Young onset hemoptysis: A rare cause of pulmonary arterial aneurysm

Sir,

A 19-year-old gentleman from Tamil Nadu, presented to us with complaints of cough and mild-to-moderate hemoptysis for four months and dyspnea on exertion (grade 1 Modified Medical Research Council) over the past two months. It was not associated with fever, chest pain, loss of weight or loss of appetite.

There has been no history of joint pain, skin rashes, oral ulcers, claudication pain, diminution of vision, double vision or hoarseness of voice. He does not have any other comorbidity and has no addictions.

His vitals were stable on presentation. Peripheral pulses were well felt. General examination was within normal limits. There were no signs suggestive of uveitis, and there were no skin rashes or skin discoloration. Systemic examinations were also within normal limits.

His total blood count was elevated (12,300 cells/mm^3) with neutrophilic predominance. X-ray of the chest showed right hilar prominence [Figure 1]. His sputum examinations were negative for acid fast bacilli (AFB) on direct smear and culture. Polymerase chain reaction (PCR) for *Mycobacterium tuberculosis* was also negative, but bacterial culture grew *Haemophilus Parainfluenzae*, which was treated with antibiotics. Computed tomography (CT) of the thorax with a pulmonary angiogram, which was done as part of hemoptysis evaluation, showed multiple segmental pulmonary artery aneurysms with filling defects - thrombus [Figure 2], consolidation in the right middle lobe and ground glass opacities in the right middle and lower lobes – which likely represented alveolar hemorrhage. On further evaluation, a venereal disease research laboratory test was negative and other bacterial and fungal infections were ruled out. The vasculitis workup was negative. Clinical screening for Marfan syndrome was negative. Echocardiography was done, which ruled out any cardiac anomaly and pulmonary hypertension. Thus, the differential diagnosis was narrowed down to Bechet's disease and Hughes Stovin syndrome.

He did not have oral or genital ulcers. His fundus examination ruled out uveitis and the pathergy test was negative. HLA B-51 analysis was also negative. An ultrasound Doppler screening of both the lower limbs was negative for any deep venous thrombosis. Thus, a diagnosis of Hughes Stovin Syndrome was made. He was started on pulse steroid and later on Mycophenolate Mofetil and tapering doses of steroids.

Hughes Stovin syndrome (HSS) is a rare autoimmune disorder, first described by John Patterson Hughes and Peter George Ingle Stovin, two British physicians, in 1959.[1] It is characterized by pulmonary artery aneurysms and deep vein thrombosis, it can present with recurrent fever, cough,

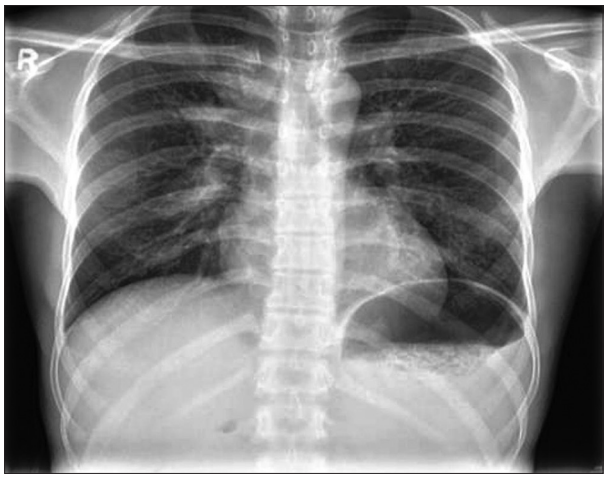

Figure 1: X-ray chest showing right hilar prominence

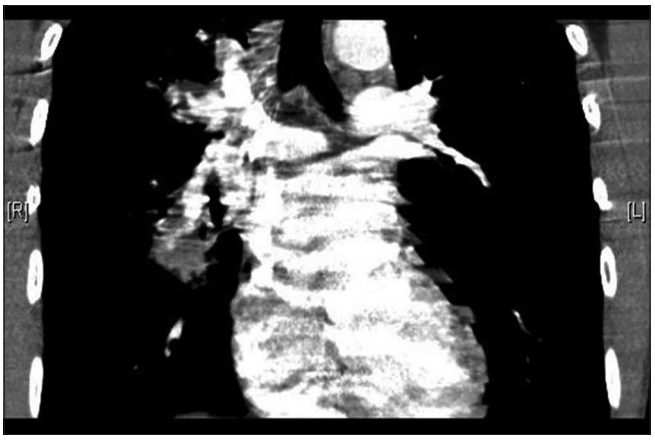

Figure 2: Computed tomography pulmonary angiogram (CTPA) showing multiple segmental pulmonary artery aneurysms with filling defects — thrombus

