

Case Report

Insidious Hughes Stovin Syndrome: Journey From Pulmonary Embolism to Pulmonary Arterial Aneurysm

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Abstract

Hughes-Stovin syndrome (HSS) is a rare disease characterized by deep vein thrombosis and pulmonary and / or bronchial artery aneurysms. A 28-year-old female patient was followed-up for 5 months with pulmonary embolism. When she presented with hemoptysis at her 5th month, pulmonary artery aneurysm was detected on thoracic CT angiography. The abdominal magnetic resonance (MR) angiography revealed thrombus in the inferior vena cava. Because she didn't carry the criteria for Behçet's disease, she was diagnosed as Hughes-Stovin Syndrome (HSS) and steroid treatment was started. The patient who had a response to the treatment, stay in remission for a long time. The prognosis was poor in patients with HSS, and aneurysmal rupture was the main cause of death. In order to emphasize the fact that, when the pulmonary arterial aneurysm is seen, in view of the possibility of Behçet or its variant HSS, rapid onset of treatment can be life-saving. A rare female case is presented in the light of the literature.

KEYWORDS: Hughes stovin syndrome, hemoptysis, aneurysm

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INTRODUCTION

Hughes-Stovin Syndrome (HSS) was first described in 1959 [1]. It is a very rare disease characterized by deep vein thrombosis (DVT) and pulmonary and/or bronchial artery aneurysms. There are no defined diagnostic criteria due to the rarity of the disease. It is diagnosed when an aneurysm associated with DVT and other conditions that may cause this is ruled out [2]. It has been described as "incomplete Behçet disease" because of its clinical and histopathological similarities with Behçet disease. Thus, these two pathophysiological conditions may not be differentiated in terms of pulmonary involvement [3, 4]. We presented a young woman patient with pulmonary artery aneurysms who was first diagnosed as pulmonary thromboembolism to emphasize the fact that, when the pulmonary arterial aneurysm is seen, the possibility of HSS must be kept in mind so that early treatment can be life-saving.

CASE PRESENTATION

A 28-year-old female patient was admitted to the emergency department with complaints of fever, right side pain, hemoptysis and fatigue for 5 days. Vital signs, complete blood count and biochemical tests were normal. The chest X-ray was normal (Figure 1). Wells score for pulmonary thromboembolism (PTE) was considered as moderate probability. The D-dimer was 5145 ng/ml. The thoracic CT showed filling defects in the segmental branches of the right pulmonary artery (Figure 2). Low molecular weight heparin (LMWH) and vitamin K antagonist (VKA) were started. The treatment continued with VKA.

3 months later, the new embolus was detected. Because PTE developed under VKA treatment, LMWH was restarted. She was examined for possible reasons for recurrent PTE. Anticardiolipin and antiphospholipid antibodies and all other rheumatologic auto-antibodies were negative. Tumor markers CA-125, CA-15-3 and carcinoembryonic antigen (CEA) were negative. There was no mutation for genetic risk factors for PTE. The treatment continued with LMWH.

In the 5th month of LMWH, the patient applied to the emergency department with hemoptysis (200 cc/day), dyspnea. There was bilateral hilar enlargement on the chest X-ray.

Thrombotic aneurysmatic dilatations were observed in the left pulmonary artery on the thoracic CT (Figure 3. a, b).

The patient consulted with rheumatology. Recurrent oral ulceration, recurrent genital ulceration, eye lesions, skin lesions and positive pathergy test which are necessary for the diagnosis of Behçet's disease were not present in the patient.

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On the thoracic MR angiography, it was reported that there was an aneurysm in 3.5 cm long and 3 cm wide with complete thrombus in the left pulmonary artery. A similar appearance was seen in the right pulmonary artery as non-aneurysmatic with short segment thrombosis.

The abdominal MR angiography showed thrombosis of the inferior vena cava. There was a mobile, 2.4x0.9 cm mass (thrombus?) with peduncle attached to the interatrial septum in the right atrium on Echocardiography (ECHO).

In light of all these data, the patient was diagnosed as HSS. Because the patient had massive hemoptysis, the pulse steroid treatment (500 mg/day) was given for 5 days. Afterward, maintenance treatment was continued with prednisolone. No hemoptysis was observed during follow-up.

In the 3rd month of the treatment, bilateral hilar enlargement was significantly decreased on the chest X-ray. The thoracic CT angiography showed that thrombotic aneurysmatic dilations of the pulmonary artery were reduced (Figure 4. a, b). There was no thrombus in the right atrium that was seen in the previous ECHO.

It was learned that the patient, who did not come to follow-up, discontinued her medication because she got pregnant and applied to an emergency department of a hospital with massive hemoptysis and intubated and followed in the intensive care unit. The thoracic CT angiography performed at that center revealed an aneurysm with a diameter of approximately 30 mm in the left pulmonary artery. With these results, it was learned that methylprednisolone 1000 mg/day for 3 days and then methylprednisolone 0.8 mg/kg/day was continued with maintenance therapy and the pregnancy was terminated as abortion. In addition to corticosteroid treatment, infliximab (5 mg/kg) was administered to the patient at 0, 2 and 6th weeks. The patient, who is still in the third month of new corticosteroid therapy, is in our follow-up and stable.

