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Case Report

Hughes-Stovin syndrome: a case report and literature review

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ABSTRACT

Hughes-Stovin syndrome (HSS) is a rare clinical entity comprised of thrombophlebitis and multiple pulmonary and bronchial aneurysms. Medical literature has documented fewer than 40 cases of HSS. Its pathogenesis and etiology are still unknown. It is considered a variant of Behcet's disease (BD). HSS is being managed either medically or surgically. We report the case of a 32-year-old Saudi male patient with HSS who initially presented with a pulmonary embolism (PE). Later, he developed hemoptysis that was treated successfully with methylprednisolone. The genetic, etiologic, and pathologic basis of HSS should be explored more in the future to elucidate the disease. Exploration of better and optimal medical and surgical therapeutic regimens is a need for time to treat and prevent morbidity in BD and HSS. There is a dilemma of anticoagulation in these patients that needs to be addressed and investigated properly.

Keywords: Hemoptysis, Pulmonary artery aneurysms, Huges-Stovin syndrome

INTRODUCTION

Hughes-Stovin syndrome (HSS) was first documented by Hughes J. P. in 1959.[1] It is a rare clinical disorder presented by multiple pulmonary/bronchial artery aneurysms and peripheral venous thrombosis. Until now, the medical literature has documented only about 40 cases of HSS. HSS's exact etiology and pathogenesis are still undefined and under research. Researchers think HSS may be a variant of BD. HSS predominantly affects men aged 12-40 years. This disease may present with cough, dyspnea, hemoptysis, chest pain, and clinical signs of pulmonary hypertension.[2] Other observed associated features include fever and signs of intracranial hypertension. Aneurysms can occur at any site in systematic circulation, but most commonly, pulmonary, and bronchial arteries aneurysms present as hemoptysis.[3] Recurrent peripheral venous phlebitis commonly results in thrombus formation that sometimes leads to vena cava and right atrium thrombosis.[2,4] The pathogenesis of HSS is not fully established. Tissue histopathology observed the destruction of the arterial walls and perivascular lymphomonocytic infiltration of capillaries and venules. [5]

The current consensus is that HSS results from a vasculitis phenomenon like that seen in BD.[2] BD pathognomonic feature is vasculitis which leads to systemic involvement of arterial occlusion, arterial aneurysm, venous occlusion, and varices.[6] Furthermore, it is supposed that HSS represents a cardiovascular manifestation of the same phenomenon as BD.[7] However, other clinical features can overlap significantly, although histopathological features

of the aneurysms in both entities are almost similar.[8] Different imaging modalities like conventional angiography, contrast-enhanced MRA contrast-enhanced MDCTA, and multi-detector row helical CT angiography are options for diagnosing an aneurysm.[9,10] The most common cause of death in HSS is hemoptysis due to pulmonary/bronchial arterial aneurysm rupture.[11] However, systemic bronchial artery hypertrophy secondary to ischemia could also be the origin of bleeding.[3,12] Generally, patients with HSS have a poor prognosis, and aneurysmal rupture is the leading cause of death. There is consensus that early diagnosis and optimal management for HSS are imperative to standardizing the quality of care to improve prognosis.

CASE REPORT

A 32-year-old Saudi male patient presented to our hospital for the first time complaining of a dry cough for one month, followed by hemoptysis for two days, associated with shortness of breath. He also gives a history of left leg swelling 6 months ago and an episode of transient loss of vision in the left eye 1.5 years ago, diagnosed as an ischemic stroke and started on aspirin and clopidogrel. His vital signs were normal. The clinical examination was unremarkable. Doppler ultrasonography of the left lower limb documented sub-acute deep venous thrombosis (DVT), Computed tomography-pulmonary angiography (CTPA) showed findings of bilateral PE with small aneurysmal dilatation as a sequel of vasculitis (Figure 1 and 2).

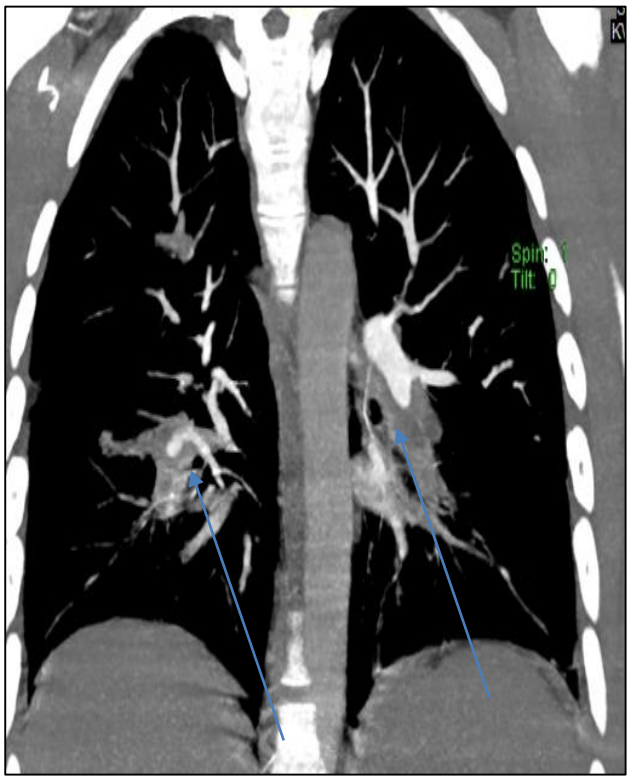

Figure 1: CT angiogram of the chest showing bilateral PE.

