Case report

Spontaneously resolving idiopathic bilateral chylothorax

Mohammed Ismail Nizami, G. K. Paramjyothi, Narendra Kumar N, R. P. Boddula

Department of Respiratory Medicine, Nizam’s Institute of Medical Sciences, Panjagutta, Hyderabad-500082, Telangana, India

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Abstract

Chylothorax is a relatively rare clinical entity caused by injury or obstruction of the thoracic duct or its tributaries leading to accumulation of chyle in the pleural cavity. It is characterized by a high concentration of neutral fat and fatty acids in the pleural fluid. Bilateral chylothorax is even more unusual, the commonest etiology being malignancy and trauma. However, we report a case of idiopathic bilateral chylothorax which resolved with medical management.

Key words: chylothorax, chyle, lymph, intercostal drainage, lymphoscintigraphy, medium chain triglycerides (MCT)

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Chyle in the pleural space was first described by Bartolet in 1633. The pleural fluid in chylothorax consists of high amounts of chylomicrons and very low-density lipoproteins. Inspite of being rare, chylothorax may have serious clinical consequences including cachexia, immunodeficiency and fibrothorax. A prompt diagnosis needs to be made based on the analysis of pleural fluid. Treatment can be divided into conservative management and surgery. There are no evidence based guidelines to assist in the management of this disease. Initial conservative therapy includes therapeutic thoracocentesis along with nutritional support in the form of total parenteral nutrition and reduction of chyle formation with somatostatin or octreotide. Surgical interventions include thoracic duct ligation, pleuroperitoneal shunt and percutaneous embolisation.

Case report

A 40 year old male with no significant previous medical history presented to emergency department with a right sided chest drain in-situ containing milky fluid (Fig 1). He was referred from a peripheral hospital where diagnosis of right sided chylothorax was made. His clinical examination was unremarkable except for diminished air entry in the right hemithorax. His pleural fluid examination done outside was suggestive of an exudate with a triglyceride level of 1719mg/dl and cholesterol level of 146mg/dl. All routine blood investigations were found to be within normal limits. A detailed workup was planned including a contrast enhanced CT scan of the chest and lymphoscintigraphy(Fig 3). During the course of hospital stay, he continued to drain around 500ml of chyle daily which progressively reduced to less than 50ml on the 14th day of admission. His CECT chest reported right sided pleural effusion with normal mediastinal vascular structures and no hilar lymphadenopathy. Lymphoscintigraphy with 2 mci of Tc 99m sulphur colloid could be traced into the intercostal drainage bag but the site of leak could not be identified.

The clinical and radiological improvement was observed in the 2nd week of hospitalisation following which the ICD tube was removed and patient discharged in a stable condition. He was found to be normal at the follow-up visit after 2 months.
Fig 1. Chest radiograph showing right sided pleural effusion with intercostal drainage tube in situ.

Fig 2. Chest radiograph showing a newly developed left sided pleural effusion.

Fig 3. CT image showing right sided pleural effusion.

Fig 4. Lymphoscintigraphic image depicting the collection of tracer material in the intercostal drainage bag, however the site of chyle leak could not be identified.

Subsequently he presented with left sided pleural effusion of chylous etiology, five months later (Fig 2). Analysis of the pleural fluid confirmed the diagnosis of chylothorax but the other investigations were, as expected, unrevealing.

Repeat lymphoscintigraphy also could not identify the site of leak (Fig 4).

However, the patient was managed conservatively this time with repeated thoracocenteses. He was prescribed a fat free medium chain triglyceride (MCT) diet. Following three pleural fluid aspirations, each around one week apart, the patient is now asymptomatic and doing well on regular follow-up visits.

Discussion

A chyle leak may manifest in a different ways—as a chylethorax (chylous effusion) into the thoracic cavity, as a chyloperitoneum (chylous ascites) into the abdomen, as a chylopericardium around the heart, or as an external draining fistula. Less commonly it may present as chyloptysis (chyle in the sputum) and chyluria (chyle in the urine). Chylothorax is the most frequent cause of pleural effusion in fetuses and neonates. However, in adults it accounts for only 3% of the cases of pleural effusion1. The main causes of chylothorax are malignant tumors, 75% of which are lymphomas. Other rare causes are lymphangiomymomatosis, sarcoidosis, tuberculosis, venous thrombosis, congenital lymphatic malformations, trauma, nephrotic syndrome, hypothyroidism, cirrhosis, decompensated heart failure and idiopathic chylothorax13. In some of the cases, hyperextension of the dorsal spine or a Valsalvamanoeuvre may have caused the rupture of a previously altered duct.

