CHEST

Recurrent Pulmonary Aneurysms

Hughes-Stovin Syndrome on the Spectrum of Behçet Disease

Sami M. Bennji, MD; Leonard du Preez, MD; Stephanie Griffith-Richards, MD; Derrick P. Smit, MD; Jonathan Rigby, MD; Coenraad F. N. Koegelenberg, MD, PhD; Elvis M. Irusen, MD, PhD, FCCP; and Brian W. Allwood, MD, PhD

In this report, we describe a male patient who presented with recurrent life-threatening hemoptysis due to the sequential formation of multiple pulmonary aneurysms. Both pulmonary artery coil embolization and right lower lobectomy were performed, with limited success. The patient experienced extensive bilateral femoral DVT extending into the inferior vena cava, with massive hemoptysis, fulfilling the diagnosis of Hughes-Stovin syndrome. A final diagnosis of Behçet disease was made following extensive investigation, and the patient responded well to prednisone 20 mg orally and azathioprine 100 mg orally. CHEST 2017; 152(5):e99-e103

KEY WORDS: Behçet disease; Hughes-Stovin syndrome; pulmonary aneurysm

Case Report

A 34-year-old South African man initially presented to the ophthalmology clinic with a painful right eye, photophobia, and a decrease in visual acuity. He had no known medical history or significant family history and denied any history of skin rashes or rheumatologic, respiratory, or renal symptoms. Following slit-lamp examination, he was diagnosed as having panuveitis with an occlusive vasculitic component (Fig 1) possibly due to sarcoidosis. Subsequent conjunctival biopsy results were nondiagnostic, and a fundus fluorescein angiogram revealed occlusive vasculitis in both eyes. A chest radiograph obtained at the same time was noted to be normal. He was treated with dexamethasone 0.1% eye drops with good symptom resolution, and 1 year later was lost to follow-up.

Two years later, he was admitted to our hospital with massive hemoptysis. His presentation was preceded by a 1-month history of cough, fever, loss of weight, and night sweats. On examination, he was pale and had decreased breath sounds bilaterally. Laboratory values included a C-reactive protein level of 68 mg/L and a pretransfusion hemoglobin value of 10 g/dL.

CT angiography showed multiple bilateral pulmonary vascular abnormalities, initially interpreted as arteriovenous malformations but subsequently diagnosed as pulmonary artery aneurysms (Fig 2). The patient underwent coiled embolization of the pulmonary aneurysms bilaterally through a transfemoral vein catheter, with resolution of the hemoptysis (Fig 3).

He was discharged but presented again 3 months later with recurrent massive

AFFILIATIONS: From the Division of Pulmonology (Drs Bennji, Koegelenberg, Irusen, and Allwood), Department of Medicine, the Division of Cardiothoracic Surgery (Dr du Preez), Department of Surgery, the Department of Radiology (Dr Griffith-Richards), the Division of Ophthalmology (Dr Smit), Department of Surgery, and the Division of Anatomical Pathology (Dr Rigby), Department of Pathology and National Health Laboratory Services, Tygerberg Academic Hospital/Stellenbosch University, Cape Town, South Africa.

CORRESPONDENCE TO: Sami M. Bennji, MD, Department of Medicine, Stellenbosch University, PO Box 241, Cape Town, South Africa; e-mail: saminj12@gmail.com

DOI: http://dx.doi.org/10.1016/j.chest.2017.07.015

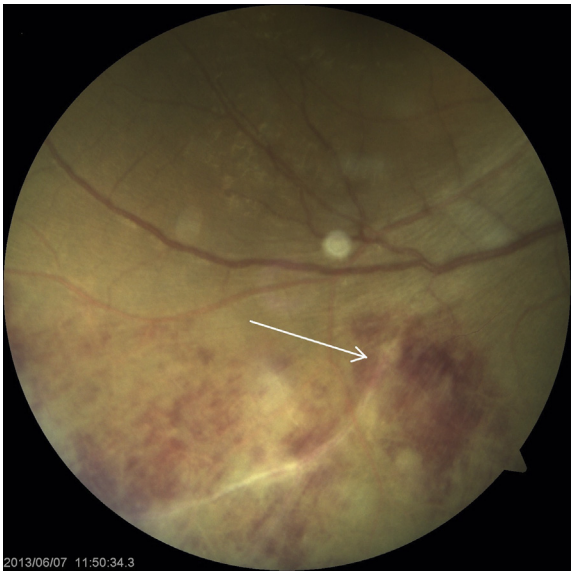

Figure 1 – *Color fundus photograph of the inferior retina of the right eye showing occluded blood vessel (arrow) with surrounding intraretinal hemorrhage.*

