American Journal of Medical Case Reports, 2019, Vol. 7, No. 10, 223-227
Available online at http://pubs.sciepub.com/ajmcr/7/10/1
Published by Science and Education Publishing
DOI:10.12691/ajmcr-7-10-1

Concurrent Occurrence of Chylothorax and Chyloperitoneum in Non-Hodgkin's Lymphoma

Tariq Sharman*, Jeffrey Song

Department of Internal Medicine, Southern Ohio Medical Center, Portsmouth, U.S.A.
*Corresponding author: tysharman@yahoo.com

Received April 03, 2019; Revised July 08, 2019; Accepted July 24, 2019

Abstract Chyle is a milky bodily fluid consisting of lymph and emulsified fats, or free fatty acids. It is formed in the small intestine during digestion of fatty foods, and taken up by lymph vessels specifically known as lacteals. Chylothorax refers to the presence of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries. Chylothorax is classified as non-traumatic or traumatic. Non-traumatic chylothorax is mainly caused by malignant etiologies accounting for more than 50% of chylothorax diagnoses. Lymphoma is the most common etiology. Chyloperitoneum is the extravasation of milky chyle rich in triglycerides into the peritoneal cavity, which can occur as a result of trauma or obstruction of the lymphatic system. Chylous ascites is an uncommon finding that can be caused by malignancy, cirrhosis and lymphatic disruption after abdominal surgery. The combination of chyloperitoneum and chylothorax is rare. When abdominal lymphatics are obstructed, chylous ascites results and eventually leads to a chylothorax. The Patient is an 82 year old Caucasian male who presented with worsening shortness of breath and abdominal distension. Chest X-ray showed moderate left sided pleural effusion. Bedside ultrasound-guided paracentesis and thoracentesis revealed milky ascetic and pleural fluid respectively. Triglyceride level was 271 mg/dl in the ascetic fluid, and 221 mg/dl in the pleural fluid. Pleural fluid was exudative with lymphocytic predominant cell count and negative cytology. Lymph node biopsy confirmed Non-Hodgkin's Lymphoma causing the concurrent chylothorax and chyloperitoneum.

Keywords: *chyloperitoneum, chylothorax, chyle, pseudochylothorax, chylous ascites, lymphoma, thoracentesis, pleural effusion, triglycerides, lymphoperitoneal fistula, giant lymph node hyperplasia, sarcoidosis, tuberculosis, histoplasmosis, yellow nail syndrome, systemic lupus erythematosus, noonan syndrome, nephrotic syndrome*

Cite This Article: Tariq Sharman, and Jeffrey Song, "Concurrent Occurrence of Chylothorax and Chyloperitoneum in Non-Hodgkin's Lymphoma." *American Journal of Medical Case Reports*, vol. 7, no. 10 (2019): 223-227. doi: 10.12691/ajmcr-7-10-1.

1. Introduction

Chyle is a milky bodily fluid consisting of lymph and emulsified fats, or free fatty acids. It is formed in the small intestine during digestion of fatty foods, and taken up by lymph vessels specifically known as lacteals. The lipids in the chyle are colloidally suspended in chylomicrons. Chylothorax refers to the presence of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries. A tear or leak in the thoracic duct causes chylous fluid to collect in the pleural cavity. Chylothorax is classified as non-traumatic or traumatic. Non-traumatic chylothorax is mainly caused by malignant etiologies accounting for more than 50% of chylothorax diagnoses. Lymphoma is the most common etiology, representing about 60% of all cases, with non-Hodgkin lymphoma more likely than Hodgkin lymphoma to cause chylothorax. Other malignancies include lung cancer, mediastinal cancers, chronic lymphocytic leukemia, kaposi sarcoma and multiple myeloma. Non-malignant causes of non-traumatic chylothorax include yellow nail syndrome, histoplasmosis, tuberculosis, sarcoidosis, Castleman's disease, liver cirrhosis and Waldenström macroglobulinemia.