Injury to the thoracic duct below the T-5 level results in a right-sided chylothorax, while damage to the thoracic duct above T-5 leads to a left sided chylous effusion. The thoracic duct usually crosses from the right to the left side of the thorax at the
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The symptoms of chylothorax are nonspecific and related to the presence of fluid in the pleural space causing dyspnea, fatigue and thoracic discomfort on the affected side. Fever and pleuritic pain are rare because the chyle is not irritating to the pleural surface. The loss of chyle might result in hyponatremia, hypocalcemia, acidosis, hypovolemia, reduction of the venous return and lymphocyte depletion in acute settings and weight loss with immunological impairment in chronic cases. The diagnosis of the chylothorax is made through laboratory tests, since clinical signs and imaging tests are inconclusive. Computed tomography of the chest is useful to rule out the presence of lymphoma or metastasis but is ineffective in diagnosing chylothorax. Lymphoscintigraphy using 99mTc human serum albumin is a simple and non-invasive modality that can visualize lymph ducts up to the thoracic duct. It is used to detect the site of chyle leakage in combination with advanced imaging modalities. Lymphoscintigraphy with SPECT/CT is a latest technique in which a gamma camera and CT are juxtaposed in order to obtain fusion images for accurate identification of lesions. The white, milky appearance of the drainage is often identifying, although the color of chyle may vary from clear to reddish-brown (in the presence of red blood cells). When the levels of triglycerides are higher than 110 mg/dl, there is a greater than 99% probability of pleural fluid being chylothorax. However, if the triglyceride levels are lower than 50 mg/dl, with normal serum levels of cholesterol and triglycerides, the probability of chylothorax drops to 5%. If the levels of triglycerides are inconclusive, the presence of chylomicrons in the pleural fluid confirms the diagnosis.

It is important to review the factors that affect the production and flow of chyle in the management of chylothorax. Interestingly, there are several factors other than fat intake involved in the pathogenesis. Any activity that increases blood flow will increase the flow of chyle. This includes exercise, especially torso or upper extremity exercises or increase in the intraabdominal pressure (such as during coughing or straining). Although the primary focus of nutrition therapy is on reducing fat in the diet, it has been shown that peristalsis and any enteral intake, even ingestion of water, can increase lymph flow by 20%. However, high fat intake (in particular long chain fat), will augment the flow rate of chyle.

Octreotide, a somatostatin analog, has recently been tried as a pharmacological means to manage a chyle leak. It is a potent inhibitor of growth hormone, glucagon, and insulin. It also suppresses gastrointestinal hormones including gastrin, motilin, secretin and pancreatic polypeptide thereby decreasing the splanchnic blood flow. Although the exact mechanism of action of Octreotide in chyle leaks is not well understood, it is attributed to deceleration in lymph flow.

The conservative treatment of spontaneous chylothorax consists of preventing dehydration, nutrition maintenance and reduction of the chyle formation rate. Options for nutritional management include a low fat or fat free diet, enteral nutrition with a specialized formula or total parenteral nutrition. Nutritional status should be monitored carefully in patients on a fat free diet. Fat-soluble vitamins and essential fatty acids (EFA) may need to be supplemented. Medium chain triglycerides (MCT) are frequently ordered for the treatment of chyle leaks. The advantage of MCT is that they do not require transport via the lymph system. Hydrolyzation to medium chain fatty acids (MCFA) occurs rapidly, allowing absorption across the brush border where the MCFA then binds to albumin and is transported directly to the liver via the portal vein. Very few cases of spontaneous bilateral chylothorax have been reported in the literature so far.

Conclusion
In small lesions, the duct frequently regenerates spontaneously and no surgical intervention is required. In our case, intercostal drainage, nutritional supplementation and regular outpatient monitoring were the cornerstone in management.

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References
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