Written informed consent was obtained from the patient who participated in this case.

DISCUSSION

Most of pulmonary artery aneurysms are associated with congenital cardiovascular lesions, but include less common cases with degenerative changes in the pulmonary arteries with syphilis or trauma. Multiple pulmonary artery aneurysms are generally observed in patients with Behçet disease or HSS [5, 6]. HSS is observed in three clinical stages: thrombophlebitis symptoms (first stage), pulmonary artery aneurysms forma-

tion (second stage), and aneurysm rupture with massive hemoptysis (third stage), which usually results in death [7]. This patient presented to us at the third stage.

Approximately 25% of patients with HSS have arterial and venous occlusions with vascular thromboembolism, arterial aneurysms, and nonspecific vasculitis. Seven percent of the reported cases have arterial, 25% have venous, and 68% have both. This patient had arterial aneurysm. The most common findings are pulmonary vascular lesions and DVT of the lower extremities. Aneurysms can be single or multiple, unilateral or bilateral. In this case aneurysms were bilateral. Aneurysms in other anatomical locations have also been identified in HSS. Peripheral vein thrombosis may affect the vena cava, jugular veins, iliac veins, femoral veins, hepatic veins, or cardiac cavities [8]. This patient was initially diagnosed with PTE, and there was no thrombus in the lower extremities on Doppler ultrasonography. After the development of pulmonary artery aneurysms, abdominal MRI showed thrombus in the inferior vena cava and in the right atrium with ECHO.

The etiology of this syndrome is still unknown. Some researchers argue that this syndrome is a type of Behçet disease characterized by multifocal vasculitis affecting small arteries



Figure 1. The chest X-ray, normal



Figure 2. The thoracic CT axial image, filling defects in the segmental branches of the right pulmonary artery

MAIN POINTS

- HSS is a very rare clinical disorder characterized by DVT and pulmonary and/or bronchial artery aneurysms.
- There is no standard treatment for HSS. The use of steroids alone or in combination with immunosuppressive drugs has been proposed.
- Anticoagulants are generally contraindicated due to an increased risk of fatal hemorrhage.

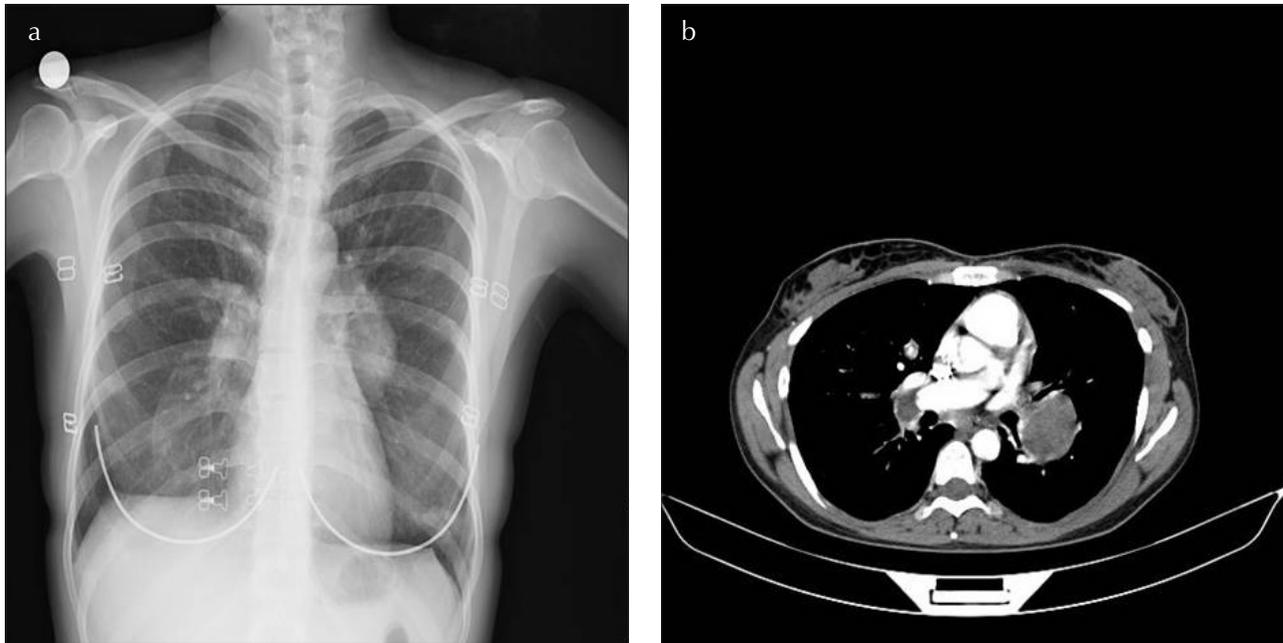


Figure 3. a, b. The chest X-ray showing bilateral hilar enlargement and the thoracic CT axial image showing thrombotic aneurysmatic dilatations in the left pulmonary artery