Traumatic chyothorax can be surgical or non-surgical. Surgical procedures in the area of the thoracic duct or nearby structures, which account for the majority of cases of traumatic chylothorax, can disrupt the thoracic duct or tear lymphatic tributaries. Esophagectomy, pulmonary resection with lymph node dissection, and surgery for congenital heart disease are among the common causes. Non-surgical traumatic chylothorax can be a complication of central line placement, pacemaker implantation, embolization of pulmonary arteriovenous malformations, and chest trauma.

Chylous ascites is the extravasation of milky chyle rich in triglycerides into the peritoneal cavity, which can occur de novo as a result of trauma or obstruction of the lymphatic system. Chylous ascites is an uncommon finding that can be caused by malignancy, cirrhosis and lymphatic disruption after abdominal surgery. We are presenting a case of concurrent occurrence of chylothorax and chylous ascites in an 82 year old Caucasian male caused by non-Hodgkin lymphoma.

2. Case Presentation

The patient is an 82 year old Caucasian male with history of coronary artery disease post coronary artery bypass grafting surgery, hypertension and hyperlipidemia who presented with worsening shortness of breath mainly on exertion associated with abdominal distention that has been getting worse over a month. He also reported multiple episodes of fever, chills and night sweats, in addition to unintentional weight loss of 40 pounds over the past few months. Patient denied any symptoms of diarrhea, constipation, heat intolerance, chest pain, palpitations or hemoptysis. He also denied any exposure to tuberculosis at present or in the past. His social history is significant for smoking. Physical examination revealed normal vital signs with no acute distress. Lung auscultation revealed decreased air entry on the left lung base with no crepitations or wheezes. Cardiovascular examination showed median sternotomy scar, normal S1 S2 and no murmurs. His Abdomen was distended with positive shifting dullness. Lower limbs with trace bilateral edema. He also had a palpable left axillary lymph node.

Laboratory investigation including complete blood count (CBC), complete metabolic profile (CMP), Hepatic Panel and Thyroid panel were within normal limits. Chest X-ray (Figure 1) showed moderate left sided pleural effusion. Bedside ultrasound-guided paracentesis and thoracentesis revealed milky ascetic (Figure 3) and pleural fluid respectively. Triglyceride level was 271 mg/dl in the ascetic fluid, and 221 mg/dl in the pleural fluid. Pleural fluid was exudative with lymphocytic predominant cell count and negative cytology. Computed tomography (CT) scan of chest (Figure 2), abdomen and pelvis showed multiple bulky lymphadenopathy involving the pelvic, retroperitoneal and mediastinal region. Left axillary excisional lymph node biopsy showed the histopathologic findings of lymphoblastic non-Hodgkin lymphoma.

