CASE STUDIES

Massive pleural effusion in a patient with systemic lupus erythematosus

Durdu Mehmet Yavsan[1], Ali Yavuz Karahan[2], Muhammed Sahin[2], Tayfun Gungor[2], Adem Kucuk[3]*, Emin Maden[1], Recep Tunc[3]

[1]Departments of Chest Diseases, Meram School of Medicine, Necmettin Erbakan University, Meram, Konya, Turkey
[2]Department of Physical Medicine and Rehabilitation, Meram School of Medicine, Necmettin Erbakan University, Meram, Konya, Turkey
[3]Department of Internal Medicine, Division of Rheumatology, Meram School of Medicine, Necmettin Erbakan University, Meram, Konya, Turkey

Abstract

Systemic lupus eryhtematosus (SLE) is a chronic systemic inflammatory disease of unknown etiology, which may cause life-threatening tissue and organ damage. Pleural effusion in SLE tends to be small and bilateral. The present case study focuses on the medical management of a 24-year-old SLE patient with massive pleural effusion who presented with cough and shortness of breath.

Introduction

Systemic lupus eryhtematosus is a chronic systemic inflammatory disease of unknown origin that causes an overactive immune system. It usually affects women of childbearing age.[1-2] SLE may manifest itself with several symptoms including fever, fatigue, joint pain, mild rash, and arthritis. It can also cause life-threatening damage to tissues and organs (such as kidneys or heart).[1] Pleural effusion affects 16-50% of SLE patients and tends to be small and bilateral.[3] The aim of this case report is to draw attention to medical management of massive pleural effusion, which rarely occurs in SLE patients.

Case report

A 24-year-old female patient presented with shortness of breath and cough that became worse with exercise during the previous month. She reported a history of photosensitivity and episodes of intermittent arthritis, which affected her wrists and proximal interphalangeal joints. She did not have any family history of connective tissue disease. She rarely consumed alcohol and smoked 4 packs of cigarettes per year. Her heart sounds were distant upon physical examination. During ausculation, the respiratory sounds could not be obtained from the middle area of the left hemithorax and onwards, and the percussion sounds were dull. Lymphadenopathies were present in the bilateral submandibular area. Chest radiography revealed elevated cardiothoracic index as well as increased density in the middle and lower zones of the left hemithorax, which was consistent with pleural effusion (Fig. 1). A left hemithorax thoracentesis was performed for therapeutic and diagnostic purposes, which revealed that the thoracentesis fluid appeared serous, which met the Light criteria for exudative pleural effusion (Table 1). Cytological examination of the pleural fluid did not reveal signs of malignancy, nor were acid-resistant bacilli present in the pleural fluid. Direct Gram staining and culture were negative for bacteria and fungi.

Results of arterial blood gas analysis were as follows: pH - 7.44, PCO2 - 33.7, PO2 - 81.8, and HCO3 - 22.7. Therefore, the patient appeared to have metabolic acidosis. Whole blood analysis showed the following: hemoglobin level - 8.4 g/dl, platelet count - 420x10^3/u, MCV - 65.2 fL, red blood cell count - 4.38x10^6/uL, red cell distribution width - 13.2%, and ferritin - 132 ng/mL. Her partial thromboplastin time was 37.1 and the international normalized ratio was 1. Hemoglobin electrophoresis revealed hemoglobin A2 to be 5.3%, which was consistent with thalassemia minor. Complete urinalysis did not show any particular feature. Rheumatologic blood tests were highly positive. The anti-nuclear antibody was granular and homogenous (1:320 with indirect fluorescent antibody). Additionally, she was found to be positive for anti-dsDNA, anti-Smith (Sm) and anticardiolipin IgM antibodies. Complement

levels were within normal range, and rheumatoid factor was negative. Protein electrophoresis revealed hypergammaglobulinemia, IgG of 33.5 g/L, IgM of 2.06 g/L, and IgA of 4.3 g/L. ECG revealed right bundle branch block, while echocardiography showed 60% ejection fraction and minimal pericardial effusion. Multiple lymph nodes with hilar structures were observed during the neck ultrasonography, with the largest nodes being 13x5 mm at the right side in the mandibular zone and 20x6 mm at the left side.

The patient was treated with hydroxychloroquine, acetylsalicylic acid, methylprednisolone, and azathioprine. High-dose, pulse intravenous methylprednisolone of 1 g was given daily for three days and the dose was decreased gradually. Her respiratory symptoms and pleural effusion were considerably regressed during the follow-up visits (Fig. 2). In addition, regression of bilateral lymphadenopathies was observed.

