Case Report

Pseudochylous Pleural Effusion in a Case of Rheumatoid Arthiritis

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ABSTRACT

Chyliform (pseudochylous) pleural effusions accumulate gradually due to the breakdown of cellular lipids in long standing pleural effusions, such as Rheumatoid Arthritis (RA) and Tuberculosis. These are also known as cholesterol effusions due to high cholesterol levels in pleural fluid and presence of cholesterol crystals on wet mount. We present the case report of a 60 year old lady, who came to the medicine out patient department with breathlessness and generalized body ache. Radiological investigation revealed a left sided pleural effusion with atelectasis. Based on the characteristic biochemical and cytological findings in the pleural fluid, a diagnosis of pseudochylous effusion was made. Test for rheumatoid factor was done subsequently due to the presence of joint pains, and it was positive.

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Introduction

A chyliform (pseudochylous) pleural effusion is an uncommon disease condition developing as a sequela of a long standing exudative effusion.[1] The most common cause of this pleural reaction is Tuberculosis.[2] The other reported underlying diseases include rheumatoid lung disease, syphilis, diabetes, alcoholism, Meig's syndrome, paragonimiasis, malignancy, and trauma or hemothorax.[3] This entity was so named because unlike chylous effusion, it is not due to disruption of the thoracic duct, but due to a high lipid content, which causes it to be turbid.[2] The exact pathogenesis of chyliform pleural effusions is unknown.

Case Report

A 60 year old lady presented with breathlessness, generalized body ache, joint pains and loss of appetite since two months. She was a known case of bronchial asthma. There was no history of diabetes, hypertension, tuberculosis or any drug allergy.

On complete blood count, the patient had hemoglobin of 12.9 gm/dl and a high white blood cell (WBC) count of 19400/cumm, with neutrophilic predominance (76%). The platelet count was 3.7 lakh/cumm. ESR was 56 mm in the first hour. CRP (C Reactive protein) showed an elevated value of 40.2 mg/L. The RA factor was 35 IU/ml (normal value less than 30 IU/ml).

On radiological evaluation, x ray showed left sided pleural effusion and a homogeneous opacity in the left lower zone, with rising level and blunting of left CP angle (Figure 1). Ultrasound chest showed a moderate pleural effusion on left side. HRCT chest was also done, which confirmed the above findings and showed basal atelectasis on left side along with few fibrotic lesions in the right upper lobe.

Thereafter, thoracocentesis was done and 30 ml of pleural fluid was sent for cytological evaluation. The fluid was turbid and slightly hemorrhagic. Pleural fluid WBC count done by Modified Neubauer's chamber was 50 cells/µl. Manual differential count revealed lymphocytic predominance. Smears showed numerous macrophages, some of which were epithelioid (Figure 2), along with few multinucleated giant cells and mesothelial cells (Figure 3). The background showed amorphous extracellular granular debris. On wet mount, cholesterol crystals with characteristic "shattered glass appearance" were also noted and confirmed with the help of a polarizer (Figure 4).

Pleural fluid protein was raised at 9.7 gm/dl; while glucose levels were reduced at 16 mg/dl (normal reference value for pleural fluid protein is less than 3 gm/dl, while that for glucose is 70-210 mg/dl). Adenosine Deaminase (ADA) levels were done to rule out tuberculosis; the value was normal (25 U/L). Pleural fluid cholesterol levels were raised (230 mg/dl), while triglyceride levels were normal (70 mg/dl). Pleural fluid culture was negative.

The patient was managed conservatively with thoracocentesis and anti-rheumatoid drugs. Subsequently, the patient was discharged in a satisfactory condition.

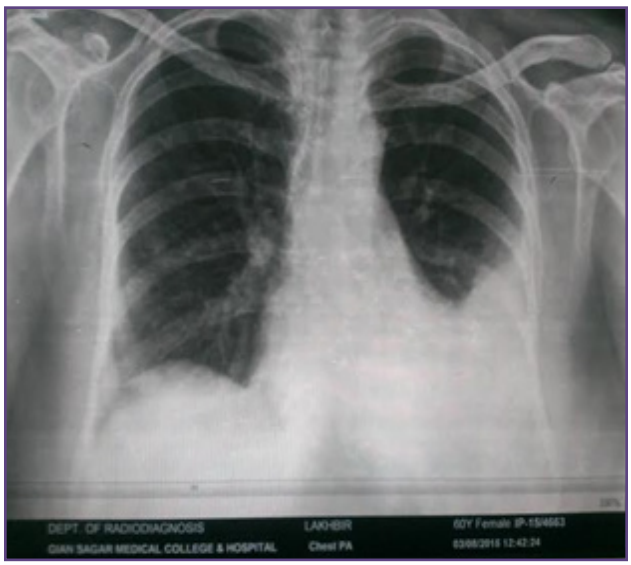

Fig. 1: Chest XRay Postero-Anterior(PA) view showing left sided pleural effusion.

