

Pulmonary artery aneurysm: Harbinger of an ominous disease

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ABSTRACT

An 11-year-old girl presented with chronic cough and recurrent hemoptysis. On examination, she had features of right heart failure and cyanosis, with severe pulmonary hypertension on echocardiogram. Computed tomography pulmonary angiography showed aneurysmal dilations of the pulmonary artery with elevated erythrocyte sedimentation rate and C-reactive protein and positive human leukocyte antigen B51. A diagnosis of Hughes–Stovin syndrome (vascular variant of Behcet’s syndrome) was confirmed, and she was started on immunosuppression, on which there was improvement.

Keywords: Hughes–Stovin syndrome, pulmonary artery aneurysm, pulmonary artery thrombus

INTRODUCTION

Aneurysmal dilatation of pulmonary artery is a rare cause of hemoptysis. We report a 11 year old girl who was diagnosed with this uncommon disorder, and had a much sinister cause underneath.

CASE REPORT

An 11-year-old girl presented with a history of nonproductive cough for 1 year, with few episodes of hemoptysis, followed by progressive exertional dyspnea for the past 2 weeks. There was no history of oral or genital ulcers, rash, or eye involvement. On examination, she had mild cyanosis (SpO₂ 86% on room air) with normal mean jugular venous pressure with prominent A waves. Cardiovascular examination revealed features of elevated pulmonary artery pressure. Systemic examination was otherwise unremarkable.

Electrocardiogram showed sinus tachycardia with right-axis deviation and right ventricular hypertrophy. Echocardiogram showed severe

pulmonary hypertension (right ventricle systolic pressure [RVSP] = 60 + right atrial pressure [RAP], Peak pulmonary regurgitation pressure 36mm Hg) with good biventricular function and no evidence of thrombus in the proximal pulmonary artery.

Chest X-ray showed dilated main pulmonary artery and retrocardiac soft tissue shadow on the left side [Figure 1]. Computed tomography (CT) pulmonary angiogram showed dilated proximal pulmonary arteries with thrombotic occlusion of the right pulmonary artery distal to hilum; descending branch of the left pulmonary artery was aneurysmally dilated (40 mm × 42 mm × 68 mm) with eccentric mural thrombus [Figure 2]. Right-sided pulmonary blood flow was maintained by hypertrophied bronchial arteries. CT aortic angiogram showed a small saccular aneurysm of the infrarenal abdominal aorta with eccentric thrombus [Figure 3]. Pulmonary parenchyma was normal. Ventilation perfusion scan showed absent perfusion in the entire right lung and patchy perfusion loss in the left lung.

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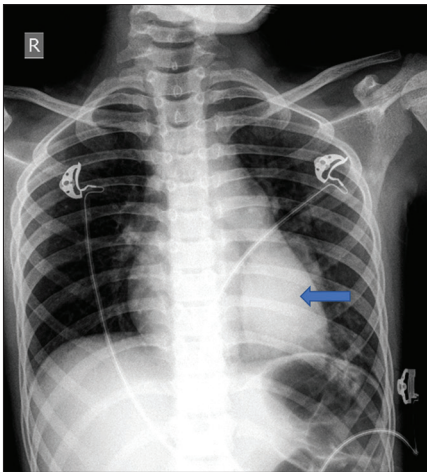


Figure 1: Chest X-ray showing dilated main pulmonary artery with retrocardiac soft tissue shadow (arrow)

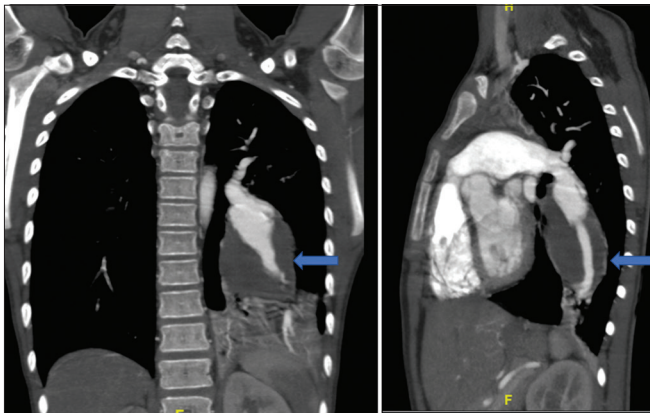


Figure 2: Contrast enhanced computed tomography showing aneurysmally dilated left lower pulmonary artery with thrombus (arrow)

