Constrictive pericarditis presenting as chylothorax

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ABSTRACT

Chylothorax is a rare clinical condition that can be attributed to a damaged thoracic duct. The condition is suggested by aspiration of milky white fluid from the pleural cavity and is commonly associated with either malignant diseases or trauma (e.g. cardiothoracic surgery). Other reported causes include thrombosis of the superior vena cava or subclavian veins, pulmonary lymphangiomatosis, filariasis, Kaposi’s sarcoma in acquired immunodeficiency syndrome (AIDS), heart failure, amyloidosis, sarcoidosis, Behcet’s syndrome, tuberculosis (TB) and constrictive pericarditis. Clinical features of chylothorax depend on the rate of development of effusion. Rapid effusions are associated with hypovolaemia and breathing difficulty, whereas effusion of large volumes may be associated with immunosuppression due to the loss of immunoglobulins and lymphocytes in the chyle. We present the case of a 15-year-old boy with treated pleural and pericardial TB leading to constrictive pericarditis and chylothorax.

CASE REPORT

A 15-year-old boy with no known comorbidities presented to our outpatient pulmonology clinic with complaints of dyspnoea on exertion and persistent dry cough for the past three months. He had experienced similar symptoms associated with fever and weight loss three years ago. Radiological imaging revealed moderate left-sided pleural effusion and asctes, which were therapeutically drained. Analysis of the pleural fluid revealed protein content > 4.5 g/dL, and a white blood cell count of 1,200 cells/ml, with 90% lymphocytes. Echocardiogram showed the presence of significant pericardial effusion. The patient was diagnosed with pleural/pericardial TB and started on a four-drug antituberculous treatment regimen for nine months at another centre. His symptoms as well as clinical and radiological findings resolved upon completion of the medical therapy. A year following the cessation of therapy, his pleural effusion returned, and the fluid was drained multiple times at various medical centres in another city. There was no fever, weight loss, sputum, haemoptysis, joint pain or pedal oedema associated with his condition, and there was no history of tobacco or drug use.

The patient then presented to our department for an opinion regarding the cause of his recurrent pleural effusion. On examination, he was a lean and of average height. His blood pressure was 110/70 mmHg, pulse...
was 100 beats per minute and respiratory rate was 20 breaths per minute, while maintaining oxygen saturation of 97%–98% on room air. Chest examination revealed percussion dullness across the left middle and lower chest areas, with absent breath sounds. His jugular venous pressure was markedly elevated at 10 cm above the clavicle.

Chest radiography revealed moderate left-sided pleural effusion (Fig. 1). Thoracocentesis of the pleural fluid revealed a milky white fluid (Fig. 2). Analysis of the pleural fluid revealed 112 mg/dL of glucose, 1,036 mg/dL of proteins, 672 mg/dL of triglycerides and a total white cell count of 600 cells/ml, with 90% lymphocytes and 10% polymorphonuclear cells. Microscopy was negative for bacteria after Gram and Ziehl Neelsen staining. Cytology was also negative for malignant cells. The patient’s serum triglyceride level was markedly raised at 672 mg/dL. The clinical impression indicated chylothorax.

Two-dimensional (2D) echocardiography revealed a mildly dilated right atrium and a moderately dilated left atrium. The left ventricular systolic function was normal, with an estimated ejection fraction of 55%. Septal bounce was also noted. The transmirtal inflow pattern showed pseudonormalisation of left ventricular filling. Pulmonary vein Doppler showed prominent diastolic flow, which was consistent with increased left atrial pressure. Traces of pericardial effusion and thickening of the pericardium (4 mm) were also noted. Further investigations such as computed tomography were not performed, as the patient had financial constraints and could not afford confirmatory diagnostic tests. Furthermore, the 2D-echocardiography findings, when correlated with the clinical picture, confirmed the diagnosis of constrictive pericarditis.

Histopathological examination of the pericardium showed fibrocollagenous and fibroadipose tissues with mild, chronic and nonspecific inflammation. No evidence of malignancy was observed. Postoperative radiograph of the chest showed resolution of the pericardial and pleural effusions. The patient was discharged following the removal of the chest tube, and had been under regular follow-up. Chest radiography at six months showed no signs of effusion (Fig. 3). His clinical examination at the 12-month follow-up was unremarkable, with no recurrence of symptoms. He has since started playing football for his local team without experiencing any dyspnoea.

DISCUSSION

Chylothorax is a rare clinical condition that is suggested by the presence of milky pleural fluid. The fluid is attributable to the obstruction and/or disruption of the thoracic duct or one of its major divisions. It has been proposed that elevated right-sided venous pressure in any context, such as dilated cardiomyopathy, severe tricuspid regurgitation, constrictive pericarditis and right heart failure, may potentially lead to chylothorax and chyloous ascites. However, the leading cause of chylothorax is malignancy, in particular, lymphomas and trauma, especially from thoracic surgery. Other causes include sarcoidosis, amyloidosis, superior vena cava thrombosis and congenital duct abnormalities.

Clinical and radiological tests alone are inadequate for the diagnosis of chylothorax. Aspiration of milky pleural fluid may lead to an empirical diagnosis. However, this must be supported by analysis of the pleural fluid and identification of the disruption to the thoracic duct, where possible. As the fluid is derived from the thoracic duct, its constituents include dietary fat, along with immunoglobulins and leucocytes. Studies have reported that analysis of such aspirate should show a triglyceride level > 1.2 mmol/L (110 mg/dL) and white
blood cells (400–6,800 cells/ml), with lymphocytic predominance.\(^{10}\) However, this is not absolute, as wide variations in the composition and colour of the chyle have been noted.\(^{10,11}\) A definitive diagnosis of chylothorax can be established by demonstrating the presence of chylomicrons in the aspirated fluid.\(^{12}\)

Our patient presented with signs of chylothorax, which was confirmed on aspiration of milky fluid and its analysis, which revealed raised levels of triglycerides. However, no aetiological cause could be identified. Nevertheless, the presence of constrictive pericarditis on echocardiography and the rapid resolution of chylothorax following pericardiectomy strongly suggested that it was secondary to constrictive pericarditis. Constrictive pericarditis rarely causes chylothorax. It has been postulated that chylothorax and chylous ascites in constrictive pericarditis are the result of increasing effective capillary filtration secondary to central venous hypertension and reduced lymphatic drainage due to high pressure in the left subclavian vein.\(^{13}\)

Constrictive pericarditis may occur with the healing of acute fibrinous, serofibrinous pericarditis or chronic pericardial effusion, which in turn results in obliteration of the pericardial cavity and formation of granulation tissue.\(^{14}\) TB is a common cause of constrictive pericarditis; other causes include purulent infections, trauma, cardiac operation, mediastinal irradiation, histoplasmosis, neoplastic disease, acute viral or idiopathic pericarditis, rheumatoid arthritis, systemic lupus erythematosus and chronic renal failure treated by chronic dialysis.\(^{10}\) The rapid resolution of our patient's chylothorax following pericardiectomy strongly suggests that it was secondary to constrictive pericarditis.

Chylothorax can be treated conservatively or surgically; however, the definitive treatment of chylothorax is the identification and management of the underlying pathology. We suggest that when dealing with a case of chylothorax, constrictive pericarditis should be considered in the aetiology.

REFERENCES