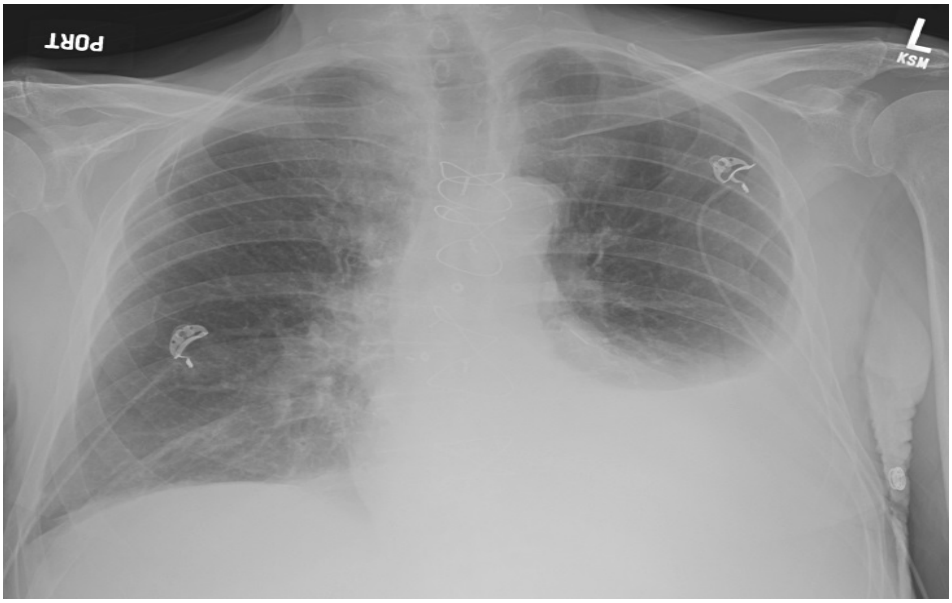

Figure 1. Chest X-ray showing left side pleural effusion secondary to chylothorax

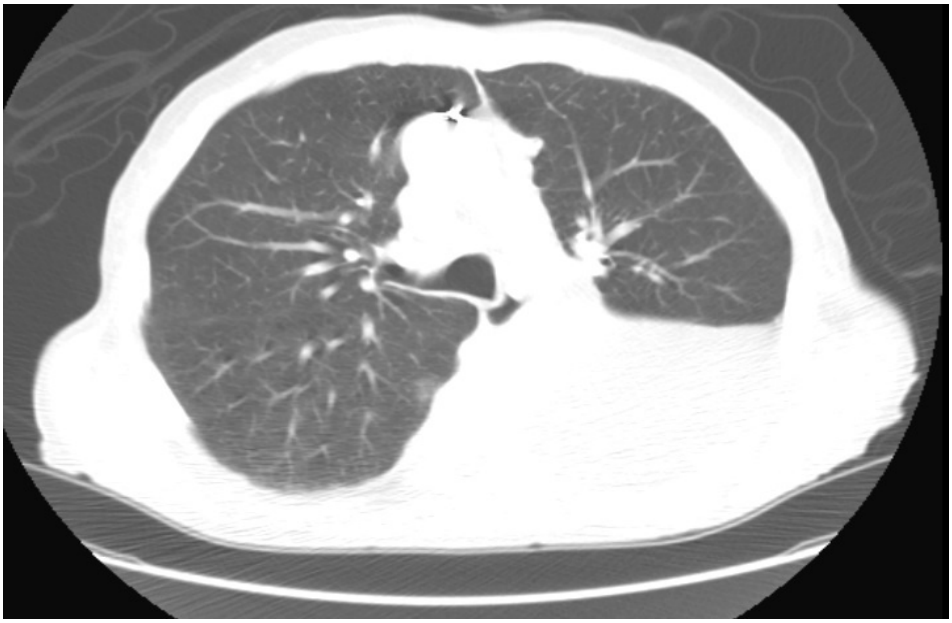

Figure 2. Computed Tomography of the Chest showing left side pleural effusion secondary to chylothorax

