

CASE REPORT

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Hugh-Stovin syndrome: the ‘incomplete Behcet’s disease’. A case study of a young adult with recurrent pulmonary embolism and pulmonary arterial aneurysms

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ABSTRACT

Hugh-Stovin Syndrome (HSS) is characterized by recurrent thrombophlebitis and multiple pulmonary and/or bronchial artery aneurysms indistinguishable from the cardiovascular features seen in Behcet’s disease (BD). Our case describes a 30-year-old male with recurrent pulmonary embolism and bilateral pulmonary aneurysms. Autoimmune, hypercoagulable, and infectious work up were negative. Elevated inflammatory markers and absence of the typical clinical findings seen in BD led to the diagnosis of Hugh-Stovin syndrome (HSS). Immunosuppression using steroids and azathioprine led to clinical response. Anticoagulation was continued based on risk/benefit ratio.

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1. Introduction

Hugh-Stovin syndrome (HSS) is characterized by recurrent thrombophlebitis and multiple pulmonary and/or bronchial aneurysms first described in 1959 by two British physicians, Drs. John Hughes and Peter Stovin in a case report of four male patients with deep venous thrombosis (DVT) and segmental pulmonary artery aneurysms (PAA) [1–5]. Less than 40 cases have been described in medical literature [6]. Young adult males between ages 12 to 48 years are predominantly affected with cases reported in diverse geographical areas including North America, Europe, Africa, and Asia [2–4]. The genetic bases and familial predisposition of HSS remain unclear [3].

Recurrent thrombophlebitis usually involves large vessels leading to thrombosis of the vena cava, heart chambers, jugular, iliac and femoral veins, and dural sinuses [3,4]. Aneurysms observed in HSS may be single, multiple, unilateral, or bilateral and generally involves the pulmonary and bronchial arteries but can occur anywhere in the systemic circulation [3,4,6].

Patients with HSS can present with cough, hemoptysis, dyspnea, and recurrent episodes of pulmonary embolism (PE) despite effective anticoagulant therapy [2]. We describe a case of a young African American man presenting with recurrent episodes of unprovoked PE complicated by PAA.

2. Case presentation

A 30-year-old African American male with a remote history of smoking, recent unprovoked bilateral PE and pneumonia presented with 2-week history of

cough, chest pain, and dyspnea. Seven months prior, patient was diagnosed with pneumonia. One month later, he was diagnosed with bilateral unprovoked PE without DVT and initiated on coumadin which unfortunately he stopped after 1 month. Of note, there is no family history of DVT or PE or recent prolonged immobilization.

On presentation, he was afebrile with stable hemodynamics without evidence of hypoxia. Body mass index was 23.7 kg/m². Physical examination was unremarkable and without cutaneous or mucosal ulcerations.

Complete blood count and comprehensive metabolic panel were normal except for mild anemia (hemoglobin 11.4 g/d, normal range: 14–18 g/dl). Both erythrocyte sedimentation rate and C-reactive protein were elevated 125 mm/hour (normal 0.0–15.0 mm/hour) and 9.59 mg/l (normal 0.0–4.9) respectively. IgM cardiolipins were slightly elevated but other hypercoagulable parameters were normal. Antinuclear antibodies, antineutrophil cytoplasmic antibodies, rheumatoid factor, complement, and HLA-B51 were normal. Human immunodeficiency virus 1 and 2, rapid plasma regain, hepatitis viral studies and interferon gamma release assay were also normal.

Computed Tomography (CT) of the chest showed enlarged pulmonary arteries, bilateral pulmonary artery occlusions involving both upper and lower lobes containing organizing thrombus, multiple bilateral PAAs, peripheral right pulmonary infarcts with mediastinal vascular collaterals and right ventricular thrombus. Transthoracic echocardiogram showed a left ventricular ejection fraction of 60%, dilated right ventricle with normal wall thickness, and a 5 mm thrombus. Venous doppler of the lower extremity was negative for DVT.

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Anticoagulation with heparin infusion was initiated. Thrombolysis was not performed given the chronicity of the thrombus.

3. Discussion

PAAAs have been associated with congenital heart disease, trauma, infection, lung cancer, pulmonary hypertension, connective tissue disorders, and systemic inflammatory diseases [3,5–9]. Marfan and Ehlers-Danlos syndromes can be associated with aneurysmal vascular dilatations although specific diagnostic criteria and disease classification must be established, i.e., the revised Ghent Nosology criteria and the revised Villefranche Classification, respectively [10,11]. Our patient fulfilled neither criteria.

BD, HSS, and Takayasu's arteritis are vasculitides associated with PAAAs [6]. HSS has been described as 'incomplete Behcet's' or categorized as 'cardiovascular manifestation of BD' [2–4]. Although they share similar cardiovascular manifestations, HSS lacks the typical clinical manifestations of Behcet's disease such as arthralgia, recurrent orogenital ulcers, pathergy, and eye lesions [2,3,6]. Clinical manifestations of HSS are triphasic. First, findings of thrombophlebitis are seen; second, formation of large pulmonary and/or bronchial aneurysms are characteristic; finally, if left untreated, aneurysmal rupture, massive hemoptysis, and death can ensue [3,4,6].

Given shared similarities, medical management of HSS resembles BD [6]. Aggressive treatment of the PAAAs is necessary because of its significant morbidity and mortality [6,12]. Potential treatment options for pulmonary aneurysms are: immediate surgical intervention, embolization, or observation with management of underlying disease [3,12]. Surgical lobectomy or pneumonectomy are options for massive hemoptysis secondary to large pulmonary aneurysms, expanding aneurysms or lesions confined to one segment or one lung [13]. Primary surgical repair of proximal main pulmonary artery aneurysms may be feasible [6]. Transcatheter PAA embolization is a less invasive option [3,13]. In our case, the role of surgery is limited given extensive bilateral PAAAs, which confers high operative morbidity and mortality as well as recurrence in 25% of cases [3].

As in BD, immunosuppression remains the cornerstone of treatment of HSS. The European League Rheumatism (EULAR) recommends corticosteroids, azathioprine, cyclophosphamide, or cyclosporine A [3,14]. Anti-TNF inhibitors (infliximab) may be used [6]. Our patient responded well to treatment with prednisone and azathioprine. The role of anticoagulation in the management of BD-associated thrombotic disease is unclear [4,6]. Given elevated clot burden and recurrence, anticoagulation was initiated using apixaban.

4. Conclusion

Early diagnosis and treatment are important and lead to improved outcomes in HSS. Immunosuppression as used in BD remains the best medical treatment option. The role of anticoagulation remains unclear.

Disclosure statement

No potential conflict of interest was reported by the authors.

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