



Case Report

A Rare Case of Hughes Stovin Syndrome

Tanmay Bhat¹, Ruqia Asna Rabah², Sirinivas Kudwa³

¹Assistant Professor, Department of Medicine, K.S. Hegde Medical Academy, Mangalore.

²Senior Specialist, District Hospital Gulbarga, Karnataka

³Junior Resident, K.S. Hegde Medical Academy, Mangalore, India.

Corresponding Author: Ruqia Asna Rabah

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ABSTRACT

This is a case report of a young male patient with history of cough and hemoptysis since 4 months. He used to cough out blood tinged sputum daily. There was no history of exertional dyspnea or fever. He was admitted one year before in our hospital with right lower limb deep vein thrombosis involving the popliteal vein and superficial femoral vein. Basic investigations ruled out tuberculosis. Detailed evaluation showed that the patient had a very rare syndrome Hughes- Stovin syndrome.

Key Words- Hemoptysis, Pulmonary Artery, Aneurysm, Hughes Stovin Syndrome.

INTRODUCTION

Hughes-Stovin syndrome (HSS) was named after two British physicians, Drs. John Patterson Hughes and Peter George Ingle Stovin. They first described the findings of the syndrome (deep venous thrombosis and segmental pulmonary artery aneurysms) in a total of four male patients with pulmonary artery aneurysms in 1959.

[1] It is a very rare disease with fewer than 30 cases reported in the literature. It is defined by the presence of pulmonary artery aneurysm in association with peripheral venous thrombosis. The combination of pulmonary artery aneurysm and thromboembolic disease is uncommon but is reported in association with Behcet's disease. [2-3] Aneurysms observed in HSS maybe single, multiple, unilateral or bilateral. [4] These aneurysms generally involve the pulmonary and bronchial arteries

but can also occur anywhere in systemic circulation. The exact etiology and pathogenesis is unknown; possible causes include infections and angiodysplasia. The disease affects mainly adults, especially men. Typical symptoms are recurrent fever, chills, haemoptysis and cough. For cases of massive haemoptysis due to large pulmonary aneurysms or those with lesions confined to one segment or one lung, lobectomy or pneumectomy can be carried out. We present a case of Hughes Stovin syndrome where the thrombotic event preceded the development of pulmonary aneurysm by one year.

CASE HISTORY

A 25 year old male patient came with history of cough and hemoptysis since 4 months to our OPD. He used to cough out blood tinged sputum daily. There was no history of exertional dyspnea or fever. He

was admitted one year before in our hospital with right lower limb deep vein thrombosis involving the popliteal vein and superficial femoral vein. Routine blood tests and chest X-ray were normal. Protein C, protein S and antiphospholipid antibodies were negative. ANA was normal. He was on warfarin for past one year with regular follow up, maintaining INR in therapeutic range.

Examination and Investigations;

At admission, general physical and systemic examination was unremarkable. Vitals – pulse rate: 78/min; BP- 130/80 mm of Hg; Spo2- 98% in room air. Complete blood count, Renal function tests and liver function tests were within normal limits. PT – test: 30.9 and control: 14.8 with INR of 2.23, aPTT and BT were normal. ECG and Echocardiogram were within normal limits. Venous Doppler of right lower limb was suggestive of recanalization of old DVT. Chest X-ray showed a homogenous opacity in left lower perihilar area. Hence CT angiogram thorax was done that revealed thrombus with recanalization of pulmonary artery with aneurysm of distal left pulmonary artery measuring 4cm. Pulmonary angiogram showed a large fusiform aneurysm measuring 3.6 into 3.5 cms arising from left pulmonary artery lower branch with no thrombus or any filling defects seen within the aneurysm.