Table 1: Pulmonary artery aneurysm differentials[3]

Infection	Tuberculosis, Syphilis, other bacterial and fungal- can arise from right sided endocarditis
Structural cardiac abnormalities	Congenital and acquired cardiac abnormalities, atherosclerosis, Marfan's, Behcet's
Pulmonary hypertension	
Idiopathic vasculitis syndromes	Hughes Stovin Syndrome, Behcet's disease
Trauma	Eg. By Swan- Ganz catheter
Miscellaneous	

and hemoptysis. Radiologically it is similar to Bechet's disease, however, the other features of Bechet's, like oral or genital ulcers, uveitis, joint involvement, positive pathergy test, and the like, are absent in the Hughes Stovin syndrome. Therefore, it is also called as 'Incomplete Bechet's Disease'. Even though HSS is considered as a vasculitis syndrome, the exact pathogenesis of HSS is still unknown. As the clinical and radiological features are similar to that of Bechet's, a common pathogenesis is assumed.

The Hughes Stovin syndrome occurs predominantly in young men, between the second and fourth decades of life. A male predilection is noticed in the incidence cases of HSS that have been described in literature from various geographical locations, including North America, Africa, Asia, and Europe. There can be multiple pulmonary artery aneurysms associated with this syndrome, and in 50% of patients, this can be bilateral. However, there will not be any other lung involvement.[2] The differentials to be considered when a patient presents with pulmonary artery aneurysms are listed in the Table 1.[3]

Various hypothesis of pathogenesis of HSS have been postulated. Some of them are infections with Hepatitis A, B, C, E viruses, Herpes Simplex Virus (HSV), Parvovirus B19, *Chlamydia pneumoniae*, *Helicobacter pylori*, *Streptococcus mitis*, *Streptococcus salivarius*, *Streptococcus sanguis* and *Saccharomyces cerevisiae*.[4] Another leading hypothesis is of angiodysplasia of the bronchial arteries leading to lack of nutritional supply to pulmonary arteries via the vasovasorum leading to pulmonary artery wall degeneration.[5]

The disease is clinically graded into three stages

- Stage I - thrombophlebitis
- Stage II - formation and enlargement of pulmonary aneurysms
- Stage III - aneurysmal rupture

Stage-III can lead to massive hemoptysis and it can be fatal to the individual.

Radiological features of HSS seen in a CT-Pulmonary angiogram are systemic thrombi in the vena cava or cerebral sinuses, bronchial artery, and one or more pulmonary artery aneurysms and pulmonary artery stenosis on account of thrombus or emboli. Histopathological findings of pulmonary artery in HSS include diffused dilatation of the artery causing aneurysms with partial occlusions and lymphocytic perivascular infiltration. Diffused proliferative sclerosis can also be noticed.

Immunosuppressive therapy is the mainstay of therapy for HSS; Glucocorticoids and cyclophosphamide are the first-line of management. Steroids are introduced as intravenous pulse therapy, followed by a tapering dose of oral steroids. Cyclophosphamide will be continued up to one year following complete remission, but the steroid can be tapered and stopped depending upon the clinical improvement. This treatment can stabilize small pulmonary artery aneurysms and sometimes it can result in even regression of the smaller aneurysms. In our patient we have used Mycophenolate Mofetil because of the young age, better safety profile, and tolerability. Anticoagulation might have been of advantage, as HSS is a pro-thrombotic state, with the risk of embolism. However, on account of

the possibility of fatal hemorrhage, anticoagulation is of high risk. After stabilization of the condition with adequate immunosuppression, cautious anticoagulation may be able to provide a beneficial effect, but it requires support from further research.

Arterial aneurysm is a poor prognostic factor in comparison to venous aneurysms. In HSS, arteriovenous involvement is more common (68%), isolated venous involvement is 25%, and isolated artery involvement is 7%. Angiodysplastic bronchial artery rupture can be the cause of death rather than pulmonary artery aneurysm rupture, according to Mahlo *et al*.

Hughes Stovin Syndrome is an autoimmune disorder, which is similar to Behcet's disease in clinical presentation and radiological features. It is a diagnosis of exclusion. Other causes of pulmonary artery aneurysms should be looked for before establishing the diagnosis of HSS, as it is very rare. Rupture of aneurysms can lead to massive hemoptysis, which can be lethal to the patient. Early identification of the condition and initiation of immunosuppression can be lifesaving.

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Conflicts of interest
There are no conflicts of interest.

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