Case reports

Chylothorax; a description of four cases and a brief review

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Abstract

Chylothorax probably accounts for less than 1% of pleural effusions and early recognition of the condition is imperative to prevent sequelae and there is usually a need for extensive subsequent investigations to arrive at the final diagnosis. We report four cases of this uncommon condition with different aetiologies, namely chronic pleural tuberculosis, mediastinal sarcoidosis or thoracic surgery, mediastinal lymphoma and gastric adenocarcinoma, that presented during a period of two years to the specialized respiratory medicine unit, Teaching Hospital Kandy, Sri Lanka. We also briefly review the pathophysiology, aetiology, diagnosis and management of chylothorax.

Key words: pleural effusion, chyle, chylous, fibrothorax.

Introduction

Chylothorax is defined as the presence of "chyle" or "lymph of intestinal origin" in the pleural cavity. The pleural fluid in chylothorax consists of chylomicrons and very low-density lipoproteins, resulting in its characteristic "milky" appearance due to the high triglyceride level (Figure 1). Although this accounts for only a small proportion of clinical pleural effusions, prompt recognition is warranted, to plan out an investigating strategy to elucidate one out of diverse possible aetiologies and to avoid the possible hazardous complications, including malnutrition, immunodeficiency and fibrothorax[1,2].

Chylothorax may be idiopathic, traumatic or non-traumatic in origin. In many series, malignancy was the most common cause of chylothorax in approximately one half of patients. Lymphoma and bronchogenic carcinoma were the commonest of malignancies. The second most common cause of chylothorax was trauma, including neck and thoracic surgery[3,4]. "Pseudochylothorax" or cholesterol effusion may macroscopically mimic a chylothorax, but is rare and usually occurs secondary to chronic pleural effusions lasting for several months to years with thickenend or calcified pleural surfaces[5].

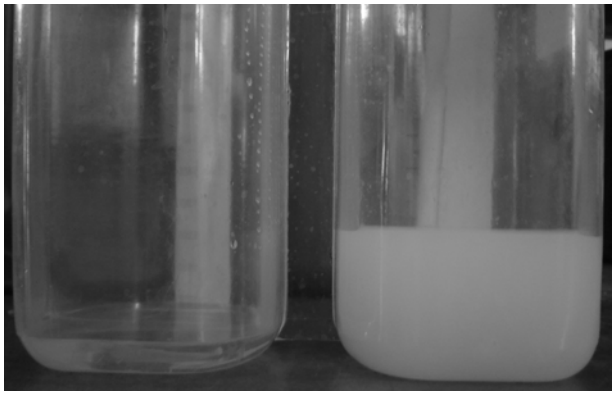

Figure 1. Macroscopic "milky" appearance of chylothorax.

Case 1

A young male with recurrent unilateral pleural effusion for two years, presented during the third episode and was investigated and diagnosed to have chylothorax (Figure 2). He had a high ESR and positive Mantoux test, without a past history of tuberculosis. His sputum was negative for acid-fast bacilli (AFB). Pleural fluid testing was negative for AFB but the adenosine deaminase level was high (95 iu/l). Pleural fluid culture did not grow mycobacteria.

Pleuroscopy revealed the parietal pleura to be coated with thick exudative material and histology of pleural biopsy revealed granuloma with caseation. He was diagnosed with chronic pleural tuberculosis and commenced on anti-tuberculosis therapy (ATT). CT scan of thorax did not reveal lymphoma, malignancy or structural anomalies. He showed symptomatic improvement with normalization of inflammatory markers. Marked resolution of effusion was noted at the end of ATT, with some residual pleural thickening on the CT scan of chest performed at the end of treatment. He was followed up for about two years without recurrence of the chylothorax.

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Case 2

A middle aged man was investigated for vague throat discomfort and progressive dyspnoea. He had prominent paratracheal shadows on chest x-ray and CT chest revealed bilateral paratracheal and tracheobronchial lymphadenopathy (Figure 3). Sputum AFB and Mantoux were negative and blood picture was normal. ESR was only moderately elevated (56 mm/hr), while serum angiotensin converting enzyme (ACE) level was high.