Figure 3. Chylous ascetic fluid

3. Discussion

3.1. Chylothorax

Chylothorax refers to the presence of lymphatic fluid in the pleural space secondary to leakage from the thoracic duct or one of its main tributaries. Any disruption or dysfunction of the flow of chyle through the thoracic duct can cause chylothorax. Etiologies of chylothorax can be broadly categorized as non-traumatic or traumatic. Malignancy is the leading cause of non-traumatic chylothorax while thoracic surgery is the major cause of traumatic chylothorax. Abnormalities of the thoracic duct or lymphatics can occur via multiple mechanisms including direct involvement (eg, malignant or infectious lymphadenitis, tear or rupture from trauma or surgery), compressive obstruction (eg, malignancy), transfer of chyle across the diaphragm from abdominal or retroperitoneal chyle accumulations, dysfunction (eg, reverse flow of chyle toward the lung) and anomalies (eg, Gorham syndrome, lymphangiomatosis). Non-traumatic chylothorax can be caused by malignant and nonmalignant conditions. Common malignancies that can be complicated by chylothorax include mediastinal cancers, lung cancer, lymphomas, multiple myeloma and kaposi sarcoma. Non-malignant causes of non-traumatic chylothorax include benign tumors of the mediastinum, giant lymph node hyperplasia, sarcoidosis, tuberculosis, histoplasmosis, yellow nail syndrome, systemic lupus erythematosus, Noonan syndrome, nephrotic syndrome, filariasis and thoracic irradiation. Traumatic causes are classified into surgical and non-surgical. Surgical procedures in the area of the thoracic duct or nearby structures, which account for the majority of cases of traumatic chylothorax, can disrupt the thoracic duct or tear lymphatic tributaries like surgery for congenital heart disease, esophagectomy and pulmonary resection with lymph node dissection. Injury to the intra-abdominal portion of the thoracic duct can cause chylous ascites that passes through the diaphragm, resulting in a chylothorax. Nonsurgical traumatic chylothorax can complicate central line placement, pacemaker implantation, embolization of pulmonary arteriovenous malformations, and chest trauma. Approximately 6 to 14 percent of chylothoraces are idiopathic.

The clinical presentation of chylothorax depend on the rate of chyle accumulation as well as the concomitant effect of the aetiology. Usually patients with chylothorax remain asymptomatic until a large amount of chyle accumulates in the pleural space. The most common presentation is dyspnea, other symptoms include heavy feeling in the chest, fatigue, weight loss and rarely chyloptysis (expectorating chylous fluid).

Rarely, patients may experience a rapid accumulation of fluid in the pleural space, causing a tension chylothorax. This is of particular concern following a pneumonectomy. These patients experience a rapid hemodynamic and respiratory compromise, similar to the classic tension pneumothorax. Patients may be malnourished or have fat-soluble vitamin deficiencies since chyle contains protein and fat, also patients with chylothorax are more prone to infection due to immunosuppression from loss of immunoglobulins. Patients will also have the symptoms and signs of the underlying disorder causing the chylothorax. The onset of symptoms is gradual in patients with non-traumatic chylothoraces (eg, malignancy) while the onset of a posttraumatic or postsurgical chylothorax may be immediate if the volume is high (>500mL/day) or occur within a few days after the traumatic event (2 to 10 days) for those in whom accumulation is slower. In postsurgical patients who have a slow leak, accumulation may begin soon after resuming oral intake. In surgical patients, the chylothorax may be first detected as a pleural effusion on serial radiographic evaluations or by the persistent drainage of pleural fluid from a preexisting chest tube.

On physical examination, findings of decreased breath sounds and stony dullness to percussion may be present depending on the size and location of the effusion.

Laboratory findings in chylothorax are non-specific and usually laboratory abnormalities are related to the disease causing the chylothorax. Serum glucose, LDH, total protein and triglycerides are usually sent for comparisons with pleural fluid values. Rarely, when severe, electrolyte loss into the pleural space may result in hyponatremia, hypocalcemia, and metabolic acidosis.