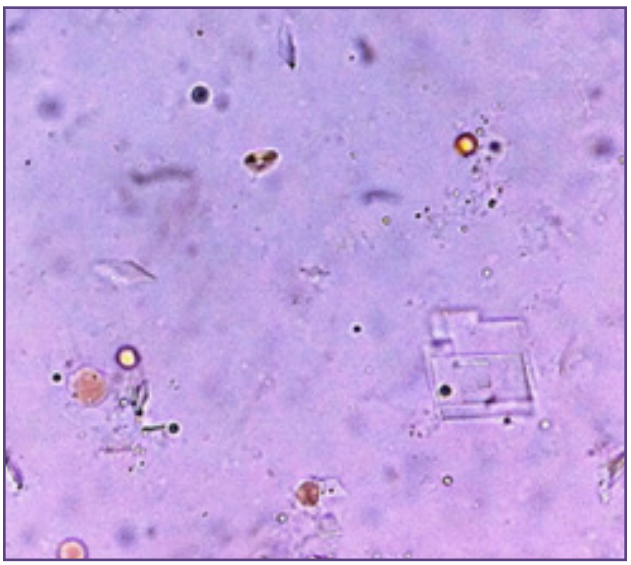

Fig. 2: High power view of wet mount of pleural fluid showing amorphous debris in the background, against which are present cholesterol crystals having a characteristic shattered glass appearance. (X400).

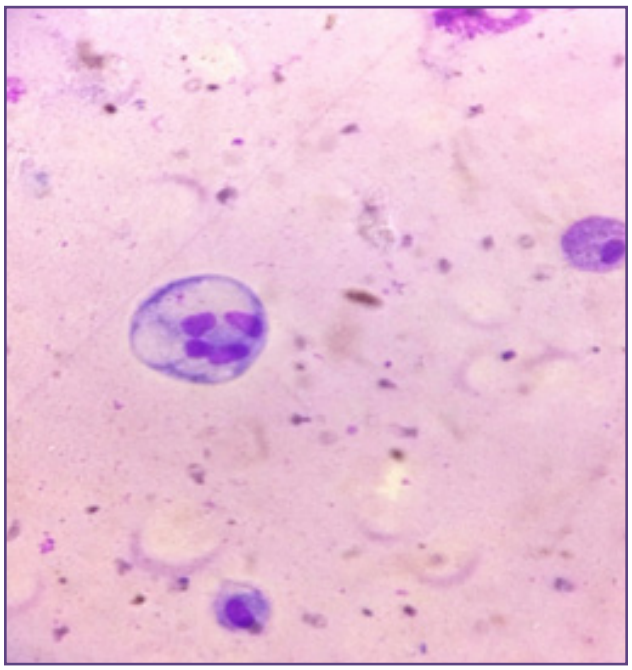

Fig. 3: High power view of cytological smear of pleural fluid showing an epithelioid multinucleated giant cell and few histiocytes. (LeishmannX400).

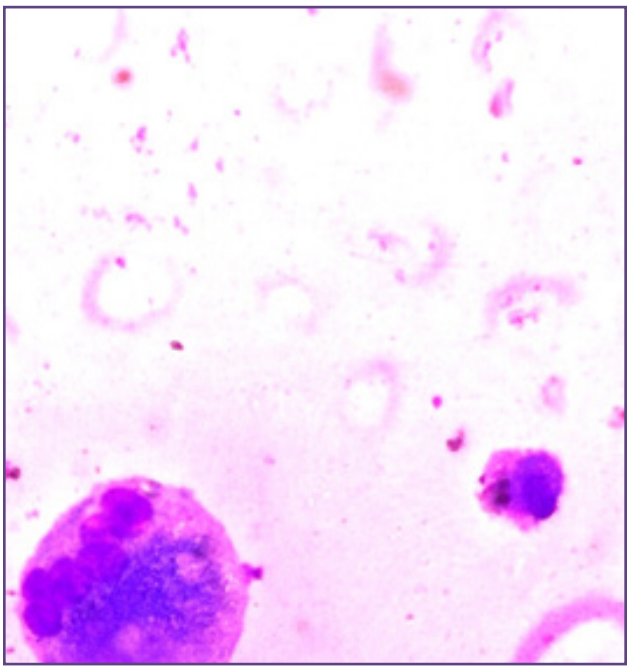

Fig. 4: High power view of cytological smear of pleural fluid showing many foamy histiocytes and a multinucleated giant cell. (LeishmannX400).

