Case Report

Hughes-Stovin syndrome: A rare cause of hemoptysis

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ABSTRACT

Multiple pulmonary artery aneurysms are seen along with venous thrombosis in Hughes-Stovin syndrome, which many investigators believe is an incomplete form of Behcet's disease. We present a case of hemoptysis with multiple pulmonary artery aneurysms, femoral vein thrombosis, and oral ulcers with emphasis on its CT features.

KEY WORDS: Aneurysms, hemoptysis, thrombosis, ulcers

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INTRODUCTION

Hughes-Stovin syndrome is an exceedingly rare disorder characterized by the combination of multiple pulmonary artery aneurysms and deep venous thrombosis; the cause is unknown.[1] Some authorities consider it as a forme fruste of Behcet's syndrome. We present a case of a middle-aged man in whom multiple pulmonary aneurysms were demonstrated by CT angiography and DSA. The patient had history of bilateral iliofemoral thrombosis and recurrent oral ulcers.

CASE REPORT

A 40-year-old man was evaluated for bilateral hilar opacities on radiographs by a contrast-enhanced CT of chest. The patient had significant history of bilateral femoral vein thrombosis two years back for which the patient was hospitalized; the patient also had recurrent episodes of hemoptysis for last one and a half year for which he did not undergo any imaging and which was managed conservatively with antibiotics. The patient had also developed aphthous ulcers in the past year that lasted for two to three weeks. There was no evidence of genital ulcers, uveitis, or skin lesions either historically or on physical examination. Laboratory studies were remarkable for a raised ESR. ANA and RF were absent; coagulation studies were normal. CECT chest revealed large saccular aneurysms involving the segmental branches of lower lobe arteries bilaterally and right upper lobe artery [Figure 1]. The bronchial arteries were hypertrophied; eccentric, organized thrombi were seen in few aneurysms [Figure 2]. Pulmonary angiography was performed through the jugular vein as transfemoral cannulation was not possible because of bilateral femoral vein thrombosis which confirmed the presence of multiple pulmonary artery aneurysms. The final diagnosis of Hughes-Stovin syndrome was made. The possibility of incomplete Behcet was also considered as the patient did not fulfill the diagnostic criteria of the complete Behcet's syndrome. The patient was put on topical steroids (as mouth paint for oral ulcers), systemic steroids, and azathioprine (in view of pulmonary vasculitis and aneurysm formation); the steroids were subsequently tapered and withdrawn after 6 months (aspirin and anticoagulation were avoided in view of hemoptysis). More than one year of follow up has documented the resolution of oral ulcers without any fresh recurrence; there has been no fresh episode of deep venous thrombosis. The patient had two episodes of mild hemoptysis which was managed conservatively. There was no evidence of enlargement of the pulmonary artery aneurysms on follow-up CT scans.

DISCUSSION

Hughes and Stovin first described cases of deep vein thrombosis and multiple pulmonary artery aneurysms with mural thrombi. The typical clinical syndrome is characterized by a stage of thrombophlebites, followed by formation and enlargement of pulmonary aneurysm, and the final stage of aneurysmal rupture with massive hemoptysis and death.

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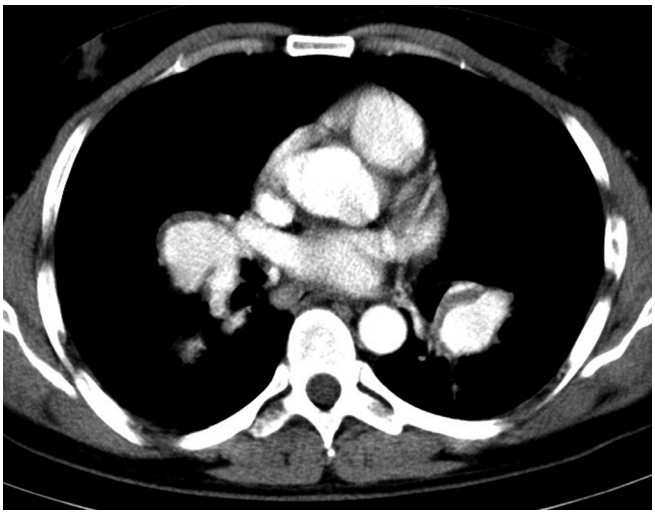

Figure 1: Contrast-enhanced CT chest shows bilateral pulmonary artery aneurysms arising from lower lobe arteries with organized eccentric thrombi in the wall

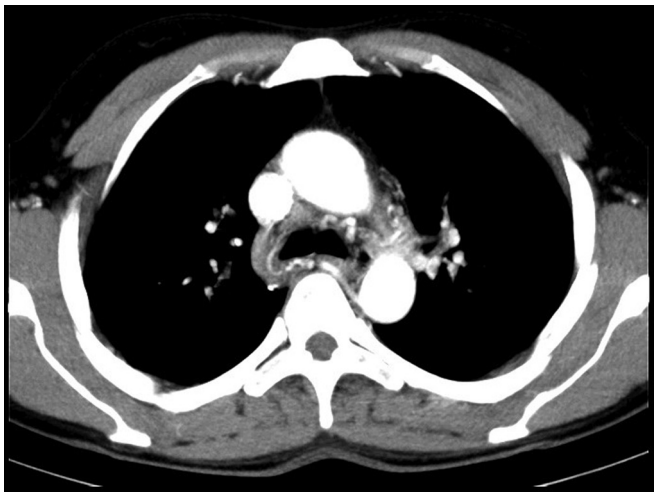

Figure 2: MIP images of CT angiography demonstrate the hypertrophic bronchial artery collaterals in the mediastinum

The cause of this entity is an idiopathic systemic vasculitis. Many investigators considered this entity as an incomplete form of Behcet's syndrome.[1-3]

The pulmonary artery aneurysms develop at the sites of prior thrombosis; weakening of the vessel wall due to inflammation is the likely mechanism. In our patient, eccentric organized thrombi were well visualized in few of the aneurysms on CT angiography. Some authors have reported dilated bronchial arteries on DSA; they believe that hemoptysis likely results from rupture of angiodysplastic bronchial arteries rather than due to rupture of pulmonary arteries.[2] Enlarged, hypertrophied bronchial vessels were clearly visualized in our case as well, which we believe was the cause of recurrent hemoptysis in this case. One author has described multiple bronchial artery aneurysms with a hepatic artery aneurysm in one patient.[4] CT angiography not only allows visualization of vessel lumen (like pulmonary angiography), it also depicts the mural thrombus, vessel wall, and mediastinal structures including the hypertrophied bronchial circulation.[5] The treatment of Hughes-Stovin syndrome is similar to Behcet's disease–steroids alone or in combination with immunosuppressants. Anticoagulation and platelet inhibitors are not indicated owing to risk of hemoptysis.[2] Our case emphasizes the overlap between the clinical and radiological features of Behcet's disease and Hughes-Stovin syndrome.

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