Discussion

SLE, an autoimmune disorder affecting multiple organs, predominantly occurs in women. Isolated pulmonary involvement is infrequent in SLE. Although pleuritis is the most common respiratory symptom, pleural effusion occurs at a rate of 1-2% as the initial sign in SLE patients. Pleuritis is more commonly observed in patients with late-onset SLE.[4-5] Typical signs of pleural involvement include acute chest pain accompanied by fever, cough,

Fig 1: Increased density in the middle and lower zones of the left hemithorax, which was consistent with pleural effusion

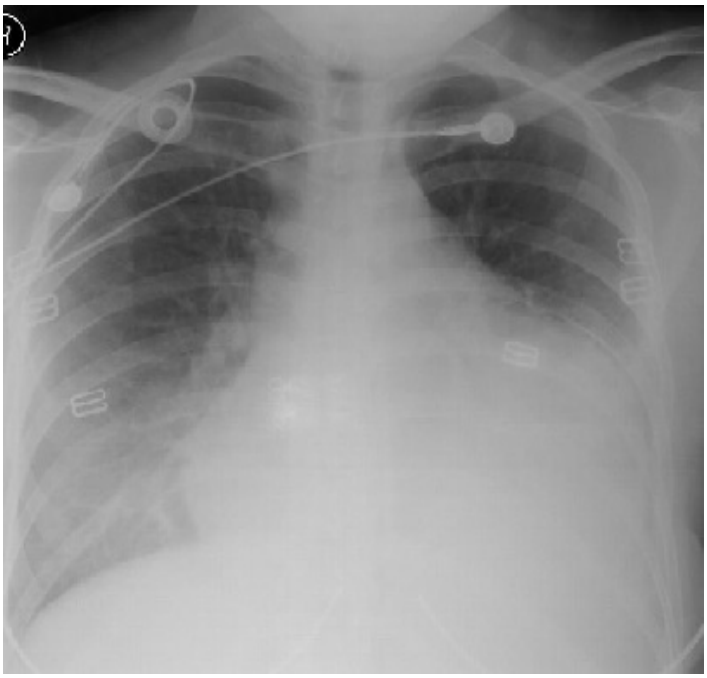
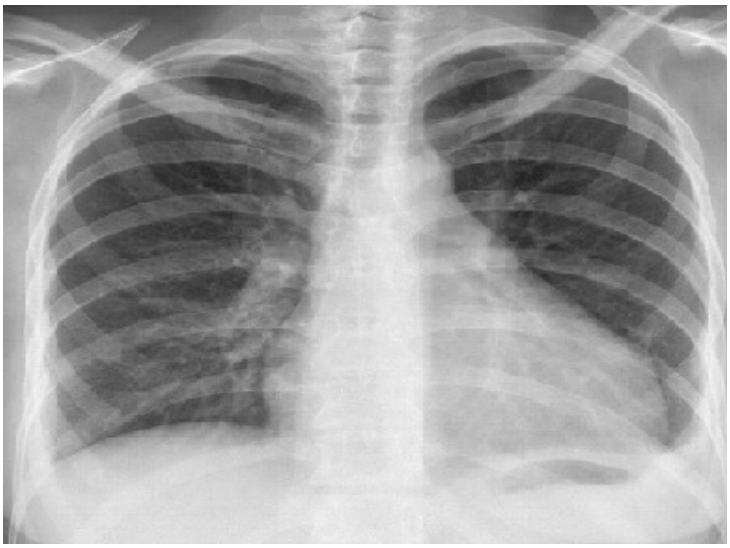


Table 1: Serum and pleural fluid characteristics of the patient

Parameters	Serum level	Pleural fluid level	Light criteria (Pl. fluid/serum)
Glucose	83 mg/dl	69 mg/dl	
Lactic dehydrogenase	333 u/l	684 u/l	>0.6
Total protein	8.6 g/dl	5.8 g/dl	>0.5
Albumin	3.2 g/dl	2.4 g/dl	

Fig. 2: Chest X- ray after treatment



and dyspnea. Although the occurrence of serositis is frequent, the cause of pleural effusion in SLE is usually renal, cardiac or pulmonary embolism. Pleural effusion is typically small and bilateral, and is rarely massive. In SLE, pleuritis develops as a result of inflammation due to the production of immune complex and complement activation. Pleural fluid specimens often have protein levels above 3.5 g/dL, LDH levels below 500 U/L, and decreased glucose concentrations. In addition, low complement levels and high anti-nuclear antibody levels (>1/160) are suggestive of SLE. However, ANA titers may also increase in neoplastic effusions.

Lupus eryhtematosus (LE) cells are highly specific.[5-10] There are different manifestations of respiratory system involvement in SLE, in addition to pleuritis (with or without effusion). One end of the wide spectrum comprises of interstitial lung diseases, acute lupus pneumonitis, diffuse alveolar hemorrhage, pulmonary arterial hypertension, and mostly pulmonary infections. Whereas, the other end includes minor conditions of lesser concern like acute reversible hypoxemia.[6, 11] In the present case report, the patient had shortness of breath, cough, arthritis, lymphadenopathy, and photosensitivity. She also reported that she used to consume alcohol and smoke cigarettes. Smoking generally increases the risk of lupus flares. On the other hand, moderate/reasonable alcohol intake has been reported to confer a protective effect.[12-14]

Even though there are many potential causes of pleural effusion and congestive heart failure (CHF), the most common etiologic factors include malignancy, pneumonia and pulmonary embolism. Several disorders may cause effusion, despite the involvement of many different organs. History and a careful physical examination are very helpful for diagnosis. Weight loss and hemoptysis suggest malignancy, and fever may be indicative of empyema, pneumonia and tuberculosis. However, articular, ophthalmic and skin symptoms suggest connective tissue disorders.[15-17]

