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CASE REPORT

Chylothorax as the initial presentation of systemic lupus erythematosus: a case report

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Pulmonary involvement in systemic lupus erythematosus (SLE) is common, and all the elements of the respiratory system can be affected. Chylothorax is a rare manifestation of SLE, and only three cases have been reported in English literature [1, 2]. In this study, we describe a case of SLE whose initial presentation was in the form of chylothorax.

Case presentation

A previously healthy 43-year-old woman presented with a 2-month history of dry cough. She denied constitutional symptoms such as fever, malaise, poor appetite, or body weight loss. Physical examination revealed no abnormalities. A chest radiograph showed blunting of the left costophrenic angle, suggesting the presence of pleural effusion. Thoracocentesis yielded 30 ml of odorless milky fluid containing 4,608/mm^3 white blood cells with a differential of 59% lymphocytes, 6% neutrophils, 2% eosinophils, and 33% tissue cells. Biochemical analysis of the pleural fluid demonstrated the following: total protein, 7.8 g/dl; lactate dehydrogenase, 228 U/l; glucose, 86 mg/dl; amylase, 29 U/l; cholesterol, 89 mg/dl; and triglyceride, 857 g/dl, supporting the diagnosis of chylothorax [3]. Gram's stain, acid-fast stain, and culture of bacteria and mycobacteria all gave negative results. Cytology of the pleural fluid was negative for malignant cells. The computed tomographic (CT) scan of chest demonstrated small amounts of left-sided pleural effusion without evidence of pulmonary parenchyma lesion or mediastinal lymphadenopathy. There was no history of antecedent chest trauma or thoracic surgery. Further immunological studies of serum showed positive for antinuclear antibodies (ANA) at a titer of 1:1,280 with diffuse chromatin and fine and large speckled pattern. Tests for anti-dsDNA, anti-SSA/Ro, anti-SSB/La, anti-Sm, and anti-RNP antibodies all gave positive results. A test for ANA of the pleural fluid was also positive. The diagnosis of SLE was made by fulfilling the 1997 ACR criteria of serositis, lymphopenia, and positive results for anti-dsDNA, anti-Sm, and ANA. She was treated with prednisolone 20 mg and hydroxychloroquine 400 mg daily. Her cough tailed off within a few days, and the chylothorax cleared rapidly. At present, she is treated alone with 50 mg of azathioprine daily, and there has been no recurrence during a follow-up period of 4 years.

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Discussion

Chylothorax is defined as the accumulation of chyle within the pleural space due to disruption of the thoracic duct or one of its main divisions. The diagnosis is made by analysis of the fats within the pleural fluid. Triglyceride levels greater than 110 mg/dl are highly suggestive of a chylous effusion [3]. In our patient, the high triglyceride content (857 mg/dl) in pleural fluid supports the diagnosis of chylothorax.

In addition to establishing the presence of chylothorax, further evaluation concerning the underlying disease should be conducted. Surgical procedures and chest trauma account for most cases of chylothorax. If there is no history of trauma or surgery, i.e., nontraumatic chylothorax, malignancy is a leading cause and should be excluded first. Other less common etiologies of chylothorax encompass liver cirrhosis, nephrotic syndrome, congestive heart failure, lymphangioleiomyomatosis, and infectious diseases such as filariasis, tuberculosis, and histoplasmatosis. Some connective tissue diseases have also been associated with chylothorax, including sarcoidosis [4], Behcet's disease [5], and SLE [1, 2]. Strausser and Flye [1] described a 24-year-old woman with an established diagnosis of SLE complicated by a left-sided chylothorax and chylous ascites. Lee et al. [2] reported two cases with previously undiagnosed SLE presenting as bilateral chylothorax, chylous ascites, and protein-losing enteropathy. The concurrence of malar rash and/or proteinuria prompted further investigation, leading to the diagnosis of SLE. In contrast to the three cases described above, our patient had chylothorax as the single clinical presentation. Identification of SLE as the underlying disease facilitated appropriate corticosteroid therapy to resolve her chylothorax.

Because of the rarity, the mechanism of SLE causing chylothorax is not well understood, and theories about its pathophysiology are lacking. Owing to the dramatic response to corticosteroids in our case, we speculate that the mechanism might be related to the inflammatory process of the thoracic duct or its divisions, i.e., lymphangitis, causing damage to lymphatic ducts and the subsequent chyle leakage.

In conclusion, chylothorax is an exceedingly rare manifestation of SLE. Nevertheless, it could be the first and sole clinical presentation of SLE as described in this case. We suggest that in patients with idiopathic chylothorax, SLE should be considered as a possible cause.

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