Respiratory Medicine Case Reports 19 (2016) 98–102

Contents lists available at ScienceDirect

Respiratory Medicine Case Reports

journal homepage: www.elsevier.com/locate/rmcr

Case report

Life threatening hemoptysis from Hughes Stovin syndrome: Is it that rare?

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ARTICLE INFO

Article history:
Received 6 July 2016
Received in revised form
11 August 2016
Accepted 12 August 2016

Keywords:
Deep venous thrombosis
Thrombophlebitis
Hemoptysis
Pulmonary embolism
Pulmonary aneurysm

ABSTRACT

Introduction: Hughes-Stovin syndrome is a life-threatening disorder of unknown etiology. This condition is characterized by vasculitis, deep venous thrombosis and aneurysms that mainly involve the pulmonary arteries resulting in hemoptysis. It has been described in literature less than 40 times. However, we believe it is not very uncommon as it might be diagnosed as pulmonary embolism solely. In such cases, anticoagulation therapy augments the risk of life-threatening hemoptysis.
Materials and methods: We report the case of a 35 years old, Egyptian female patient with Hughes-Stovin syndrome, who initially presented with lower limb deep vein thrombosis and coughing of blood. Anti-coagulation regimen for pulmonary embolism was given. This resulted in massive hemoptysis that was successfully controlled by medical therapy.
Conclusion: Adults who present with venous thrombosis and hemoptoic cough, with no predisposing factors of thrombosis, normal platelet count and coagulation, the possibility of Hughes-Stovin syndrome has to be considered.

1. Introduction

Hughes-Stovin syndrome (HSS) is a life-threatening disorder of unknown etiology. This condition is usually presented by vasculitis, deep venous thrombosis and aneurysms that usually involve the pulmonary arteries as well as the bronchial arteries resulting in hemoptysis. In such cases, using anticoagulant therapy might be life-threatening as it augments the risk of hemoptysis [1,4].

2. Case report

A 35 years old female presented at the emergency room with coughing of blood (about half a cup all over the day) associated with bilateral below knee level non-pitting edema of two days' duration, more evident on the right side. She reported that her illness started four years ago by lower limb edema, tense and tender calf muscles and was diagnosed as bilateral acute deep venous thrombosis. She received anticoagulation therapy and stopped by herself after few months. One year ago, she developed dyspnea on moderate exertion, hemoptoic cough, chest pain and erythema nodosum. At that time, she was advised to take anti-inflammatory drugs and steroids to control the erythema nodosum, underwent lower limb duplex scan and multislice CT angiography of the pulmonary arteries that revealed bilateral chronic DVT and bilateral pulmonary embolism respectively. Vascular surgery didn't support the idea of IVC filter placement and advised to keep the patient on LMWH that resulted in partial improvement.

Based on the clinical presentation and patient's medical history she was admitted, kept on LMWH 80 IU twice per day and Warfarin 5mg/day. A new lower limb venous duplex scan and Multislice CT scan of the pulmonary arteries revealed Subacute bilateral DVT with similar findings in the CT pulmonary angiography Which stated the existence of thrombotic filling defects that involved the main right pulmonary artery, extending partially into the segmental lower lobe artery branches with adjacent ectatic bronchial arteries (Fig. 1), also with a smaller thrombotic circumferential filling defect at the left lower lobe artery branch, preserving a centrally patent residual lumen (Fig. 2). Unfortunately, such findings of suspicious pulmonary aneurysms with thrombotic process inside as well as dilated ectatic bronchial arteries could not be correlated at the time of presentation to Hughes Stovin syndrome

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http://dx.doi.org/10.1016/j.rmcr.2016.08.003

