



Patient Presenting with Hemoptysis: A Case of Hughes-Stovin Syndrome

Hemoptizi ile Başvuran Hasta, Hughes-Stovin Sendromlu Bir Olgu

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Abstract

Hughes-Stovin syndrome (HSS) is a very rare autoimmune clinical disorder that has been described as the presence of thrombophlebitis and multiple aneurysms in pulmonary and/or bronchial arteries. The pathogenesis is still unknown, but this syndrome is often thought of as a manifestation of Behçet disease. Herein, we describe a 59-year-old male patient who was admitted to massive hemoptysis. HSS was diagnosed on the basis of imaging pulmonary artery aneurysms and a history of lower extremity thrombosis. It differs in terms of the occurrence of this rare syndrome in an elderly patient. In this syndrome, which has a high mortality, the results are satisfactory when the treatment is started with a rapid diagnosis.

Keywords: Hughes-Stovin syndrome, deep vein thrombosis, hemoptysis, pulmonary artery aneurysm

Öz

Hughes-Stovin sendromu (HSS), pulmoner ve/veya bronşiyal arterlerde tromboflebit ve çoklu anevrizma varlığı olarak tanımlanan, çok nadir görülen bir otoimmün klinik bozukluktur. Patogenezi hala bilinmemektedir, ancak bu sendrom sıklıkla Behçet hastalığının bir varyantı olarak düşünülmektedir. Burada masif hemoptizi nedeniyle başvuran 59 yaşında bir erkek hastayı sunduk. HSS, görüntüleme pulmoner arter anevrizmaları görülmesi ve alt ekstremitte tromboz öyküsü temelinde konuldu. İleri yaş bir olguda bu nadir sendromun görülmesi açısından farklılık göstermektedir. Yüksek mortalitesi olan bu sendromda tedavisi hemen başladığında sonuçlar yüz güldürücü olmaktadır.

Anahtar kelimeler: Hughes-Stovin sendromu, derin ven trombozu, hemoptizi, pulmoner arter anevrizması

Introduction

Hughes-Stovin syndrome (HSS) is a very rare autoimmune clinical disorder which has been described as the presence of thrombophlebitis and multiple aneurysms in pulmonary and/or bronchial arteries, first described in 1959 by two British physicians (1).

The etiology and pathogenesis is still unknown, although vasculit is thought to be the underlying mechanism, various infectious agents are thought to be the cause for HSS (2). Although HSS is considered a variant of Behçet's disease (BD), it is thought to be a different entity. Patients with HSS usually present with symptoms such as cough, shortness of breath, fever, chest pain and hemoptysis (3,4). It is assumed to be a result of angiodysplasia and vasculitis, as in BD.

The disease affects predominantly young males between the second and fourth decades of life (5). We considered this case worth presenting in terms of his older age and a previous history of deep vein thrombosis (DVT).

Case Report

Herein, we described a 59-year-old male patient was admitted to the emergency department because of hemoptysis following 1 week of intermittent non-productive cough and sudden onset of dyspnea. He reported no chest pain. Hemoptysis was massive (about 300 mL) on admission. His medical history revealed that he was a ex-smoker (20 packs/year) and had oral aphthous ulcers occurring 3-4 times a year. There was no medical history of fever, weight loss, sweating, genital ulcer, uveitis and skin

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lesions or tuberculosis. He gave a history of DVT in left leg in 2019. At that time, a chest tomography was taken and he was called to the controls for further research, but he did not come. He has been receiving warfarin 5 mg/day ever since.

On physical examination, his blood pressure was 140/85 mmHg, body temperature was 37 °C, respiratory rate 22/minute. Musculoskeletal examination revealed normal range of motion and no arthritis was found. There was an increase in diameter in the left leg. He had no rash lesion on extremities. Respiratory sounds were decreased in the bilateral lower zones of the lung. Laboratory investigations showed; sedimentation first hour 25 mm/h, C-reactive protein of 1.6 mg/dL, hemoglobin 13.3 gm/dL, white blood cell count of 9,400/ μ L. Platelet count was 192,000/ μ L. Renal and liver function tests were within normal limits. Normal D-dimer levels ($n < 243$) was found.

The chest computed tomography (CCT) scan was taken due to abnormality on lung X-ray. CCT revealed an aneurysmal enlargement in the right lower lobe pulmonary artery with thrombus inside. It was reported that the right lower lobe ground glass opacity probably representing a pulmonary hemorrhage (Figure 1a, b). When the patient's previous examinations were examined, a CCT was seen in 2019. Partially thrombosed saccular aneurysmatic enlargements were observed

in the subsegment branches of the right lung lower lobe pulmonary artery, and clinical laboratory correlation in terms of the differential diagnosis of pulmonary artery aneurysm was reported as appropriate in terms of Behçet and other vasculitic involvement (Figure 2).

Finally, a diagnosis of HSS was made on the basis of pulmonary artery aneurysms and thromboses. Warfarin was discontinued due to bleeding. The patient started with pulse methylprednisolone therapy (1000 mg/day) intravenous bolus infusion and then cyclophosphamide 750mg monthly as intravenous pulses. He was treated with intravenous therapy followed by oral steroid with subsequent taper (1 mg/kg) and 6 cyclophosphamide pulses of 1 gram each per 6 months with incomplete regression of aneurysms and thromboses.

