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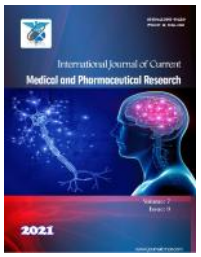

HUGHES-STOVIN SYNDROME; A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

Background: Hughes-Stovin Syndrome (HSS) is a rare clinical entity comprised of thrombophlebitis, multiple pulmonary and/or bronchial aneurysms. Medical literature has documented fewer than 40 cases of HSS. Its pathogenesis and etiology are still unknown. It is considered as a variant of Behcet's disease (BD). HSS is being managed either medically or surgically.
Case Presentation: We report the case of a 32-year-old Saudi male patient with Hughes-Stovin syndrome who initially presented with a pulmonary embolism. 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 clearly elucidate the disease.
Conclusion: 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.

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 exact etiology and pathogenesis is still undefined and under research. Researchers think HSS may be a variant of Behcet's disease (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 caval and right atrium thrombosis (2,4). The pathogenesis of Hughes-Stovin syndrome 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 Hughes-Stovin syndrome results from a vasculitis phenomenon like that seen in Behcet's disease (2). Behcet's disease pathognomonic feature is vasculitis that leads to systemic involvement of arterial occlusion, arterial aneurysm, venous occlusion, and varices (6). Furthermore, it is supposed that Hughes-Stovin syndrome represents a cardiovascular manifestation of the same phenomenon as Behcet's disease (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 and contrast-enhanced MDCTA, and multi-detector row helical CT angiography are options to diagnose 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

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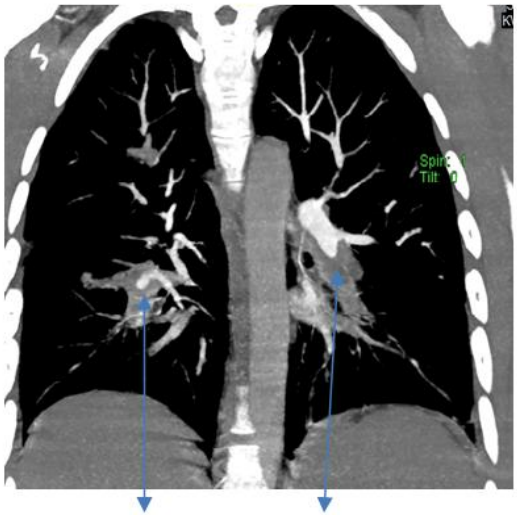
with HSS have a poor prognosis, and aneurysmal rupture is the leading cause of death. There is consensus that early diagnosis and optimal management for HSS is imperious to standardize the quality of care to improve prognosis.

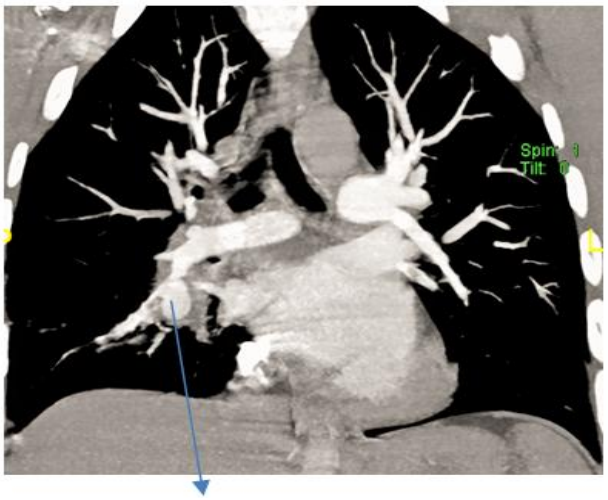
Case Report

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 pulmonary embolism (PE) with small aneurysmal dilatation as a sequel of vasculitis (see below figure1&2). 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 (see figure 3). Clinically, a diagnosis of Hughes-Stovin syndrome was made in a young patient, without clinical findings consistent with Behcet's disease 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, and then patient was shifted to higher center for possible intervention.



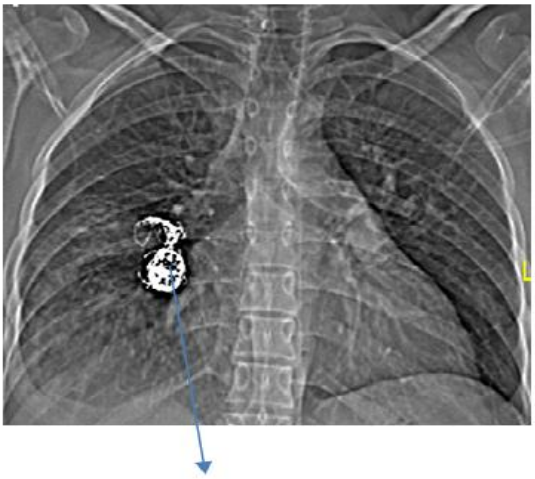
Bilateral pulmonary embolism

Figure 1 CT Angiogram of the chest showing Bilateral pulmonary Embolism (original image).



Rt. pulmonary artery aneurysm.

Figure 2 CT Angiogram of the chest showing Right sided Pulmonary artery aneurysm (Original Image)



Pulmonary artery coiling

Figure 3 CTscan of chest shows Right sided pulmonary artery aneurysm coiling (Original image).

DISCUSSION

Hughes-Stovin syndrome 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). Hughes-Stovin syndrome shares etiology and clinical features with Behcet's disease. Digital subtraction angiography data that support Hughes' hypothesis are documented by Mahlo *et al.* (13) and Herb *et al.* (3)document digital subtraction angiography findings that correspond to Hughes' theory. The leading cause of death in Behcet's disease is hemoptysis. Similarly, Hughes-Stovin syndrome has bad prognosis because of massive hemoptysis. Finally, hemoptysis may be caused by aneurysmal rupture or pulmonary artery occlusion (3, 12).Helical CT angiography provides better evaluation of pulmonary aneurysms, and bronchial/non-bronchial systemic arteries than does conventional angiography (12).The HSS syndrome is most often treated to stabilize the patient with immunosuppressants, either systemic corticosteroids or cytotoxic drugs (14). A therapeuticdilemma is often 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 rupture 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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Ethics approval: It was not required for the imaging, diagnosis, and study of the case, in our institution.

Consent: Informed consent was obtained from the patient. Conflicts of interest: None

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