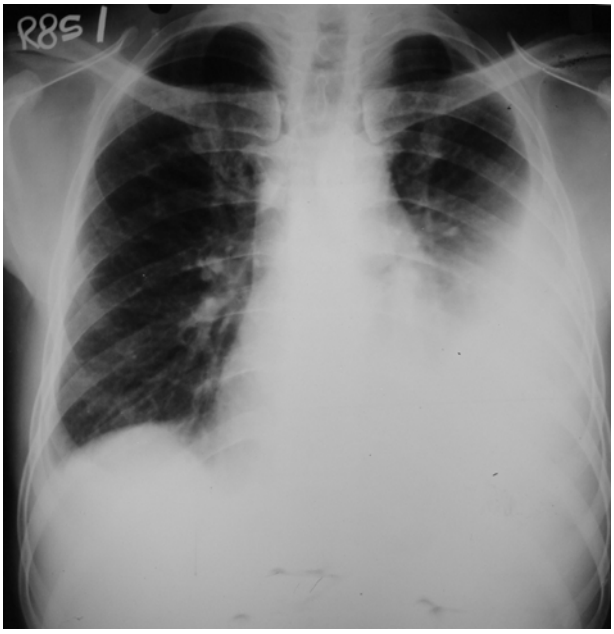

Figure 2. Chronic left sided pleural effusion without tracheal deviation.

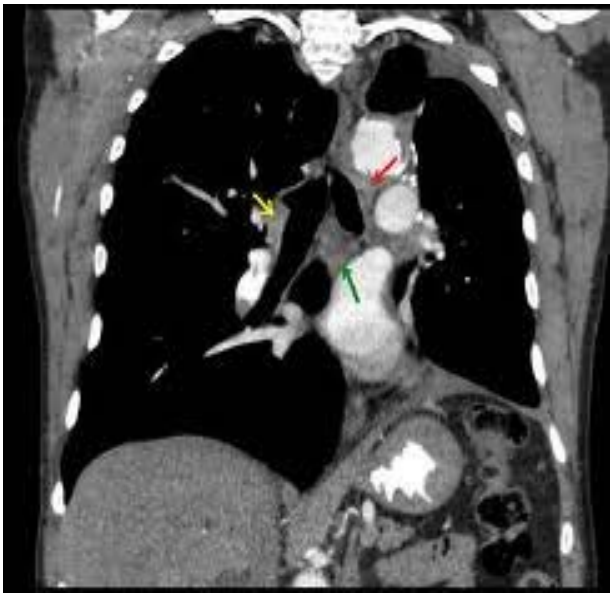

Figure 3. CT scan of chest showing bilateral paratracheal and tracheobronchial lymphadenopathy in mediastinal sarcoidosis.

Mediastinoscopy followed by sampling of mediastinal lymph nodes was performed and the histology proved the diagnosis of sarcoidosis. Subsequently he developed unilateral chylothorax on the side of surgery, requiring intercostal (IC) drainage. Condition resolved within two weeks and lymph nodes regressed with steroids. He did not have recurrent effusion.

Case 3

A young adult female presented with left sided massive pleural effusion with a mediastinal shift. IC drainage was chylous and her CT of thorax showed a large anterior mediastinal mass with pretrachal, paratracheal and hilar lymphadenopathy (Figure 4). Blood investigations showed mild normocytic anaemia, marginal thrombocytopaenia and hypoalbuminaemia.

CT guided biopsy from the lesion confirmed Hodgkin's lymphoma. She was immediately commenced on combined onco-chemotherapy, with nutritional support and reversed barrier nursing. IC drainage gradually declined over next six weeks and marked resolution noted at the third cycle of chemotherapy. She did not require decortication of the affected pleura.

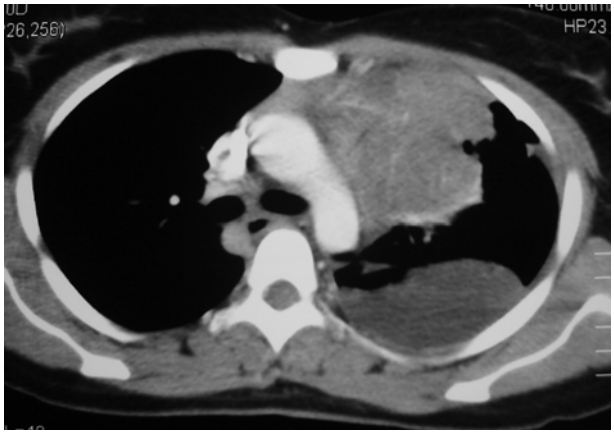

Figure 4. CT scan of chest showing mediastinal mass in lymphoma with pleural effusion.

