Case Report

Hughes–Stovin syndrome: a case report

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Abstract

Hughes–Stovin syndrome (HSS) is a very rare clinical entity, which is characterized by peripheral venous thrombosis and multiple pulmonary and/or bronchial artery aneurysms and is associated with a high mortality. Although the exact etiology of HSS is still not known, possible causes include infection and angiodysplasia. Because of its histopathological and clinical similarities with Behçet's disease (BD), it is also considered as a variant of BD. HSS usually present with cough, dyspnea, fever, chest pain, and hemoptysis, which can be massive and often fatal. We report a case of a 24-year-old man who presented with thrombosis of inferior vena cava (IVC) with recurrent hemoptysis. There were evidences of pulmonary artery aneurysm and thrombosis of IVC and lower extremity veins on contrast-enhanced computed tomography (CECT) scan of the thorax and color Doppler study, respectively. There were no orogenital ulcerations or uveitis consistent with BD. Pulmonary transcatheter embolization was planned but the patient died following massive hemoptysis probably because of rupture of the aneurysm.

KEY WORDS: Hughes–Stovin syndrome, pulmonary artery aneurysm, hemoptysis

Introduction

Hughes–Stovin syndrome (HSS) is an extremely rare condition, characterized by multiple pulmonary artery aneurysms and peripheral venous thrombosis, which was first described in 1959 by John Patterson Hughes and Peter George Ingle Stovin.[1] The disease usually affects young men in their second to third decade of life. Patients usually present with recurrent fever, chills, chest pain, dyspnea, cough, and hemoptysis. The natural course of the illness is usually fatal because of massive hemoptysis secondary to the rupture of a pulmonary artery aneurysm. The etiology of HSS is still unknown; infection and angiodysplasia have been proposed as possible causative factors of the disease, although these are not widely accepted. It is also thought to be a clinical variant of Behçet's disease (BD), because of the combination of pulmonary artery aneurysm and venous thrombosis, found in both the diseases.

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A 24-year-old man was admitted for the first time in our institute with complaints of repeated episodes of hemoptysis and low-grade fever, off and on, since 2005. On admission, the patient had cough with mild expectoration of blood-tinged sputum and a low-grade fever associated with chills. In 2011, he had features suggestive of deep venous thrombosis of bilateral lower limbs, more on the left, with mild hemoptysis, which resolved with oral corticosteroids. In 2012, he was put on course of warfarin in view of thrombosis of inferior vena cava (IVC) and pulmonary artery.

On examination, except for a mild temperature, other vital signs such as pulse, blood pressure, and respiratory rate were at normal levels. There were engorged and tortuous veins in the upper part of abdomen and lower chest wall along with dark pigmentations in both the lower limbs below the knees. There was no evidence of uveitis and oral or genital ulcerations, suggestive of BD. Hematological tests showed normal leukocyte and platelet counts, normal levels of erythrocyte sedimentation rate (ESR), and normal coagulation profile. pANCA and cANCA tests were negative. Color Doppler study was suggestive of venous thrombosis extending bilaterally from the tibial, popliteal, and iliac veins up to lower part of IVC with multiple collaterals and dilated saphenous vein. There was sluggish blood flow in the popliteal, posterior tibial, and dorsalis pedis arteries bilaterally.