If pleural effusion observed in a SLE patient is small and asymptomatic, no treatment is required. For cases with mild effusion, non-steroidal anti-inflammatory drugs (NSAIDs) may be prescribed. Patients with more severe disease should receive steroid therapy. Hydroxychloroquine and azathioprine should also be given to selected cases. While SLE often responds to medical treatment, refractory cases have also been reported. There is no specific treatment regimen for resistant cases.[3]

In summary, connective tissue disorders and SLE are mainly responsible for the development of pleural effusion, particularly among younger women. Here, we described a case of SLE with a rare cause of massive pleural effusion. Marked bilateral effusion responded well to treatment with azathioprine 2.5 mg/kg, hydroxychloroquine and steroid, and did not require pleurodesis. A limitation of this case report was the failure to perform analyses for ANA and LE cell on the diagnostic thoracentesis fluid.

Competing interests

The authors declare that they have no competing interests.

Citation
Yavsan DM, Karahan AY, Sahin M, Gungor T, Kucuk A, Maden E, Tunc R. Massive pleural effusion in a patient with systemic lupus erythematosus. IJRCI. 2015;3(1):CS1.

Received: 1 October 2014, **Accepted:** 31 December 2014, **Published:** 8 January 2015

* Correspondence: Dr. Adem Kucuk, Division of Rheumatology, Department of Internal Medicine, Meram School of Medicine, Necmettin Erbakan University, Meram, Konya, Turkey
drademk@yahoo.com

References

1. Askanase A, Shum K, Mitnick H. Systemic lupus erythematosus: an overview. Soc Work Health Care. 2012;51(7):576-86.
2. Ko HS, Ahn HY, Jang DG, Choi SK, Park YG, Park IY, et al. Pregnancy outcomes and appropriate timing of pregnancy in 183 pregnancies in Korean patients with SLE. Int J Med Sci. 2011;8(7):577-83.
3. Breuer GS, Deeb M, Fisher D, Nesher G. Therapeutic options for refractory massive pleural effusion in systemic lupus erythematosus: a case study and review of the literature. Semin Arthritis Rheum. 2005 Apr;34(5):744-9.
4. Wan KS. Pleuritis and pleural effusion as the initial presentation of systemic lupus erythematous in a 23-year-old woman. Rheumatol Int. 2008 Oct;28(12):1257-60.
5. Gulhane S, Gangane N. Detection of lupus erythematosus cells in pleural effusion: An unusual presentation of systemic lupus erythematosus. J Cytol. 2012 Jan;29(1):77-9.
6. Ferreiro L, Alvarez-Dobano JM, Valdes L. Systemic diseases and the pleura. Arch Bronconeumol. 2011 Jul;47(7):361-70.
7. Gonzalez Leija ME, Orrantia Vertiz M, Vazquez Zaragoza MA. [Massive pleural effusion in systemic lupus erythematosus]. Reumatol Clin. 2009 Nov-Dec;5(6):289-90.
8. Good JT, Jr., King TE, Antony VB, Sahn SA. Lupus pleuritis. Clinical features and pleural fluid characteristics with special reference to pleural fluid antinuclear antibodies. Chest. 1983 Dec;84(6):714-8.
9. Wang DY, Yang PC, Yu WL, Kuo SH, Hsu NY. Serial antinuclear antibodies titre in pleural and pericardial fluid. Eur Respir J. 2000 Jun; 15(6):1106-10.
10. Khare V, Baethge B, Lang S, Wolf RE, Campbell GD, Jr. Antinuclear antibodies in pleural fluid. Chest. 1994 Sep;106(3):866-71.
11. Pego-Reigosa JM, Medeiros DA, Isenberg DA. Respiratory manifestations of systemic lupus erythematosus: old and new concepts. Best Pract Res Clin Rheumatol. 2009 Aug;23(4):469-80.
12. Sibilia J. Treatment of systemic lupus erythematosus in 2006. Joint Bone Spine. 2006 Dec;73(6):591-8.
13. Kiyohara C, Washio M, Horiuchi T, Asami T, Ide S, Atsumi T, et al. Cigarette smoking, alcohol consumption, and risk of systemic lupus erythematosus: a case-control study in a Japanese population. J Rheumatol. 2012 Jul;39(7):1363-70.
14. Ghaussy NO, Sibbitt W, Jr., Bankhurst AD, Qualls CR. Cigarette smoking and disease activity in systemic lupus erythematosus. J Rheumatol. 2003 Jun; 30(6):1215-21.
15. Diaz-Guzman E, Budev MM. Accuracy of the physical examination in evaluating pleural effusion. Cleve Clin J Med. 2008 Apr;75(4):297-303.
16. McGrath EE, Anderson PB. Diagnosis of pleural effusion: a systematic approach. Am J Crit Care. 2011 Mar;20(2):119-27; quiz 28.
17. Tekeoglu I, Hiz O, Ozbay B, Toprak M, Avcu S. Pulmonary involvement in rheumatic diseases: HRCT findings. J Clin Anal Med 2011;2:36-39.