Thoracentesis and pleural fluid analysis are the criterion standards to establish a diagnosis of chylothorax. Alternatively, in a postsurgical patient, tube thoracostomy output can be analyzed. The appearance of fluid from a chylothorax can be milky, sanguineous, or serous. The detection of milky-appearing fluid is not specific for chylothorax, since milky fluid can also be seen in a cholesterol pleural effusion or an empyema. The white blood cell differential of fluid from chylothoraces typically has a predominance of lymphocytes, usually greater than 70 percent of the total nucleated cell count, reflecting the cellular composition of lymph. Most chylothoraces (up to 85 percent) have a higher protein concentration making them exudates by Light's criteria. However, transudative chylothorax has been reported in a small proportion of patients with amyloidosis, cirrhosis, nephrotic syndrome, superior vena cava obstruction, heart failure, and chylous ascites that has crossed the diaphragm into the pleural space. Chylous fluid usually has a pH that ranges from 7.40 to 7.80. The pleural fluid glucose in chylothorax is usually similar to that in plasma. A pleural fluid glucose below 60 mg/dL suggests coexisting empyema or a malignant pleural effusion. LDH levels in chylous pleural fluid are low, being in the range of a transudative pleural effusion by Light's criteria, and elevation of LDH in chylous pleural fluid was associated with an underlying cause of a chylothorax (eg, malignancy) rather than from simple chyle leakage from ruptured lymphatic channels. Measurement of triglyceride and cholesterol levels in the pleural fluid should be the initial lipid tests performed in patients with suspected chylothorax. A triglycerides level greater than 110 mg/dL reflects a 99% chance that the fluid is chyle. A triglycerides level less than 50 mg/dL reflects only a 5% chance that the fluid is chyle. If the triglycerides level is 50-110 mg/dL, use lipoprotein analysis or inspect the pleural fluid for chylomicrons or cholesterol crystals. The cholesterol level is generally less than 200 mg/dL. A ratio of pleural fluid cholesterol to triglyceride of less than 1 is also diagnostic. Chylothorax can be distinguished from pseudochylothorax by fluid analysis. In pseudochylothorax, the cholesterol level is greater than 200 mg/dL, no chylomicrons are present, and cholesterol crystals are seen at microscopy.

Chest radiographic findings are nonspecific for chylothorax and indistinguishable from other causes of pleural effusion; however, they may help to rule out other causes of the patient's symptoms, determine if effusion is bilateral and look for a mediastinal shift. Findings of a unilateral pleural effusion occur in approximately 78 percent of patients and involve the right hemithorax in 67 percent and left hemithorax in 33 percent. However, bilateral effusion can also occur. Occasionally, signs of the underlying disorder may be evident (eg, lymphatic masses, lung malignancy). If the etiology of the chylothorax is unknown, obtaining computed tomography (CT) scanning or magnetic resonance imaging (MRI) of the chest and abdomen for evaluation of the lymphatic vessels, and to rule out vascular abnormalities and/or malignancy will be helpful. Lymphangiography is a contrast-enhanced study of the lymphatic system that can delineate thoracic duct anatomy and identify a potential site of chyle leak. Lymphoscintigraphy is also useful for the localization of the leak, evaluation of the thoracic duct patency, and differentiation of partial from complete thoracic duct transection.

3.2. Chyloperitoneum

Chylous ascites is the extravasation of milky chyle rich in triglycerides into the peritoneal cavity, which can occur as a result of trauma or obstruction of the lymphatic system. Malignancy, cirrhosis, and lymphatic disruption after abdominal surgery are leading causes in adults. Three underlying mechanisms have been proposed:

1. Exudation of lymph through the walls of dilated retroperitoneal vessels lacking valves, which leak fluid through a fistula into the peritoneal cavity (ie, congenital lymphangiectasia)
2. Acquired thoracic duct obstruction from trauma or surgery, causing direct leakage of chyle through a lymphoperitoneal fistula.
3. Obstruction of the lymph flow due to malignancy, causing leakage from dilated subserosal lymphatics into the peritoneal cavity.

Chylous ascites frequently presents as progressive and painless abdominal distention, occurring over the course of weeks to months, depending upon the underlying cause. The most common presenting symptom is abdominal distension. Other clinical features include abdominal pain, anorexia, weight loss/gain, edema, weakness, nausea, dyspnea, lymphadenopathy, early satiety, fever, and night sweats. Fever, night sweats, and lymphadenopathy are usually observed in patients with lymphoma.

Routine laboratory tests may show hypoalbuminemia, lymphocytopenia, anemia, hyperuricemia, elevated levels of alkaline phosphatase and liver enzymes. Serum cholesterol and triglyceride levels are usually normal. Abdominal paracentesis is the most important diagnostic tool in evaluating and managing patients with ascites. Chyle typically has a cloudy and turbid appearance. The ascites triglyceride level is elevated in all patients. Typically, chylous ascites is diagnosed when the ascites triglyceride level is greater than 110 mg/dL. In addition to triglyceride levels, ascetic fluid should be sent for cell count, culture, Gram stain, total protein concentration, albumin, glucose, lactate dehydrogenase, amylase, and cytology.