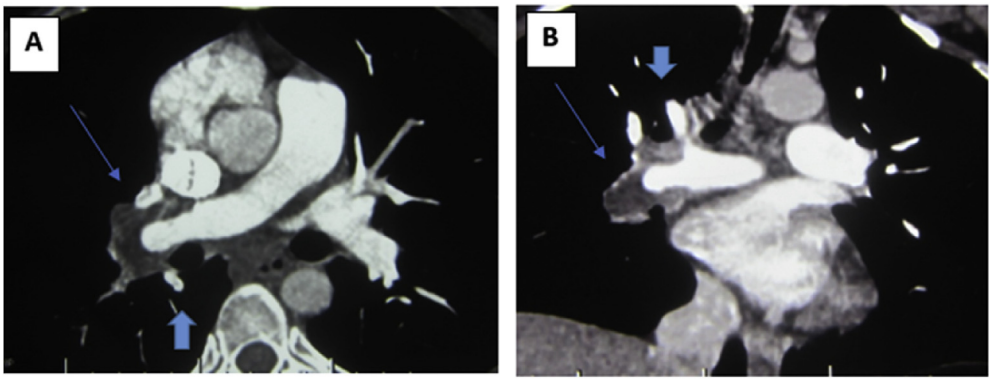

Fig. 1. CT angiography of the pulmonary arteries (a) Axial view (b) Coronal view showing right main pulmonary artery aneurysm with circumferential thrombotic filling defect (Thin arrows) and ectatic adjacent bronchial arteries (Thick arrows).

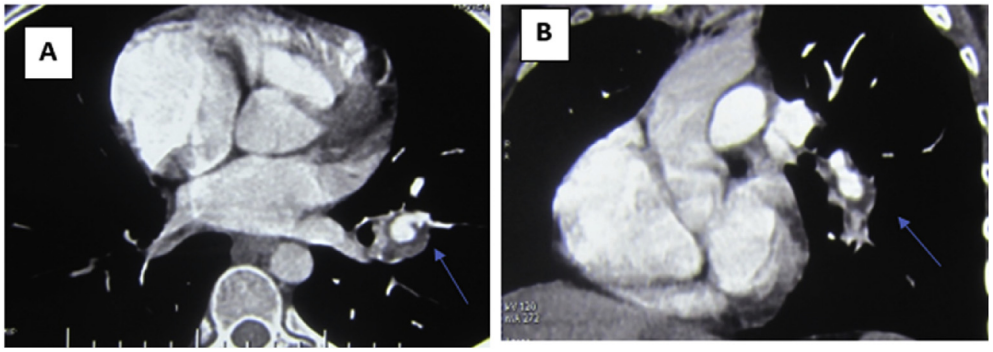

Fig. 2. CT angiography of the pulmonary arteries (a) Axial view (b) Coronal view showing left lower lobe pulmonary artery aneurysm with mural thrombotic filling defect (Thin arrows).

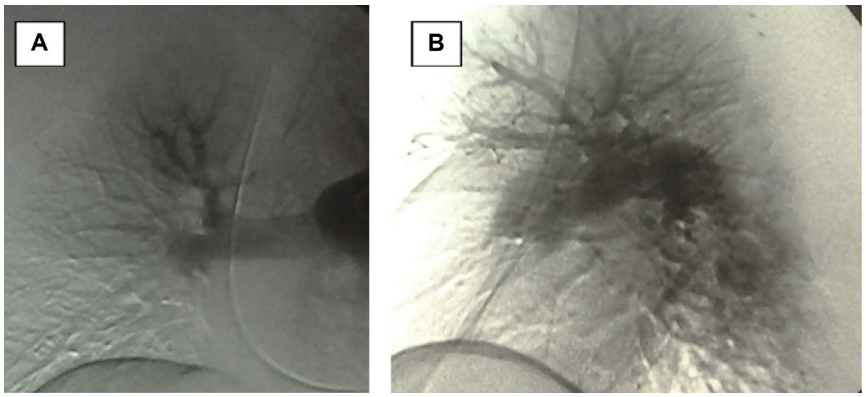

Fig. 3. Catheter angiography of the pulmonary arteries(a) Right pulmonary artery aneurysm (thick arrow) with lower lobe perfusion defect (curved arrow) (b) Left segmental pulmonary artery aneurysm (thick arrow).

(see Fig. 3).

Associated clinical findings included few primary erythema nodosum for which methylprednisolone 8mg 1 tablet/day and colchicine 0.5 mg two tablets twice daily were given. Co-existing vaginal bleeding was also investigated by transvaginal ultrasound that revealed right-sided hemorrhagic ovarian cyst.

Vital data and general laboratory work up like CBC, liver and kidney functions, T3, T4 and TSH were all fine. ESR was 74, ANA, ANCA, Antiphospholipid antibody, lupus anticoagulant, anti-cardiolipin, PCR for factor V and sputum culture and pathergy tests were all negative. Patient was diagnosed as DVT with PE and received the classical anticoagulation therapy, stayed in the