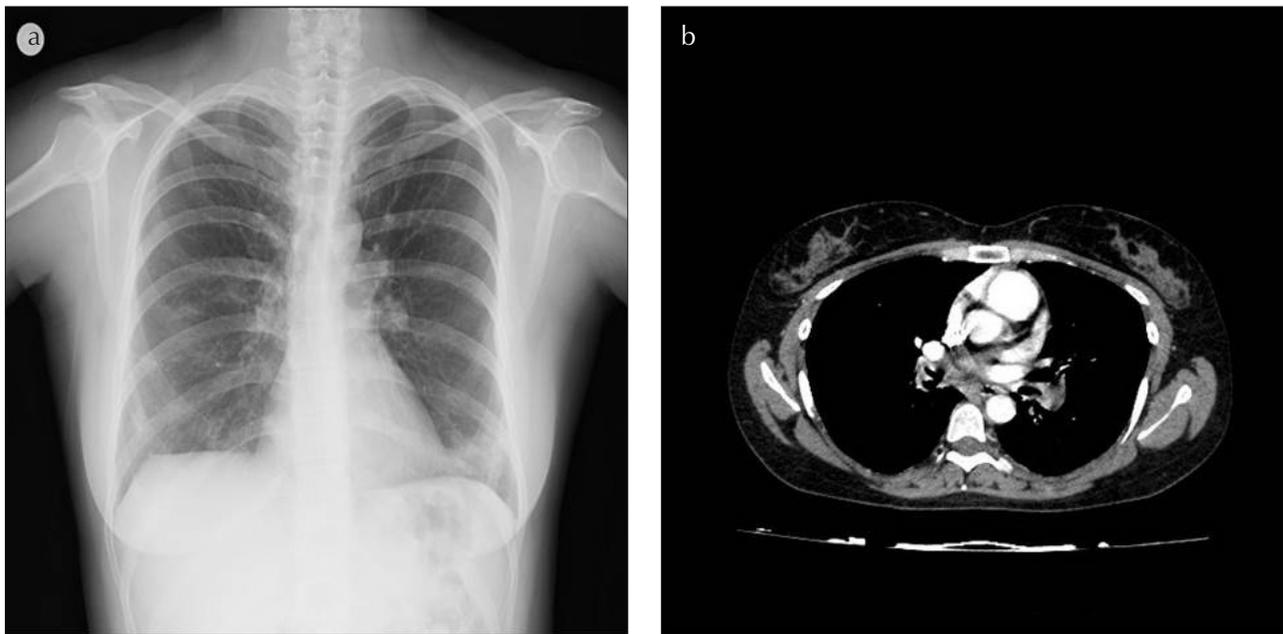


Figure 4. a, b. The chest X-ray showing bilateral hilar enlargement was significantly decreased after the treatment and the thoracic CT axial image showing thrombotic aneurysmatic dilatations of the pulmonary artery were reduced

and veins. Recurrent oral and genital ulcers and uveitis are common findings in Behçet disease. These findings have not been found in many cases of HSS [5]. Oral and genital ulceration or uveitis was not detected in this case, and Behçet disease was excluded by consulting rheumatology department. HSS, which is also considered as variant of Behçet disease, is mostly seen in men [2]. This case was a young woman, unlike other cases in the literature.

There is no standard treatment for HSS. Similar to the treatment of Behçet disease, the use of steroids alone or in combination with immunosuppressive drugs has been proposed. However, as the disease is usually fatal despite treatment, it is debatable whether this recommended treatment method

is effective [2]. In this case, we had to make a quick decision because of massive hemoptysis. We started pulse steroid treatment as soon as we noticed pulmonary artery aneurysm. Hemoptysis was controlled in a short time.

Anticoagulants are generally contraindicated due to an increased risk of fatal hemorrhage. However, it can be used cautiously where benefits are believed to outweigh the risks significantly. If there is pulmonary embolism, a therapeutic dilemma usually arises regarding the use of anticoagulants. Anticoagulation may prevent the progression of pulmonary embolism and resolve venous thrombosis [8]. In this case, LMWH was started because of pulmonary embolism. However, LMWH was discontinued when hemoptysis developed

and anticoagulant initiation was not considered. Thrombosis regressed with steroid treatment.

In patients with large, bilateral multifocal aneurysms and/or severe or recurrent hemoptysis, arterial embolization has been proposed as a treatment option. Surgical treatment (lobectomy or pneumonectomy) can be opted in complicated cases with massive hemoptysis due to large pulmonary aneurysm associated with high morbidity and mortality [8].

This is a rare case that was not considered as vasculitis since the aneurysms were not initially observed and it was thought to be vasculitis after the development of aneurysms. When pulmonary artery aneurysm is seen, it should be kept in mind that early treatment may be life-saving because it may be Behçet or its variant HSS. It is important to be aware of the fact that there may be an underlying vasculitis in recurrent embolism.

Informed Consent: Written informed consent was obtained from the patient who participated in this case.

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