Case 4

An elderly female presented with marked appetite loss and worsening dyspnoea. She had bilateral asymmetrical moderate chylothoraces, left supra-clavicular discrete lymphadenopathy and non-tender epigastric mass. She was markedly cachectic with anaemia and a high ESR.

Abdominal ultrasonography showed a mass of probably gastric origin, but no lymphadenopathy.

Peripheral lymph node sampling revealed metastatic adenocarcinoma, while gastroduodenoscopy and biopsy confirmed primary gastric carcinoma. She succumbed to her illness during treatment.

Discussion

These case histories illustrate some different aetiological factors, namely chronic tuberculosis, mediastinal lymphoma and gastric carcinoma causing chylothorax. In the case 3, it was probably secondary to thoracospy induced trauma, even though sarcoidosis itself is a recognized cause.

Pathophysiology

The thoracic duct begins at the cisterna chyli near the T-12 vertebra and ascends through the aortic hiatus of the diaphragm between the aorta and the azygous vein into the posterior mediastinum. At the level of the fifth thoracic vertebra, it crosses to the left of the vertebral column on its way to end near the junction of internal jugular and subclavian veins. Therefore obstruction of the thoracic duct by lymphoma or bronchogenic carcinoma tends to cause a right-sided chylothorax when the lower portion of the duct is involved, whereas a left-sided chylothorax results when the upper portion of the thoracic duct is involved[6].

Chylothorax results from either extrinsic compression or infiltration of the thoracic duct, that causes an increase in intraductal pressure, or following traumatic damage to the duct. This increased pressure promotes the formation of dilated collateral channels that eventually drain into the pleural space. 1,500 to 2,500 milliliters of chyle is normally emptied into the venous system daily via the thoracic duct[7].

Aetiology

The causes of chylothorax can be categorized as nontraumatic or traumatic (Table 1). Idiopathic chylothorax has included most cases of neonatal (congenital) onset as well as cases with no clear explanation for the occurrence of chylothorax. Chylothorax is the most common form of pleural effusion in the first few days of life[3,8].

Diagnosis

Non-traumatic chylothorax is usually of gradual onset with progressive dyspnoea due to mechanical effects. Since chyle is not irritating, fever and pleuritic pain are uncommon. Traumatic chylothorax usually develops within two to ten days post injury[9].

On pleural aspiration, distinctively white, odourless, and milky aspirate is found. However, the absence of a milky appearance does not exclude a chylothorax, especially if the patient is fasting or on a low fat diet. The diagnosis of chylothorax is established by measuring triglyceride levels in the pleural fluid. If triglycerides are greater than 110 mg/dL, the diagnosis is probably chylothorax; if the level is less than 50 mg/dL, chylothorax is unlikely. When levels are between 50 to 110 mg/dL, lipoprotein analysis of pleural fluid should be performed. Presence of chylomicrons in the fluid confirms the diagnosis of chylothorax[10].

Table 1. Causes of chylothorax

Non-traumatic causes	*Traumatic causes*
Malignancy	**Surgery**
1. lymphoma 2. carcinoma of lung 3. mediastinal / thoracic metastases	Cardiovascular, aortic, thoracoplasty, esophagectomy, pneumonectomy, transabdominal vagotomy, esophageal endoscopic sclerotherapy, neck surgery, Bochdalek herniorrhaphy
Non-malignant causes	**Nonsurgical causes**
1. idiopathic 2. miscellaneous causes include benign tumors, lymphangio-leiomyomatosis, thoracic aortic aneurysm, subclavian venous thrombosis, thoracic tuberculosis, sarcoidosis, amyloidosis, filariasis	1. Penetrating or nonpenetrating trauma to the neck, thorax or upper abdomen 2. forceful straining, coughing, yawning, vomiting

Chylothorax generally results in an exudative pleural effusion. However, transudative chylothorax has been reported infrequently with amyloidosis, cirrhosis, nephrotic syndrome, superior vena cava obstruction and heart failure[11]. Chylous fluid has a pH that ranges from 7.4 to 7.8. Milky or creamy pleural fluid can also be seen in pseudochylothorax. Both chylous and pseudochylous fluid remain opaque after centrifugation, but the turbidity of chylous fluid clears out on addition of two millilitres of ethyl ether. The presence of cholesterol crystals in the fluid is diagnostic of pseudochylothorax, and chylomicrons are never present on lipoprotein analysis[5].