Computed tomography (CT) of the abdomen is useful in identifying pathologic intra-abdominal lymph nodes and masses. Lymph node biopsy and laparotomy carry the highest yield of diagnostic information.

Statistics show that in patients with non-Hodgkin's lymphoma (NHL) and Hodgkin's disease (HD), 20-30% will develop a pleural effusion. However, effusions in the peritoneal and pericardial cavities are uncommon. Of all the various subtypes, T-cell originated lymphomas, particularly lymphoblastic lymphoma, usually involve Chylous effusions. Various causes may lead to serous effusions in lymphoma patients, including impaired lymphatic drainage due to obstruction in the mediastinal lymph nodes or the thoracic duct, venous obstruction, pulmonary infection, radiation therapy or pleural involvement of the tumor. The main cause of pleural effusion in Hodgkin disease has been identified as thoracic duct obstruction. However in NHL, the primary

consideration was shown to be direct pleural infiltration. Chylous effusions are always caused by an obstruction of the lymphatic trunks. In the present case, the pleural and abdominal effusions were identified to be chylous, strongly indicating that the effusions originated in the lymph trunks. Possible reasons for the effusions may be that the metastatic lymphoma cells blocked the lymph tunnels leading to obstruction and further impairment of these tunnels.

4. Conclusion

Leakage of lymph from the lymphatic ducts causes chylothorax or chyloperitoneum. Chylous effusions are unusual complications of malignant neoplasms, usually lymphomas. The combination of chyloperitoneum and chylothorax is very rare. When abdominal lymphatics are obstructed, chylous ascites results and eventually leads to a chylothorax. Management is mainly conservative with dietary modifications along with the treatment of underlying etiology.

References

[1] McGrath EE, Blades Z, Anderson PB. Chylothorax: aetiology, diagnosis and therapeutic options. Respir Med. 2010; 104: 1-8.

[2] Doerr CH, Allen MS, Nichols FC, Ryu JH. Etiology of chylothorax in 203 patients. Mayo Clin Proc. 2005; 80: 867-70.

[3] Nair SK, Petko M, Hayward MP. Aetiology and management of chylothorax in adults. Eur J Cardiothorac Surg. 2007; 32: 362-9.

[4] Smati B, Sadok Boudaya M, Marghli A, et al. Management of postoperative chylothorax. Rev Mal Respir. 2006; 23: 152-6.

[5] Maldonado F, Hawkins FJ, Daniels CE, et al. Pleural fluid characteristics of chylothorax. Mayo Clin Proc. 2009; 84: 129-33.

[6] Das DK. Serous effusions in malignant lymphomas: a review. Diagn Cytopathol. 2006 May; 34(5): 335-47.

[7] Berkman N, Breuer R, Kramer MR, Polliack A. Pulmonary involvement in lymphoma. Leuk Lymphoma. 1996; 20: 229-37.

[8] Xu K, Hu X, Tian X, et al. Clinical analysis of 123 cases of chylous effusion. Zhonghua Yi Xue Za Zhi. 2011; 91: 464.

[9] Shah RD, Luketich JD, Schuchert MJ, et al. Postesophagectomy chylothorax: incidence, risk factors, and outcomes. Ann Thorac Surg. 2012; 93: 897-904.

[10] Hillerdal, G. Chylothorax and pseudochylothorax. *Eur Respir J*. 1997; 10: 1157-1162.

[11] Nair, S.K., Petko, M., and Hayward, M.P. Aetiology and management of chylothorax in adults. *Eur J Cardiothorac Surg*. 2007; 32: 362-369.

[12] Riquet, M. et al. Chylomediastinum after mediastinoscopy. Apropos of a case. *Rev Mal Respir*. 1993; 10: 473-476.