Discussion

Pseudochylothorax (cholesterol pleurisy or chyliform effusion) is a cholesterol-rich pleural effusion that is commonly associated with chronic inflammatory disorders like Tuberculosis and Rheumatoid Arthritis.[4] It is usually seen in long standing exudative pleural effusions, in which the effusion is encapsulated in a fibrotic part of the thickened pleura.

The precise pathogenesis of chyliform pleural effusions is unknown. But various hypotheses have been postulated. In one of these, the lipid in chyliform effusions is thought to arise from breakdown of blood cells.[2] The fibrotic scar tissue, which walls off the effusion, is poorly vascularized. [5] This is thought to result in an abnormally slow transfer of cholesterol and other lipids out of the pleural space and lead to the accumulation of cholesterol in the pleural fluid. [2] In contrast to the cholesterol in acute exudates, which is mostly bound to Low Density Lipoproteins (LDL), cholesterol in Pseudochylous effusions is bound to High Density Lipoproteins (HDL). This would imply that it is derived from serum lipoproteins rather than cellular debris. To explain this anomaly, it has been hypothesized that cholesterol which enters the pleural space with acute inflammation, becomes trapped there and undergoes change in lipoprotein binding characteristics .[6]

Wrightson et al. presented six well-characterized cases of arthritis-associated pseudochylothorax, each notable due to their minimal pleural thickening. The median duration of symptoms (or arthritis, in the case of asymptomatic effusions) in their study was 15 months. The absence of pleural thickening in their patients suggested that cholesterol accumulates due to an acute or subacute process, rather than cellular breakdown and subsequent release of cholesterol within the pleural space. These unusual findings were a deviation from the pseudochylothorax seen in RA. Hence, it was suggested that pseudochylothorax should be considered clinically even in short duration nonfibrotic pleural effusions .[4]

Pseudochylous effusions need to be differentiated from the true chylous effusions and empyema. All three entities have in common, a thick milky white and opalescent fluid. Empyema can be differentiated from the other two entities by performing centrifugation. In lipid effusions, the fluid remains uniform unlike the clear supernatant that develops in empyema. Pseudochylothorax can be differentiated from chylothorax by adding 1–2ml of ethyl ether. The milkiness disappears in the former . [7] Differentiating features of various types of turbid pleural effusions are summarized in Table 1.

In our case, the diagnosis of rheumatoid pleurisy was confirmed on the basis of increased protein, decreased glucose, raised cholesterol, decreased triglyceride level, characteristic cytology and presence of cholesterol crystals

Table 1. Differentiating features of turbid pleural effusions

Type of effusion	Gross appearance	Appearance after centrifugation	Chief biochemical feature of effusion fluid
Empyema	Turbid	Clear supernatant	Glucose<60mg/dl
Chylous	Turbid	Fluid remains uniformly milky white	Triglycerides >110mg/dl
Pseudochylous	Turbid	Fluid remains uniformly milky white	High content of cholesterol>200mg/dl with no triglycerides

on wet mount preparation.[9,10] Tuberculosis was ruled out on the basis of negative ADA levels and culture.

Management of the underlying disease process is the mainstay of treatment. Although at times, the effusions may not accumulate after initial thoracocentesis, the majority require repeated drainage [12]. Decortication should be considered in symptomatic patients with restrictive lung function .[11]

Conclusion

A pseudochylous pleural effusion is an uncommon disease condition developing as a sequela of a long standing exudative effusion, commonly seen in Tuberculosis and Rheumatoid Arthritis. The conventional pathogenesis is cholesterol accumulation as a result of cellular breakdown, leading to elevated cholesterol levels in the pleural fluid. The diagnosis is confirmed by demonstration of raised cholesterol and reduced triglyceride levels in the fluid, along with visualization of characteristic cholesterol crystals on wet mount. Radiologically, in most cases, including ours, the pleura are markedly thickened.

It is important to differentiate cholesterol effusion from chylothorax and empyema because each of these entities are managed differently. In pseudochylous effusions, the mainstay of treatment is management of the underlying cause. Repeated thoracocentesis and decortications may be required in selected cases.

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Competing Interests

None

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