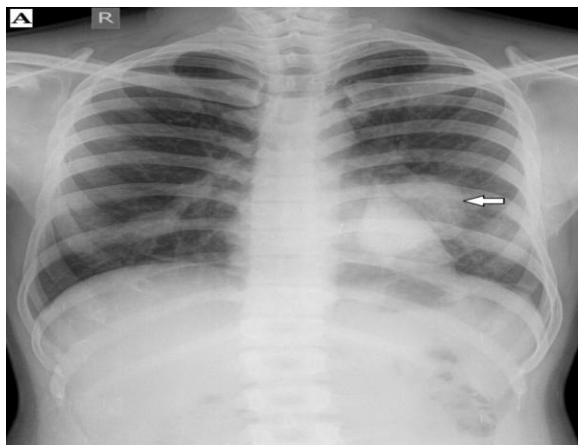


Fig. A –Chest X ray at admission

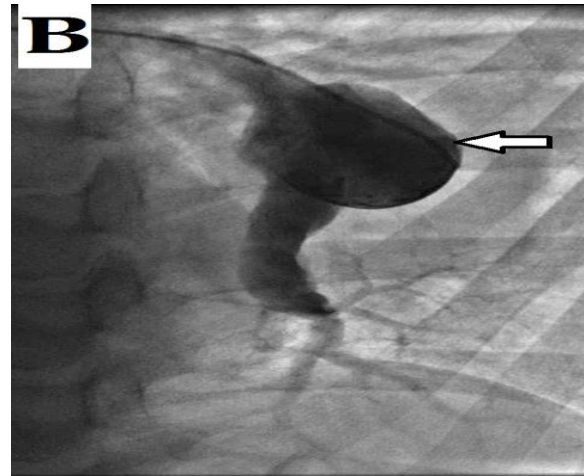


Fig B- Pulmonary angiogram at admission

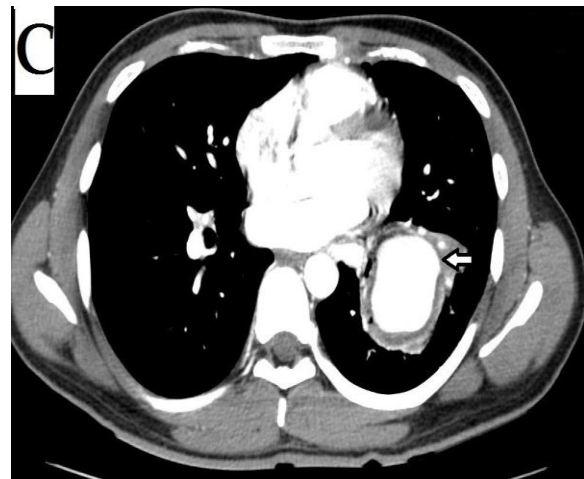


Fig C-CT scan chest at admission

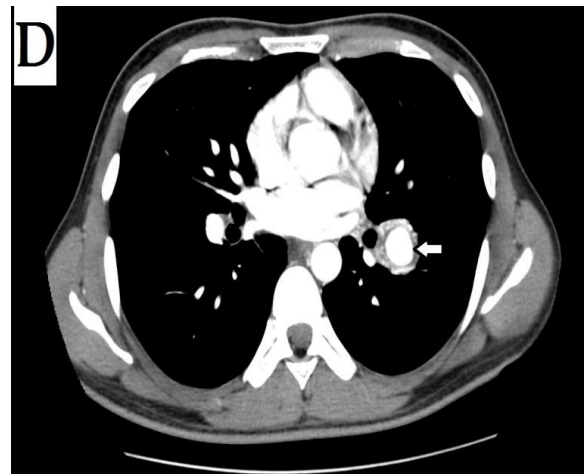


Fig D- CT scan chest during follow up

In view of persistent hemoptysis we decided to withhold warfarin. Hemoptysis

subsided within couple of days. As patient had risk of bleeding in the eventuality of aneurysmal rupture, warfarin could not be restarted. Hence patient was taken up for inferior vena caval filter insertion and also was started on Dabigatran 150mg daily to prevent pulmonary embolism in future. Patient came for further follow up after 6 months with no hemoptysis and CT angiogram showed reduction in aneurysm size to 2.4cm with surrounding irregular non-enhancing thrombus

DISCUSSION

The eponym “Hughes-Stovin syndrome” was used by Koop and Green in 1962. [4,5] Since pulmonary embolism rarely causes aneurysms, Hughes and Stovin postulated that congenital or degenerative changes of the bronchial arteries would result in inadequate nutrition of the pulmonary arteries, producing a type of embolus different from that seen in normal pulmonary arteries. [6] About 25% of patients with HSS develop thromboembolism, arterial aneurysms and vascular occlusions. The distribution of the vascular component of the syndrome is as follows: arterial (7%), venous (25%) or both (68%). The clinical paradigm of HSS can be divided into three phases: [7]

- a. Symptoms of thrombophlebitis
- b. Formation of large pulmonary and/or bronchial aneurysms
- c. Aneurysmal rupture leading to massive hemoptysis and death

Our patient had deep vein thrombosis followed by pulmonary aneurysm a year later. Hence the diagnosis of HSS was entertained. Some authors have suggested that Hughes-Stovin syndrome is a partial manifestation of Behçet’s disease. [8] The angiographic and histological findings were comparable. In addition to venous thrombosis, aneurysms of the pulmonary arteries are described in patients with

Behçet's disease. Both diseases are characterized by destruction of the wall of the pulmonary arteries and perivascular infiltration. [8] However, typical symptoms of Behçet's disease such as oral or genital ulcerations, skin lesions, iritis or arthralgia are missing in most of the patients with Hughes-Stovin syndrome.

This disorder does not have a standardized treatment. Few cases of this syndrome have been reported. A treatment similar to that used in Behçet’s disease, with isolate corticotherapy, or in association with immunosuppressants, has been suggested. The evidence is not very reassuring due to the fatal development of several cases, despite treatment. [9] In our case as patient improved after withholding warfarin and hence was managed without steroids as there is no proven benefit. Anticoagulants and thrombolytic agents are generally considered contraindicated due to an increased risk of fatal hemorrhage, even though they confer a beneficial effect in an embolic state. [10,11] Some patients with HSS already have hemoptysis at initial presentation; making these agents an unsafe therapeutic option.

CONCLUSION

Warfarin had to be withheld in the patient with this syndrome. Surgical intervention should be considered in some patients whose presentations involve life-threatening symptoms such as massive hemoptysis or who have a large aneurysm size.

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