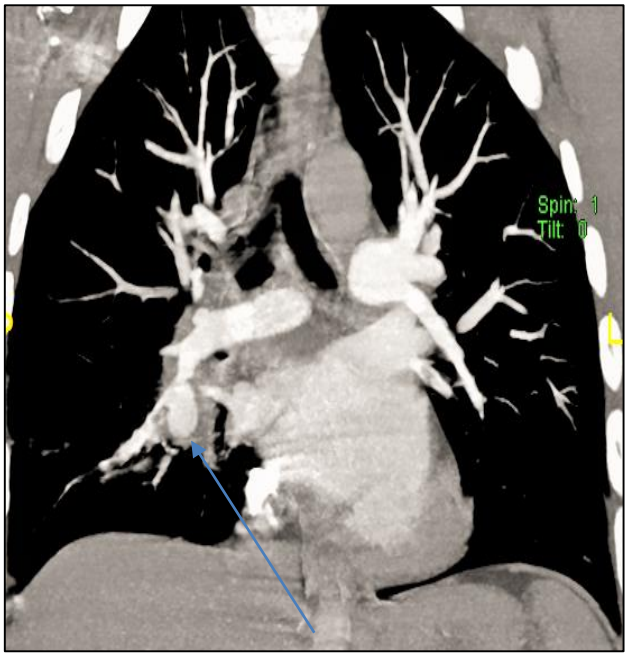

Figure 2: CT angiogram of the chest showing right sided pulmonary artery aneurysm.

He received an intravenous one-gram dose of methylprednisolone and refused admission and discharge against medical advice. Ten months later, he came again to our hospital with massive hemostasis. He gave a history of admission to other hospitals 4 months back. At that time, he was presented with hemoptysis and DVT. He had an angiography report with coils placed for his aneurysm (Figure 3).

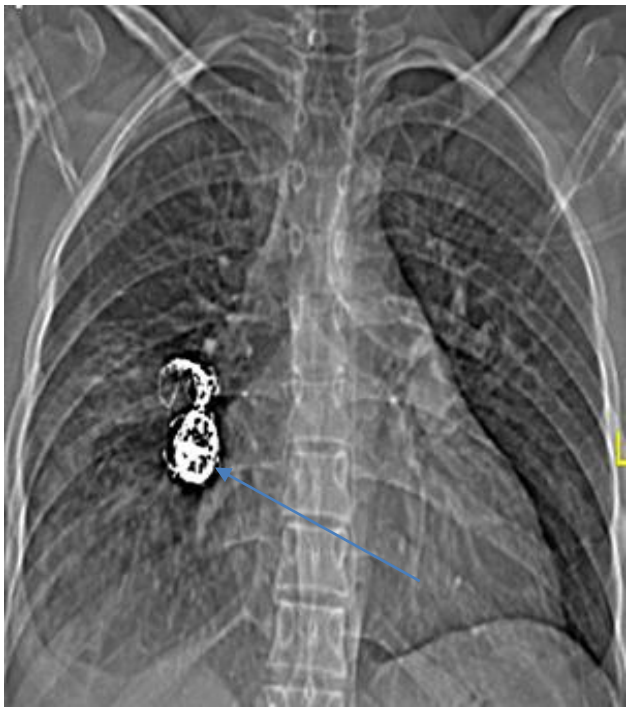

Figure 3: CT scan of the chest shows right-sided pulmonary artery aneurysm coiling.

Clinically, a diagnosis of HSS was made in a young patient, without clinical findings consistent with BD, and pulmonary artery aneurysms with PE and DVT were strong evidence of HSS. He was admitted to the intensive care unit because of unstable hemodynamic signs. He received methylprednisolone intravenous 1gram for five days followed by oral prednisolone 70 mg daily. He also received 2 units of packed red blood cells and one dose of cyclophosphamide 1 gram with good control of the hemoptysis. Then the patient was shifted to a higher center for possible intervention.

DISCUSSION

HSS is more common in men between the ages of 12 and 40. Hughes proposed that alterations in the vasa vasorum of arteries caused by degenerative bronchial arteries contribute to the formation of arterial aneurysms.[1] HSS shares etiology and clinical features with BD. Digital subtraction angiography data that support Hughes' hypothesis are documented by Mahlo et al and Herb et al document digital subtraction angiography findings that correspond to Hughes' theory.[3,13] The leading cause of death in BD is hemoptysis. Similarly, HSS has a bad prognosis because of massive hemoptysis. Finally, hemoptysis may be caused by aneurysmal rupture or pulmonary artery occlusion.[3,12] Helical CT angiography better evaluates pulmonary aneurysms and bronchial/non-bronchial systemic arteries than conventional angiography.[12] The HSS syndrome is most often treated to stabilize the patient with immunosuppressants, either systemic corticosteroids or cytotoxic drugs.[14] A therapeutic dilemma is usually seen in the use of anticoagulants in the presence of PE in HSS. Anticoagulation has the risk of increasing the severity of hemoptysis. Surgical intervention by resecting the affected segments of the lung is to be considered in cases of high-risk ruptured aneurysms.[7] However, surgery related to high morbidity and mortality is the other option for such cases. The trans-catheter approach to embolization and even embolectomy in critical patients is an alternative, safe option.[2]

CONCLUSION

Early diagnosis and optimal timely management of HSS is a fundamental and crucial step to decreasing the disease's morbidity and mortality. Genetic testing is needed to see its familial preponderance. Finally, optimizing the therapeutic guidelines is warranted to manage the disease with its serious consequences.

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