Case Report

Rheumatoid Arthritis Complicated by Pseudochylothorax Without Pleural Thickening: A Diagnostic Challenge for Clinicians

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Abstract

A 60-year-old female who was known to have rheumatoid arthritis for the preceding two-and-half years presented with difficulty in breathing associated with chest pain over the right hemithorax of two months duration. She was found to have a right-sided mild to moderate pleural effusion; there was no evidence of pleural thickening. The pleural fluid was pale yellow in appearance and diagnostic work-up confirmed it to be a pseudochylous pleural effusion. The present case highlights the rare occurrence of pseudochylothorax without pleural thickening as a complication of rheumatoid arthritis.
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Key words: Rheumatoid arthritis, Pleural effusion, Pseudochylothorax.

Introduction

Pseudochylothorax, also referred to as cholesterol pleurisy or chyliform effusion, is a condition where the pleural fluid has a very high content of cholesterol. However, triglycerides or chylomicrons are not present and the entity not related with lymphatic vessels or chyle.[1] It can occur when fluid has been present for a long time in the pleural space and, more especially, when there is pleural flibrosis. It is commonly associated with chronic inflammatory disorders, such as tuberculosis or rheumatoid arthritis.[2] Patients with rheumatoid arthritis have a high incidence of exudative pleural effusion and pseudochylothorax is rare.[3] Here we report the rare occurrence of pseudochylothorax without pleural thickening in a patient with rheumatoid arthritis. Until now, there have been only a few published cases documenting the occurrence of pseudochylothorax in patients with rheumatoid arthritis in the English language literature to the best of our knowledge.

Case Report

A 60-year-old female, presented with complaints of difficulty in breathing associated with right lower chest pain of two months duration. The pain was continuous and worsened on deep breathing. There was no associated history of fever or weight loss. There was no history of tuberculosis in the patient or her family members. She was known to have rheumatoid arthritis for the past two-and-half years and had been taking non-steroidal anti-inflammatory drugs as and when required.

On physical examination, findings suggestive of a right-sided pleural effusion were evident. Chest radiograph showed mild to moderate right-sided pleural effusion. The same was confirmed by ultrasonography and a contrast enhanced computed tomography of the thorax (Figure 1).

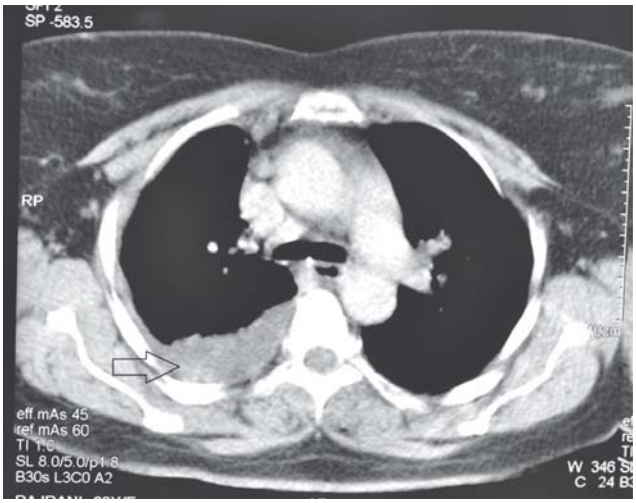

Figure 1. Computed tomography of thorax (mediastinal window) showing absence of pleural thickening with effusion (arrow).

Complete haemogram and serum biochemistry including renal and liver function tests, serum electrolytes and lipid profile were within normal limits. Rheumatoid factor was positive (46.3 IU/L).

Diagnostic thoracocentesis was performed and around 10mL pale-yellowish coloured fluid (Figure 2) was aspirated from the right pleural space and sent for

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Figure 2. Gross appearance of pleural fluid.

analysis. Pleural fluid leucocyte count was 2400 cells/mm^3 with a differential count of 60% neutrophils, 30% lymphocytes and 10% macrophages. The pleural fluid was negative for acid-fast bacilli (AFB) and Gram's staining. Biochemical examination of the pleural fluid revealed glucose 39mg/dL, lactate dehydrogenase (LDH) 20U/L, protein greater than 15g/dL, triglycerides 91mg/dL and cholesterol 1327mg/dL. On cytopathology, pleural fluid showed intact and degenerated neutrophils, macrophages and few mesothelial cells along with an occasional giant cell in a background of amorphous proteinaceous material.

The patient was diagnosed to have pseudochylous pleural effusion. She was started on prednisolone 20mg morning dose after meal with hydroxychloroquine 200mg daily as a treatment of rheumatoid arthritis. After one week of therapy the patient was discharged with significant improvement. Computed tomography of the thorax was repeated after six months of treatment for rheumatoid arthritis and it showed neither pleural effusion, nor pleural thickening (Figure 3).

