HUGHES-STOVIN SYNDROME: AN INCOMPLETE BEHCET'S DISEASE

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ABSTRACT

Hughes Stovin syndrome is very uncommon condition characterized by deep vein thrombosis (DVT) and pulmonary artery aneurysm. The etiology is still not known but it is considered to be manifestation of systemic vasculitis. It shares many clinical signs and symptoms with Behcets disease and that is why it is also called incomplete Behcets disease or variant of it. Here we report a male patient case who presented to our tertiary care hospital with DVT and pulmonary artery aneurysm. CT angiography showed aneurysmal dilatations of segmental and sub segmental pulmonary arteries. He was successfully treated with steroids and immunosuppressant.
Key Words: Aneurysm; Behcets syndrome; Pulmonary artery; Venous thrombosis.

Introduction

Hughes Stovin Syndrome (HSS) is an uncommon clinical condition which comprises of aneurysm of the pulmonary artery with thrombosis of deep veins.[1] The etiology of this disorder is still not clear.[2] It is considered variant of Behcet's disease (BD) as both sharesome of the important clinical entities.[3] HSS may present with life threatening hemoptysis and that is why early diagnosis and prompt treatment is essential.4

Case Report

A 40 years old gentleman presented to our hospital with main complaints of left leg swelling. His vitals were in normal range and the clinical examination was unremarkable except left leg edema. A color Doppler examination was performed which was suggestive of thrombosis of the left femoral and popliteal veins. He was started on low molecular weight heparin. But after cessation of anticoagulants, he developed shortness of breath and hemoptysis. Single slice computed tomography of chest was performed which showed findings suggestive of bilateral aneurysms of pulmonary arteries. He was then advised CT pulmonary angiography (CTPA) which confirmed the findings of pulmonary artery aneurysms as shown in (Fig. 1 and 2).

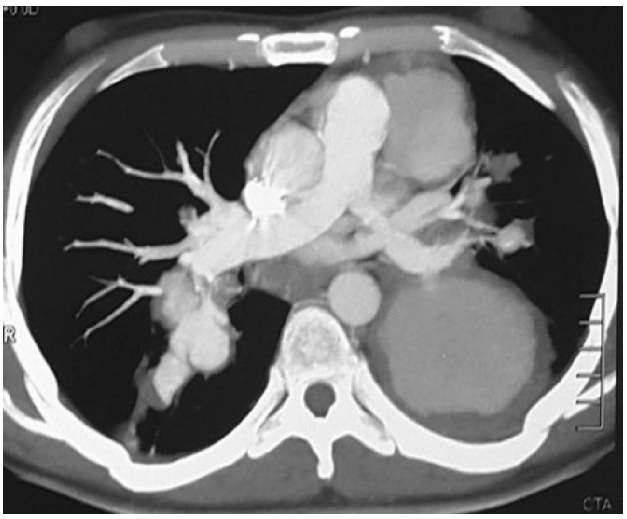

Figure 1: Axial section of CTPA showing multiple segmental pulmonary artery aneurysms with thrombi.

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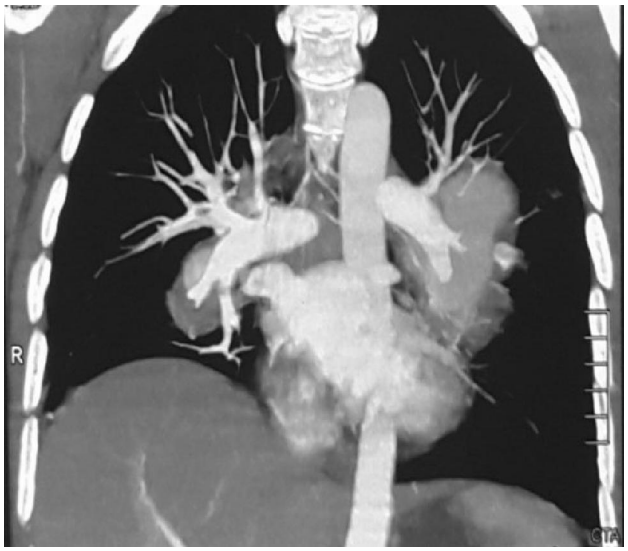

Figure 2: Coronal section of CTPA showing multiple segmental pulmonary artery aneurysms with thrombi.

Patient was investigated extensively to find the cause. Hematological profile showed decreased hemoglobin which was explained by hemoptysis, otherwise TLC, platelet counts, liver and renal function tests were normal. ESR was raised (60mm/1st hour). Antinuclear antibody (ANA) was weakly positive with negative anti double stranded DNA (anti dsDNA) and antineutrophil cytoplasmic antibody (ANCA). D-dimers and fibrinogen degradation products (FDPs) were above the limit of normal range. The patient didn't have any history of uveitis, skin lesions, oral and genital ulcers. On the basis of these findings against BD; it was ruled out and HSS was considered. The patient was started on intravenous methylprednisolone and cyclophosphamide which resolved his hemoptysis. He was not put on anticoagulants owing to the risk of massive hemoptysis secondary to rupture of pulmonary aneurysms.

Discussion

In 1959 HSS was described by two British doctors, John Patterson Hughes and Peter George Ingle Stovin. HSS was named after them, who described the findings of pulmonary artery aneurysms with deep vein thrombosis in four patients presented to them.[5] It has male predominance aged ranging from 12 to 40 years.[6] Hemoptysis can present any time during course of disease.

The etiology of this syndrome is unknown but some scientists describe it as a manifestation of idiopathic systemic vasculitis. Many physicians consider it as an incomplete form of BD.[7] Vasculitis in BD causes arterial aneurysm, venous and arterial thrombosis. It may not be wrong to call HSS as cardiovascular manifestation of BD, in fact in both these conditions; pulmonary aneurysms are main clinical findings. BD and HSS have the same course of pathophysiology and in both entities, death may occur after massive hemoptysis or due to aneurysmal rupture. Pulmonary artery thrombi may occur alone without involvement of the deep vein as a result of pulmonary artery inflammation. Hughes and Stovin theory suggested that aneurysms develop secondary to degenerative changes in the arteries of the bronchioles leading to weakness of the walls.[5] These findings were validated in other studies when they performed digital subtraction angiography on the patients.[8,9]

Conventional angiography is a good tool of evaluation in HSS as it not only describes pulmonary arteries but at the same time angiodysplastic bronchial arteries can be assessed. However, multidetector helical CT angiography is superior to the conventional angiography by giving explicit and detailed information about the bronchial arteries.[10]

Unfortunately, no guidelines are present about treatment of HSS but it is usually treated with steroids and immunosuppressants.[6] Anticoagulants are avoided as there is increased risk of massive hemoptysis which may put the life of the patient at risk.[2] Surgical resection of the lung can be an option if the aneurysm is at peak of being ruptured or the aneurysm is localized to one part of the segment.[4,8] Embolization can be another option for aneurysms which are at high risk of rupturing.[9]

Conclusion

HSS is a very rare clinical syndrome and it is considered variant of BD which may present with deep vein thrombosis and pulmonary artery aneurysm leading to massive and life threatening hemoptysis. Therefore, timely diagnosis and management is essential. Steroids and immunosuppressants are the key drugs in the management of this disease as our patient's symptoms improved with them.

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