Hughes-Stovin Syndrome: An Unusual Cause for Recurrent Hemoptysis

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Abstract

A 24 year old female presented with recurrent hemoptysis and the work up revealed a large right pulmonary artery aneurysm for which she underwent endovascular intervention with relief of symptoms. Her symptoms recurred after one month and now she had left pulmonary artery aneurysm, again treated with endovascular intervention. With systemic symptoms and absence of signs of Behcets syndrome led to diagnosis of Hughes-Stovin Syndrome. She symptom free at 18 months with immunosuppression.

Keywords: Behcet syndrome, Hughes-Stovin syndrome, pulmonary artery aneurysm, vasculitis

Introduction

Hemoptysis has varied etiology. The most common cause would be tuberculosis in tropical countries. Here, we present a patient with recurrent hemoptysis, initially thought to be tuberculosis. Later, pulmonary artery aneurysm was diagnosed and intervened with vascular plug. After 4 weeks, recurrence of pulmonary artery aneurysm on the contralateral side as well as right femoral vein thrombotic occlusion was detected and intervened again with vascular plug. The patient had symptoms such as fever, cough, hemoptysis, as well as venous thrombosis, and recurrent pulmonary artery aneurysms which were suggestive of vasculitis. Finally, it was diagnosed as a rare syndrome called Hughes-Stovin syndrome.

Case Report

Our patient is a 24-year-old unmarried female with no comorbidities. She presented with complaints of fever of 8 months duration with cough, loss of weight, and loss of appetite. She had recurrent hemoptysis which was of moderate quantity for the past 6 months. She underwent multiple investigations elsewhere and was put on anti-tuberculosis treatment. She presented to our department with massive hemoptysis. Her blood investigations were within normal limits. Her coagulation profile was normal and workup for systemic lupus erythematosus and antiphospholipid antibody syndrome was negative. Her chest X-ray showed homogenous radiopaque lesion in the right lower lobe [Figure 1a]. Her computed tomography (CT) scan chest revealed large pulmonary artery aneurysm in the right side [Figure 1b]. Pulmonary angiogram was done through the right femoral vein access which revealed inferior branch of right pulmonary artery as the feeder for the large aneurysm [Figure 1c]. The feeder artery was closed with 8 mm Vascular plug II (AMPLATZER™-St. Jude Medical, Minnesota) [Figure 1d]. The patient was symptom-free and discharged with the advice to continue anti-tuberculosis treatment.

One month later, the patient again presented with hemoptysis and fever. Chest X-ray and CT scan chest revealed a similar new pulmonary artery aneurysm in the left side [Figure 2a and b]. Pulmonary angiogram revealed a large pulmonary artery aneurysm in left side and also right iliac, femoral vein, inferior vena cava were occluded with thrombus [Figure 2c and d]. This time access was taken from...
the left femoral vein and IVC was predilated to clear of the thrombus and the aneurysm feeder was occluded again with 8 mm Vascular plug II (AMPLATZER™-St. Jude Medical, Minnesota) [Figure 2e]. Her symptoms such as fever, cough, recurrent hemoptysis, recurrent pulmonary aneurysm, and thrombophlebitis with the absence of signs of Behcet syndrome led to the diagnosis of Hughes-Stovin syndrome. She was put on cyclophosphamide this time and discharged. Now, the patient is in follow-up for almost 18 months without any recurrence of hemoptysis.

**DISCUSSION**

Hughes-Stovin syndrome is characterized by multiple aneurysms in pulmonary artery and venous thrombosis involving the peripheral veins, inferior vena cava, and even right atrium. Males aged between 12 and 40 are more affected. There have been only few reports (<40 cases) in literature. Symptoms include fever, cough, hemoptysis, recurrent hemoptysis, recurrent pulmonary aneurysm, and thrombophlebitis with the absence of signs of Behcet syndrome led to the diagnosis of Hughes-Stovin syndrome. She was put on cyclophosphamide this time and discharged. Now, the patient is in follow-up for almost 18 months without any recurrence of hemoptysis.

Our case had symptoms of cough, fever, hemoptysis, and signs of pulmonary artery aneurysm and venous thrombosis. Our diagnosis of Hughes-Stovin syndrome was based on the clinical criteria for the syndrome as there are no laboratory diagnostic criteria for the same. Behcet syndrome was ruled out because of the lack of signs such as oral and genital ulcers.

Pulmonary artery thrombus can also occur resulting from in situ thrombus formation following inflammation of the arterial wall. Pulmonary artery aneurysms can also occur secondary to inflammation and weakening of the vessel wall.

Hemoptysis is the leading cause of death in these patients. Hemoptysis may be due to pulmonary or bronchial arterial aneurysm rupture. Conventional angiography is the standard investigational modality to study about the aneurysm.

Hughes-Stovin syndrome is treated by immunosuppressants. Corticosteroids or combination of cyclophosphamide and corticosteroids are often given. Steroids, cyclophosphamide, or cyclosporine are given for managing venous thrombosis. Cyclophosphamide and steroids are given for the management of pulmonary artery and peripheral artery aneurysms. It has been recommended by European League Against Rheumatism to continue cyclophosphamide for 2 years followed by azathioprine for pulmonary artery aneurysms. Hemoptysis and small pulmonary artery aneurysms most often respond. Anticoagulants for pulmonary artery thrombus and peripheral venous thrombus are used with caution because of hemoptysis. Surgery of the affected lung segment is of high risk. Hence, transcatheter interventions such as embolization can be offered. In our case, we used vascular plug device successfully to treat pulmonary artery aneurysms.

**CONCLUSION**

Although tuberculosis is the most common cause for...
hemoptysis in tropical countries, we need to rule out very rare causes such as Behcet syndrome and Hughes-Stovin syndrome. Our patient had signs and symptoms of Hughes-Stovin syndrome and was intervened percutaneously for massive hemoptysis.

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Conflicts of interest
There are no conflicts of interest.

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