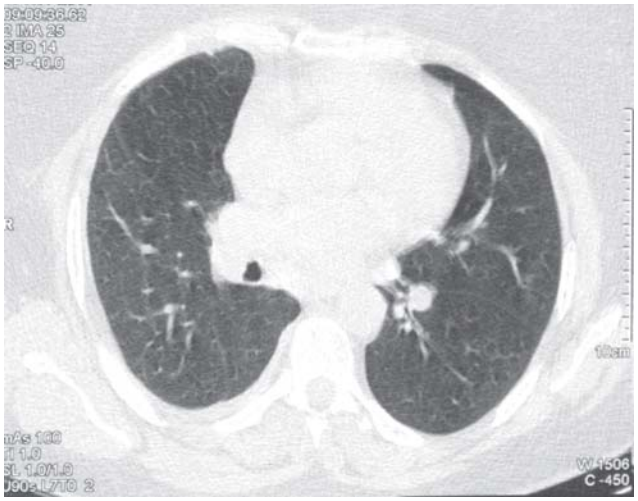

Figure 3. Computed tomography of thorax (lung window) after six months of treatment for rheumatoid arthritis.

Discussion

Pleural effusion has been reported to occur in fewer than 5% of patients with rheumatoid arthritis.[4] In fact, presentation in the form of pseudochylothorax is an uncommon pleural manifestation in patients with rheumatoid arthritis. Pseudochylothorax presents as a unilateral condition and approximately one-third of patients are asymptomatic at presentation. Unlike chylothorax, which occurs due to leakage of chyle into the pleural space, pseudochylothorax is due to the accumulation of cholesterol or lecithin-globulin complexes and is characterised by high cholesterol content and milky appearance of pleural fluid, often with cholesterol crystals seen on microscopy. The majority of pseudochylothoraces are associated with tuberculois, rheumatoid arthritis, pleural paragonimiasis, trauma among others.[5]

Since Weem's description in 1918, the medical literature has always emphasised that a hallmark of pseudochylothoraces is the presence of a grossly thickened (fibrotic) pleura, resulting from chronic intense pleuritis.[6] Yokosuka *et al*[7] reported similar case recently where parietal pleura showed slight thickening. It has been suggested that a pleural inflammatory process of at least five years is required for the development of a pseudochylothorax.[8] However, in our patient, the entire history of chest pain was only of two months duration, and no significant pleural thickening was seen on computed tomography of the thorax (Figure 2). A similar case of psudochylothorax with absence of pleural thickening and disease duration of less than six months has also been reported earlier.[6,9]

It has been postulated that lysis of erythrocytes and neutrophils in the pleural fluid releases cholesterol and lecithin-globulin complexes. These are "trapped" in the pleural cavity as the fibrotic pleural thickening blocks the drainage of fluid and particulates via parietal lymphatics.[8] The eventual concentration of these lipids gives the pleural fluid a milky appearance.

The paradox of the lack of pleural thickening in the present case reveals that an active acute or sub-acute process produces the intra-pleural accumulation of cholesterol could be responsible for the development of pseudochylothorax, rather than the previously suggested theory of cellular breakdown and cholesterol release within an entrapped pleural space. The exact pathogenesis of pseudochylothorax needs further research.[10] Radiologically, the pleural membranes are usually normal in chylothorax, but thickened and calcified in pseudochylothorax. Computed tomography of the thorax is the imaging investigation of choice in patients with pseudochylothorax.

Pleural fluid triglyceride levels greater than 110mg/dL are highly suggestive of chylothorax, which occurs due to damage or blockage of the thoracic duct. Pleural fluid triglyceride levels below 110mg/dL and cholesterol levels above 200mg/dL is considered as significant and diagnostic for pseudochylothorax.[1,11]

The treatment of pseudochylothorax remains challenging. Aggressive treatment of the underlying condition results in control or resolution of the pseudochylothorax.[1] Intervention may be needed if the patient has symptoms or increasing pleural effusion.

Therapeutic thoracentesis can be used to improve respiratory symptoms resulting from mechanical effects of the effusion. In the present case therapeutic thoracentesis was performed and hydroxychloroquin with prednisolone therapy for rheumatoid arthritis was instituted. A repeat computed tomography of the thorax after six months showed significant resolution (Figure 3).

Pseudochylothorax may be mis-diagnosed as empyema due to similar physical appearance of the pleural fluid; hence this rare diagnosis should always be kept in mind to avoid the hazards of mis-diagnosis.

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