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Case Report

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Systemic Lupus Erythematosus With Pulmonary Artery Aneurysm

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ABSTRACT

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disorder, which rarely presents with pulmonary artery aneurysm. This report presents a 51-year-old female, known case of SLE for 20 years, presented with dyspnea, productive cough, and hemoptysis. The patient was diagnosed as having a pulmonary artery aneurysm, and managed with medical therapy and follow up instead of surgery.

Introduction

Systemic Lupus Erythematosus (SLE) is a chronic inflammatory disease that has many clinical manifestations, including lung diseases [1, 2]. The lungs are commonly involved later than other organs [3]. Most common pulmonary manifestations of SLE are the pleural disease (pleural effusion, pleurisy), parenchymal disease (acute and chronic pneumonitis, diffuse alveolar hemorrhage), vascular involvement (acutely reversible hypoxemia, pulmonary hypertension, and embolism) and airway disease [4, 5].

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Pulmonary artery aneurysm is uncommon and is associated with a vascular anomaly, vasculitis, pulmonary hypertension, and cardiac anomalies [6]. Pulmonary artery aneurysm as a presentation of SLE is very rare. We present a case of calcified pulmonary artery aneurysm as a presentation of SLE. The combination of pulmonary artery aneurysm and systemic lupus erythematosus, even in the absence of any other pulmonary manifestation, should always suggest SLE as a differential diagnosis.

Case Presentation

A 51-year-old female, known case of SLE from 20 years ago, was admitted to the emergency department with a 10-day history of increasing shortness of breath, productive cough, and hemoptysis without fever and shivering. Her past medical history was unremarkable, except for SLE, which was diagnosed in 1996. Fever, malar rash, fatigue, polyarthritis, proteinuria, hematuria, and stage IV of lupus nephritis were the first presentations of the disease¬. The patient treated with cyclophosphamide and methylprednisolone pulses in the first hospitalization. Preadmission drugs included hydroxychloroquine, prednisone, losartan, pantoprazole, and alendronate.

On arrival, her blood pressure was 100/65mm Hg, heart rate 80 beat/minute, respiratory rate 24 breath/minute, O^2 saturation 89% without oxygen and 92% with nasal oxygen and temperature 37°C axillary. On physical examination, coarse crackle and wheezing were detected throughout two lungs. Cardiac auscultation was normal without murmurs and rubs. The rest of her physical examination was not notable, and she had no apparent skin rash, ulcer, and organomegaly.

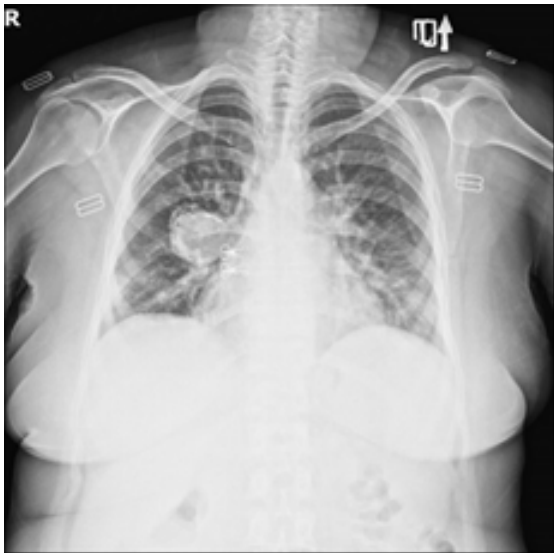

Figure 1. Opacity with a sharp border in mid-zone of the right lung

The laboratory evaluation revealed normal findings except anemia and trace proteinuria in urine analysis. Chest radiography on admission showed opacity with a sharp border in mid-zone of the right lung (Figure 1). Multislice CT angiography of the thoracic aorta with contrast ruled out pulmonary thrombi embolism but showed aneurysmal dilation of lobar branches of the right pulmonary artery (in maximum AP diameter of 34 mm) which containing soft and calcified plaques (Figure 2). Patchy ground glasses opacity in the peripheral and subpleural region of both lungs were seen that were diagnosed as pneumonia and treated.

No mediastinal or hilar lymphadenopathy was seen. There was no evidence of pleural effusion and thickening. The echocardiogram confirmed pulmonary artery pressure which was 35mm Hg. Following vascular sur-

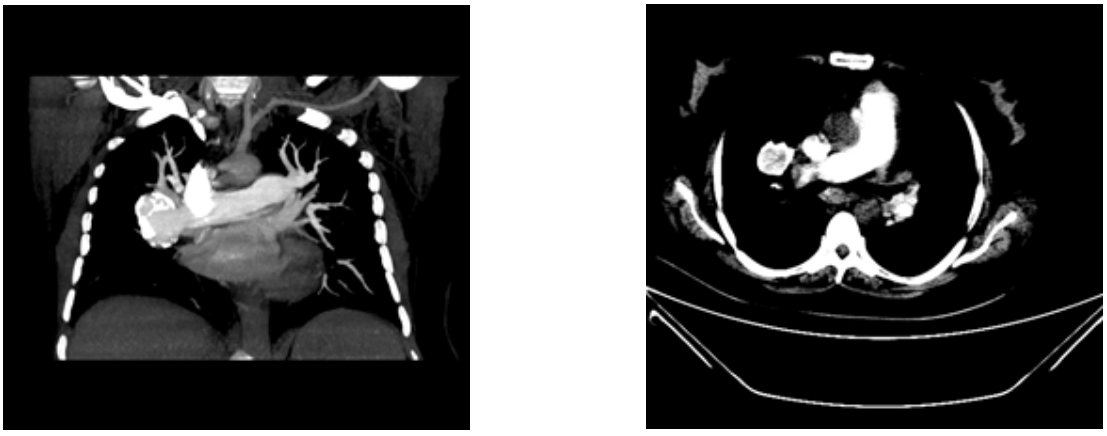

Figure 2. Aneurysmal dilation of lobar branches of the right pulmonary artery, containing soft and calcified plaques

gical consultation due to the patient's tolerance and calcification of an aneurysm, it was decided that no surgical intervention was warranted. She was followed up at the clinic.

Discussion

Pulmonary artery aneurysms (PAAs) are rare, especially calcified aneurysm [7]. PAAs are categorized into congenital, acquired, and idiopathic based on their causes. Congenital causes are the primary reason for PAA formation. There are different infectious causes for PAA such as syphilis, tuberculosis, pyogenic bacteria, pneumonia, and so on. Other causes included systemic vasculitis (Hughes-Stovin disease and Behcet's disease), collagen vascular diseases, connective tissue disorders, and idiopathic. Most patients present with nonspecific signs and symptoms such as hemoptysis, chest pain, cough, and dyspnea [6, 8, 9].

Although angiography is the gold standard for the diagnosis, it is invasive. So we can use spiral CT as an excellent diagnostic modality [8]. Treatment can be either medical or surgical. The indications for surgery are absolute diameter more than 5.5cm, compression of adjacent structures, thrombus formation in the aneurysm sack, the appearance of clinical symptoms and sign of rupture or dissection [6].

In our patient, due to an indication of surgery, patient's tolerance, and calcification, we preferred medical therapy and observation. In conclusion, pulmonary vascular manifestations of SLE are mostly pulmonary hypertension, thromboembolism, and acute reversible hypoxemic syndrome [10]. Pulmonary artery aneurysm as a pulmonary manifestation of SLE is an infrequent entity, but it should be considered.

Ethical Considerations

Compliance with ethical guidelines

All ethical principles were considered in this article.

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Conflict of interest

The authors declared no conflict of interest.

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