[13] McWilliams, A. and Gabbay, E. Chylothorax occurring 23 years post-irradiation: literature review and management strategies. *Respirology*. 2000; 5: 301-303.

[14] Bolger, C. et al. Chylothorax after oesophagectomy. *Br J Surg*. 1991; 78: 587-588.

[15] Kurekci, E., Kaye, R., and Koehler, M. Chylothorax and chylopericardium: a complication of a central venous catheter. *J Pediatr*. 1998; 132: 1064-1066.

[16] Van Renterghem, D.M. and Pauwels, R.A. Chylothorax and pleural effusion as late complications of thoracic irradiation. *Chest*. 1995; 108: 886-887.

[17] Rahman, N.M., Chapman, S.J., and Davies, R.J. Pleural effusion: a structured approach to care. *Br Med Bull*. 2004; 72: 31-47.

[18] Maldonado, F. et al. Pleural fluid characteristics of chylothorax. *Mayo Clin Proc*. 2009; 84: 129-133.

[19] Merrigan, B.A., Winter, D.C., and O'Sullivan, G.C. Chylothorax. *Br J Surg*. 1997; 84: 15-20.

[20] Diaz-Guzman, E., Culver, D.A., and Stoller, J.K. Transudative chylothorax: report of two cases and review of the literature. *Lung*. 2005; 183: 169-175.

[21] Tie, M.L., Poland, G.A., and Rosenow, E.C. 3rd. Chylothorax in Gorham's syndrome. A common complication of a rare disease. *Chest*. 1994; 105: 208-213.

[22] Worthington, M.G. et al. Isolated thoracic duct injury after penetrating chest trauma. *Ann Thorac Surg*. 1995; 60: 272.

[23] Macfarlane JR, Holman CW. Chylothorax. Am Rev Respir Dis 1972; 105: 287.

[24] Fernandez FG, Denlinger CE, Patterson GA, et al. Massive bilateral chylothoraces complicating mediastinal granulomatous disease. Ann Thorac Surg 2009; 88: 1012.

[25] Tutor JD, Schoumacher RA, Chesney PJ. Chylothorax associated with histoplasmosis in a child. Pediatr Infect Dis J 2000; 19: 262.

[26] Nadolski G. Nontraumatic Chylothorax: Diagnostic Algorithm and Treatment Options. Tech Vasc Interv Radiol 2016; 19: 286.

[27] Blankenship ME, Rowlett J, Timby JW, et al. Giant lymph node hyperplasia (Castleman's disease) presenting with chylous pleural effusion. Chest 1997; 112: 1132.

[28] Ryu JH, Doerr CH, Fisher SD, et al. Chylothorax in lymphangioleiomyomatosis. Chest 2003; 123: 623.

[29] Terzi A, Furlan G, Magnanelli G, et al. Chylothorax after pleuro-pulmonary surgery: a rare but unavoidable complication. Thorac Cardiovasc Surg 1994; 42: 81.

[30] Prakash, UBS. Chylothorax and pseudochylothorax. Eur Respir Mon 2002; 7: 249.

[31] Maldonado F, Hawkins FJ, Daniels CE, et al. Pleural fluid characteristics of chylothorax. Mayo Clin Proc 2009; 84: 129.

[32] Sarsam MA, Rahman AN, Deiraniya AK. Postpneumonectomy chylothorax. Ann Thorac Surg 1994; 57: 689.

[33] Huggins JT. Chylothorax and cholesterol pleural effusion. Semin Respir Crit Care Med 2010; 31: 743.

[34] Thaler MA, Bietenbeck A, Schulz C, Luppa PB. Establishment of triglyceride cut-off values to detect chylous ascites and pleural effusions. Clin Biochem 2017; 50: 13.

[35] Acta Clin Belg. Chylothorax and chylous ascites due to malignant lymphoma. 1995; 50(1): 20-4.