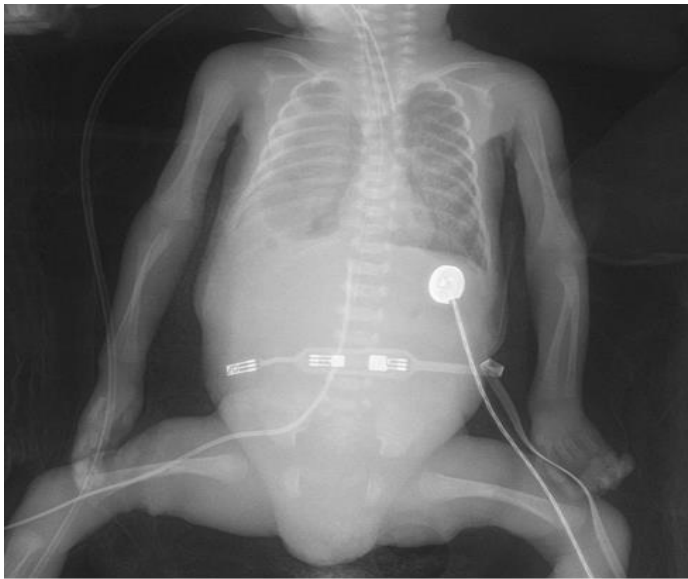


Figure 1. The chylothorax image of the patient at the base of the right lung on the direct radiograph



Figure 2. Image of chylous fluid

Discussion

Congenital chylothorax was first reported in the 17th century and is rare. It is a common cause of neonatal pleural effusion. Trauma and postcardiothoracic surgery are accepted as causes in etiology, but it is generally associated with lymphatic malformation such as lymphangiectasia in newborns, congenital cytomegalovirus, adenovirus, streptococcal infections, aneuploidy syndromes such as congenital goiter, Turner and Down syndrome, Noonan syndrome and malignancies (5,6).

The characteristics of pleural chylous fluid are as follows: triglyceride >100 mg/dl, protein >20 g/L, lymphocytosis >100 cells/ml, cholesterol 65-220 mg/dl. High triglyceride level and low cholesterol level distinguish true chylothorax from pseudochylothorax. The pleural fluid in culture is sterile (7). Laboratory findings of our patient were similar. Congenital chylothorax is associated with high morbidity and mortality (50%), especially in preterm and low birth weight infants. A small number of cases show spontaneous resolution (8).

There is no consensus on the treatment approach to chylothorax, but conservative treatment with infection control, nutrition and fluid electrolyte management and respiratory support is the common treatment. Surgical pleurodesis, thoracic duct ligation should be performed in resistant cases with excessively prolonged (3 to 4 weeks) drainage or collection >10 ml/kg/day and recurrent chylothorax. These procedures have their own complications and disadvantages as in different studies (9,10). A more conservative approach, such as octreotide infusion therapy, has had some success in the treatment of chylothorax (11). The mechanism of action of somatostatin in chylothorax is not clearly known. However, it has an antisecretory effect by reducing mild splanchnic vasoconstriction and all gastrointestinal-pancreatic secretions, intestinal absorption and hepatic venous flow. Octreotide is usually started with a slow infusion of 0.5-1 mcg/kg/hr and gradually increased until the desired effect. The normal dose range is between 0.2-10 mcg/kg/hour. In a study conducted on neonatal chylothorax, they found that the average for maximum antisecretory effect was 3 days from the start of treatment (12). As side effects of octreotide, hyperglycemia / hypoglycemia, gallbladder sludge, cardiac conduction disorder, hormone antagonism (cortisol, insulin, glucagon, thyroxine, etc.) can be observed (13). It is relatively free of side effects in newborns, but caution should be exercised in infants with NEC. Octreotide treatment was not planned in our patient because chylous fluid did not come out after thoracentesis.

In conclusion, congenital chylothorax is a rare but serious condition in newborns. Early diagnosis and intervention in the prenatal period supports the improvement of postnatal outcomes. Postpartum management includes drainage of pleural fluid, dietary changes, medication, and rarely surgery. In this case, there is no universally accepted guideline for the use of drugs and the duration of conservative treatment. Generally, the outcome of the condition depends on the underlying genetic condition and associated malformations.

References

1. Kotbi İ, Hojaili N, Alzahrani A et al. Congenital Chylothorax in Newborn with Trisomy 21. J B & Bio Engine; 2021.
2. Krishnamurthy MB, Malhotra A. Congenital chylothorax: current perspectives and trends. Research and Reports in



- Neonatology. 2017;7:53-63.
3. Paramés F, Freitas I, Fragata J, Trigo C, Pinto MF. Octreotide additional conservative therapy for postoperative chylothorax in congenital heart disease. *Rev Port Cardiol* 2009;28:799-807.
 4. Büyüktiryaki M, Okur N, Uraş N, Oğuz ŞŞ. Yenidoğanda Şilotoraks Tanı ve Tedavisinde Güncel Yaklaşımlar. *Türkiye Çocuk Hast Derg/Turkish J Pediatr Dis / 2017; 1: 76-81*
 5. Avcı A, Ulku R, Onat S. Chylothorax with Down syndrome: unusual case report. *J Intern Dent Med Res.* 2009;2:25-7.
 6. Levine C. Primary disorders of the lymphatic vessels-a unified concept. *J Pediatr Surg.* 1989;24:233-40.
 7. McGrath EE, Blades Z, Anderson PB. Chylothorax: aetiology, diagnosis and therapeutic options. *Resp Med.* 2010;104:1-8.
 8. Sahin Y, Aydin D. Congenital chylothorax treated with octreotide. *Indian J Pediatr.* 2005;72(10):885-8.
 9. Soto-Martinez M, Massie J. Chylothorax: diagnosis and management in children. *Paediatr Respir Rev.* 2009;10:199-207.
 10. Teitelbaum DH, Teich S, Hirschl RB. Successful management of a chylothorax in infancy using a pleurectomy. *Pediatr Surg Int.* 1996;11:166-8.
 11. Al-Tawil K, Ahmed G, Al-Hathal M, Al-Jarallah Y, Campbell N. Congenital chylothorax. *Am J Perinatol.* 2000;17:121-6.
 12. Radly D, Sheila V, Rama B. Octreotide therapy for chylothorax in infants and children: a brief review. *Pediatr Crit Care Med.* 2008;7:576-9.
 13. Maayan-Metzger A, Sack J, Mazkereth R. Somatostatin treatment of congenital chylothorax may induce transient hypothyroidism in newborns. *Acta Paediatr.* 2005;94:785-9.