Lipophilic dye or radio-labeled triglyceride (^{131}I-triolein) ingestion test may also be used to confirm the diagnosis of chylothorax in doubtful cases. Lymphangiography and CT scan of thorax and abdomen may be needed for further evaluation of aetiology.

Management

Chyle is rich in proteins, fats, electrolytes, bicarbonate, lymphocytes and fat soluble vitamins. The more serious sequelae of chylothorax are malnutrition, weakness, dehydration, and metabolic acidosis. Prolonged chylothorax may be associated with reversible T-cell deficiency, hypoproteinaemia and lymphopaenia, resulting in compromised immunologic status with high risk of systemic bacterial and viral infections.

The management of chylothorax is controversial as prospective studies are lacking to guide therapy. Furthermore treatment should be individualized according to the primary aetiology, volume of drainage and complications. If the primary cause is identified and treatable, the specific therapy may help to resolve the chylothorax as well. Medical management including volume, protein and nutrition supplementation is required in all symptomatic cases and prevention of secondary infection is of importance. In most instances, surgical therapy is pursued only after failure of conservative therapy (Figure 5). Surgical treatment modalities include pleurodhesis, percutaneous embolization, thoracic duct ligation and pleuro-peritoneal shunting[12,13,14].

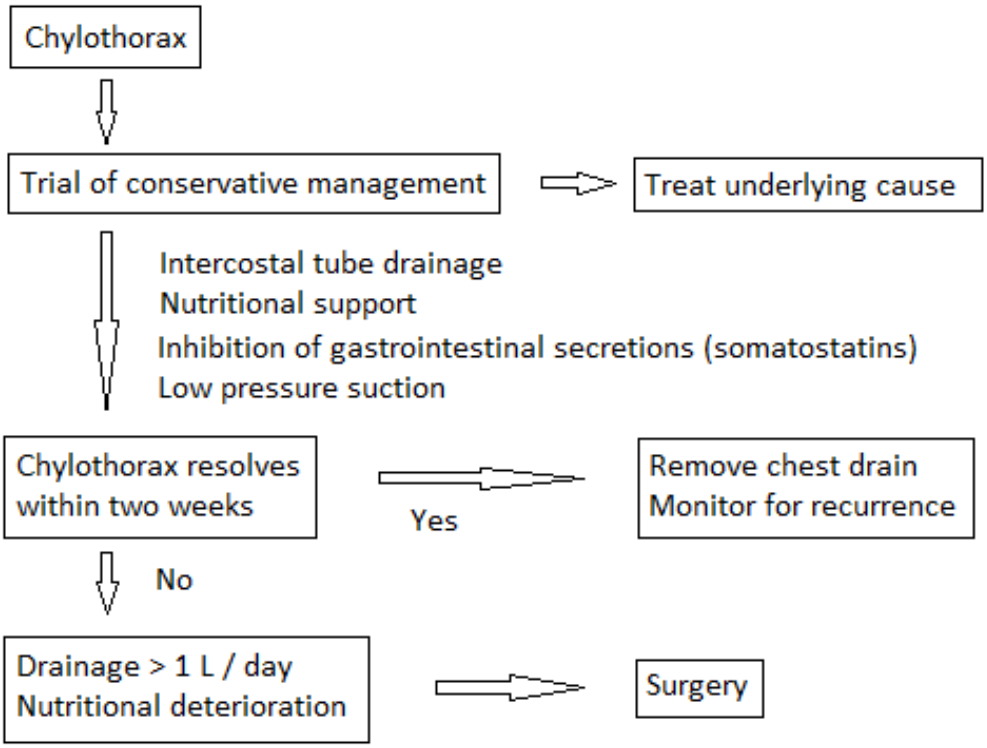

Figure 5. Management of chylothorax.

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