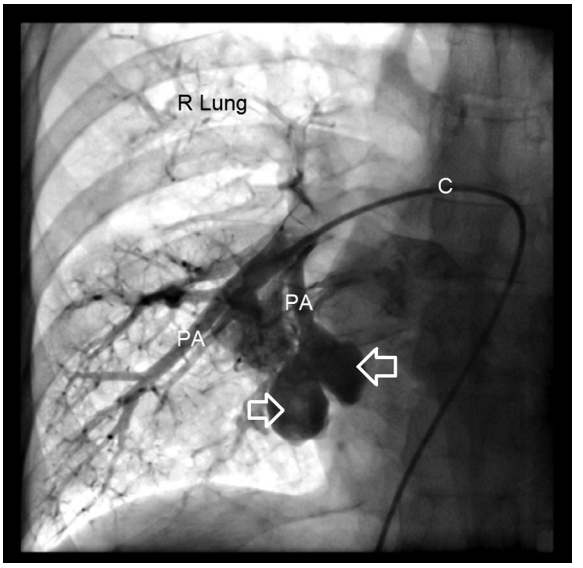

Figure 3 – *Embolization of the right pulmonary aneurysms. Note the two pulmonary artery aneurysms (arrows), as well as the catheter inside one of the pulmonary arteries of the right lung. C = catheter; PA = pulmonary artery; R = right.*

hemoptysis. Repeated radiologic examination showed a new large right perihilar mass that had evolved in the intervening period and was confirmed on CT angiography to be a new large pulmonary aneurysm in the right lower lobe (Fig 4). Repeated coil embolization was attempted but was abandoned when the patient was found to have thrombosis of his entire lower venous system. Abdominal ultrasonography revealed thrombosis extending bilaterally from the femoral veins through the iliac veins and into the inferior vena cava (Fig 5). In view of his ongoing episodic massive hemoptysis, anticoagulation was withheld and placement of a Greenfield filter was not considered technically feasible.

The patient underwent an emergency right lower lobectomy, which was technically challenging due to pulmonary engorgement because of the aneurysm and intrapulmonary hemorrhage, adhesions to the chest wall, an incomplete oblique fissure, and a difficult hilar

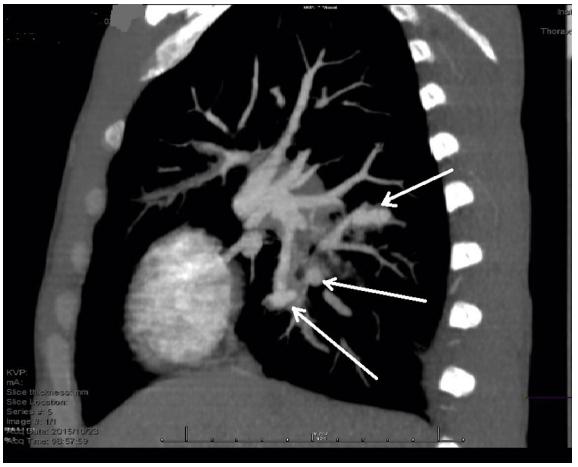

Figure 2 – *A sagittal reconstruction of the chest CT angiogram showing the right pulmonary vessels with multiple pulmonary artery aneurysms (arrows).*

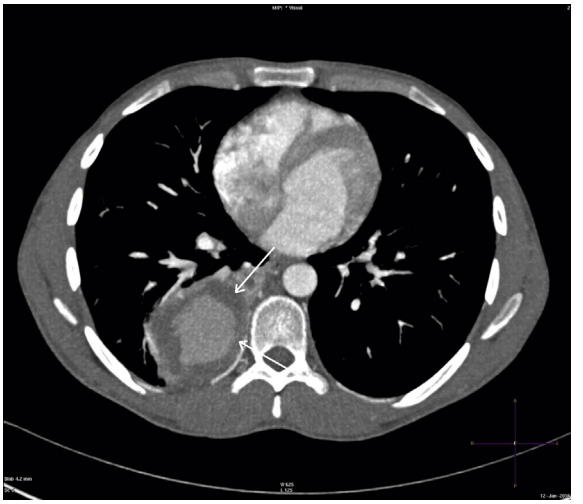

Figure 4 – *Chest CT angiogram showing a new large aneurysm in the right lower lobe with thrombus formation surrounding the aneurysm (arrows) giving rise to a heterogeneous appearance.*