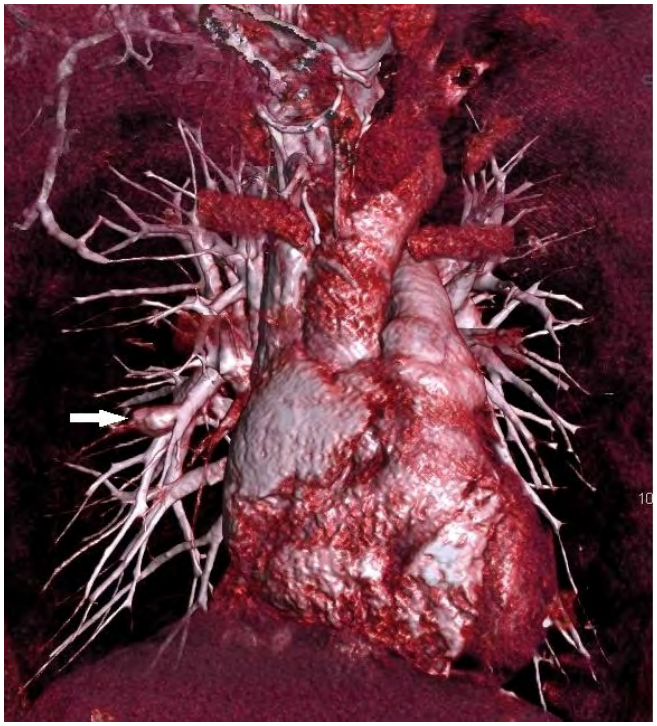

Figure 1: Volume-rendered reconstruction of CT angiography shows the descending right pulmonary artery basal segmental branch with an aneurysm (white arrow).

Contrast-enhanced computed tomography (CECT) of thorax showed fusiform type of aneurysmal dilatation of pulmonary segmental artery supplying the lateral segment of right middle lobe with secondary compression of adjacent segmental bronchiole, resulting in distal mucosal plugging and subsegmental consolidation (suggesting secondary infection). There was nonvisualization of infrarenal IVC with multiple retroperitoneal and parietal wall collaterals suggestive of chronic deep vein thrombosis [Figures 1 and 2].

A diagnosis of HSS was made on the basis of pulmonary artery aneurysms and IVC thrombosis in a young male patient, without clinical findings consistent with BD. Patient was managed conservatively with intravenous antibiotics and methylprednisolone. Pulmonary transcatheter embolization was planned, but before the procedure could be done, the patient died of a massive hemoptysis, possibly because of the rupture of the pulmonary aneurysm.

Discussion

HSS is a very rare clinical disorder characterized by multiple pulmonary and/or bronchial artery aneurysms and peripheral venous thrombosis. The disease was named after John Patterson Hughes and Peter George Ingle Stovin, two British physicians, who first described this syndrome in 1959.[1] The disease is extremely rare, and very few cases

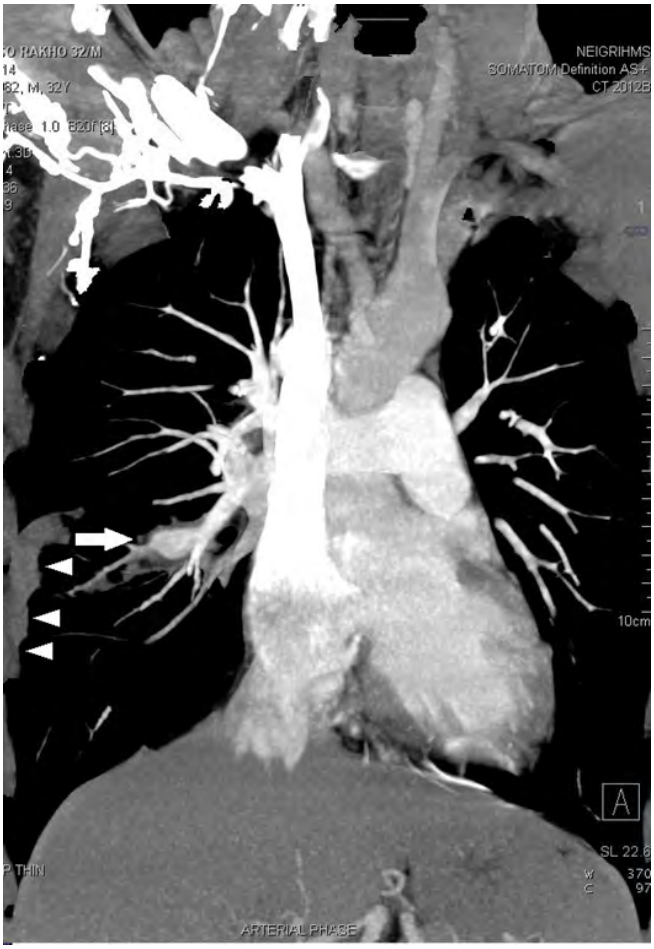

Figure 2: Coronal multiplanar reformat (MPR) image shows the aneurysm (arrow) with associated consolidation (white arrowheads).

have been reported in the literature. There is no definite clinical or laboratory test diagnostic for this syndrome. HSS has been classically described in young men with peripheral systemic venous thrombosis and multiple pulmonary and/or bronchial artery aneurysms, which is also found in BD.