Written informed consent was obtained for publication of the case report and accompanying images.

Discussion

HSS is very rare but associated with significant morbidity and mortality. The aetiology of this condition is unclear and has been defined in different ways in the literature. Some of those; "incomplete BD" and "a rare case of BD" (6,7).

Although the exact etiology and pathogenesis of HSS are unknown, it has been suggested that vasculitis may be an underlying mechanism. In this respect, it should be considered in terms of differential diagnosis with BD, which is also known as "Silk Road" disease and is common in our country. Although it seems to have similar aspects to BD in terms of pulmonary involvement and DVT, it differs in the absence of systemic involvement and the absence of orogenital ulcers (8).

While thromboembolism is seen in 25% of patients with HSS, massive hemoptysis secondary to pulmonary artery aneurysm rupture is also severely mortal. It progresses with different clinical findings from thrombophlebitis to aneurysm. Major vascular involvements of the syndrome are as follows: arterial (7%), venous (25%) or both (68%) in the studies (9,10).

Diagnosis is usually based on the clinical and radiological presentation of venous thrombosis with concomitant pulmonary

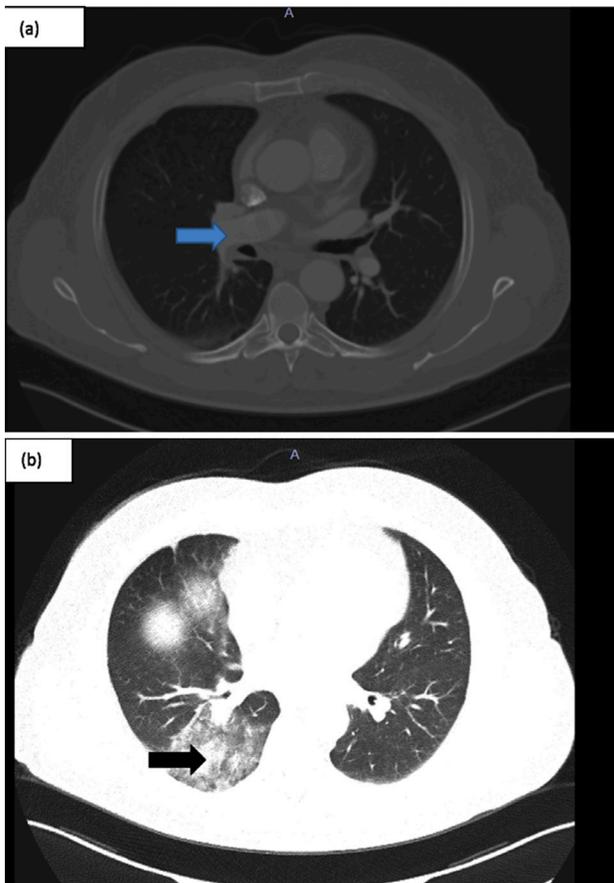


Figure 1. (a) Thoracic computed tomography (CT) showing an aneurysmal enlargement in the right lower lobe pulmonary artery with thrombus inside (blue arrow) (b) ground glass opacity representing a pulmonary hemorrhage (black arrow)

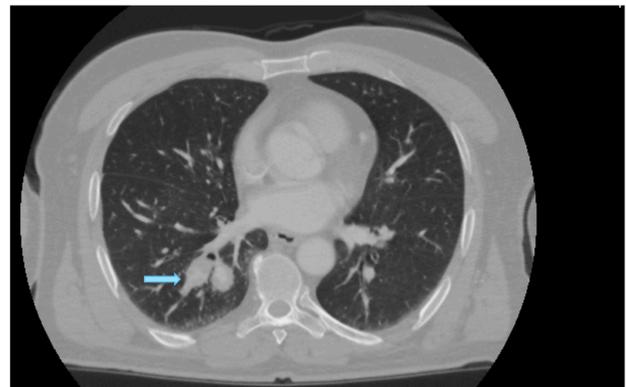


Figure 2. The blue arrow points to partially thrombosed saccular aneurysmatic enlargements were observed in the subsegment branches of the right lung lower lobe pulmonary artery

artery aneurysm in a patient. CT scan or magnetic resonance angiography is the diagnostic method of choice to detect pulmonary artery aneurysms or other visceral artery aneurysms (9,11).

In medical treatment, steroids and cytotoxic agents are used. In particular, the combination of glucocorticoids and cyclophosphamide was used as the most commonly preferred therapeutic agents in this context. Rapid recognition of this syndrome is important, especially since the presence of pulmonary artery aneurysm has a poor prognosis. If appropriate treatment is initiated promptly and early in the course of the disease, remission can be achieved quickly (12,13). In some cases, surgery may be required, except in emergencies, surgery is considered after the disease has stabilized (14).

Our case did not have any new complaints in the follow-up after completing his treatment with 6 cyclophosphamide pulses. In this case, we wanted to present the presence of aneurysm in the chest tomography taken at the time of admission with DVT and the presence of HSS at a older age.

Ethics

Informed Consent: Written informed consent was obtained for publication of the case report and accompanying images.

Peer-review: Internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: K.A.A., Ö.A., Concept: K.A.A., Ö.A., Design: K.A.A., Ö.A., Data Collection or Processing: K.A.A., Analysis or Interpretation: K.A.A., Ö.A., Literature Search: K.A.A., Writing: K.A.A.

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