HSS usually affects the young adult population, mostly male patients aged 12–48 years. The clinical feature of HSS grouped into three components: (a) features of thrombophlebitis; (b) features owing to pulmonary and/or bronchial artery aneurysms; and (c) features owing to aneurysmal rupture leading to massive hemoptysis and death. The typical presentations of HSS are cough, dyspnea, hemoptysis, venous thrombosis, chest pain, fever, chills, pulmonary hypertension, and intracranial hypertension. Patient can have seizure, diplopia, and cephalgia secondary to raised intracranial pressure as a result of cerebral venous sinus thrombosis.

Although the exact etiology and pathogenesis of HSS is not yet known, various theories have been proposed.

Infection has been thought to one possible mechanism in the genesis of HSS and in septic embolisms, and abscesses have been proposed as the cause of pulmonary aneurysm as there are many reports of aneurysm preceded by infection such as scrotal abscess and epididymitis.[2] However, the infectious etiology is not sustainable for two reasons: first, because of failure of various antibiotic regimens in the treatment of HSS and, second, there has been lack of positive blood culture in HSS. Angiodysplasia of bronchial arteries has also been proposed as a causative factor for the genesis of the vascular changes in HSS.[3] Hughes and Stovin hypothesized that the structural changes in the bronchial artery causes inadequate nutrition to the pulmonary artery via the vasa vasorum, which in turn lead to inflammation and damage to the elastic tissue and cause arterial aneurysm.

There are similarities in the angiographic and histological findings of the aneurysm in HSS and BD.[4,5] The vasculitis in both the conditions is associated with the development of pulmonary artery aneurysm. On the basis of these similarities, HSS has been described by some authors as a variant of BD, often designating different names to it such as "cardiovascular manifestation of Behçet's disease," "incomplete Behçet's disease," and "a rare case of Behçet's disease." However, typical symptoms of BD such as orogenital ulceration, uveitis, skin lesions, or arthralgia are not found in patients with HSS.

The laboratory findings of HSS are nonspecific with leukocytosis, anemia, elevated ESR, and C-reactive protein. Although conventional angiography is considered as the gold standard for the diagnosis of pulmonary artery aneurysm, in HSS, it may not be possible to do angiography in the patients showing vena caval thrombosis and there is a risk for aneurysmal rupture. Contrast-enhanced magnetic resonance angiography and multidetector computed tomography angiography may provide an alternative in such cases.[3]

Owing to the lack of controlled trials, there is no standard treatment guideline for the management of HSS. Most commonly, immunosuppressive therapy, using a combination of glucocorticoids and cyclophosphamide, forms the first-line medical management in the treatment of HSS. One such regimen is monthly pulses of cyclophosphamide (1 g) plus prednisolone (1 mg/kg/day) for at least minimum 1 year.[6] Other agents used in the treatment of HSS are colchicine, cyclosporine, and azathioprine. Antibiotics have no proven role in the management of HSS, while anticoagulants and fibrinolytic agents are generally contraindicated because of the increased risk of fatal hemorrhage.

For the cases of massive hemoptysis because of large pulmonary artery aneurysm or those with lesions confined to one segment or one lung, lobectomy, or pneumectomy can be carried out to remove the aneurysm. However, the high morbidity and mortality associated with surgery, and the bilateral and multifocal occurrence of the pulmonary artery aneurysm at the time of diagnosis, makes surgical resection difficult. Under the circumstances, transcatheter pulmonary arterial embolization is an alternative treatment modality in selected cases of HSS.

Conclusion

HSS is clearly a rare but grave clinical entity with most of the data in the form of sporadic case reports. The disease should be suspected in young male patients with peripheral venous thrombosis and pulmonary or bronchial artery aneurysm. Hemoptysis can be a presenting picture and usually suggest poor prognosis. Early diagnosis and treatment is, therefore, crucial in improving the prognosis of patients with HSS. Appropriate treatment, if instituted promptly and early in the course of the disease, has the potential to induce remission.

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