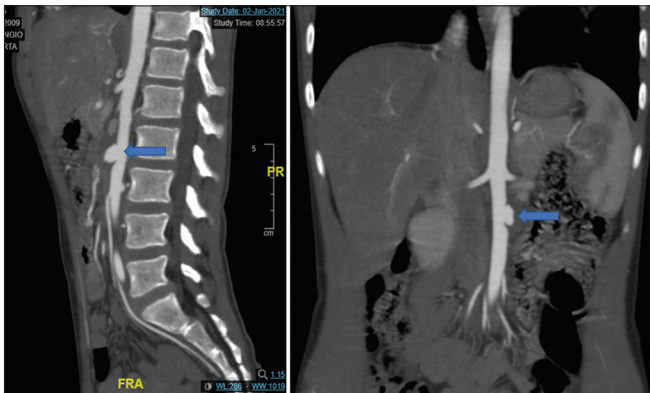


Figure 3: Contrast-enhanced computed tomography showing saccular outpouching of infrarenal abdominal aorta

Investigations revealed normochromic normocytic anemia with elevated erythrocyte sedimentation rate and C-reactive protein. HLA B51 was found to be positive; all other autoimmune workup was negative. Venous Doppler of the upper and lower limbs was normal.

As she did not fit the diagnostic criteria of Behcet's syndrome but had the typical vascular involvement associated with the disease along with HLA B51 positivity, a diagnosis of Hughes-Stovin syndrome (HSS) was made. She was treated with three pulse doses of methylprednisolone followed by oral steroids and monthly cyclophosphamide. ESR was reduced to 23 mm/h and there was improvement in SpO₂-94% on room air; echocardiogram also showed significant reduction of pulmonary hypertension (RVSP = 40 + RAP, peak PR - 30). As she has only one functioning lung, she is planned for right pulmonary endarterectomy followed by left lower lobectomy after her inflammatory markers are well controlled.

DISCUSSION

Deterling and Clagett first described pulmonary artery aneurysm in an autopsy case series.^[1] These are extremely rare and more commonly seen in main pulmonary artery (89%) and involve younger age group as compared to those with aortic aneurysms with equal sex distribution.^[2]

More than 50% of cases are associated with congenital heart disease, mainly patent ductus arteriosus, ventricular septal defect, and atrial septal defect. Acquired causes include infections such as syphilis, tuberculosis and pyogenic bacteria, pulmonary neoplasms and metastases, iatrogenic injury from interventions and surgery, chronic pulmonary thromboembolism, and vasculitis such as Behcet's disease (BD) and HSS.^[3]

BD is an autoimmune disorder characterized by aphthous oral and genital ulcers, uveitis, arthritis, chronic meningoencephalitis, pericarditis, and vasculitis. Involvement of major arteries is seen in 10%-30% of patients with predominant aneurysm formation.^[4]

HSS is a rare vasculitis characterized by multiple aneurysms of the pulmonary artery and venous thrombosis. It was first described by John Patterson Hughes and Peter George Ingle Stovin and is considered by many as the vascular variant of BD since the characteristic ulcers, arthritis, and uveitis are absent.^[5]

Less than 50 cases have been described so far in the literature. The disease affects young adults in the second decade of life with male predilection and can be divided into three phases - symptoms of thrombophlebitis, formation of pulmonary and bronchial artery aneurysms, and aneurysmal rupture, leading to pulmonary hemorrhage and death.^[6] Histologically, there is diffuse dilatation and partial occlusion of the aneurysmal arteries with predominant lymphomonocytic perivascular infiltration and diffuse proliferative sclerosis with annihilation of elastic and muscular fibers.^[7]

There are no clear management guidelines of HSS. Immunosuppression is considered the first-line therapy including pulsed cyclophosphamide and methylprednisolone followed by oral maintenance.^[8] Anticoagulants and thrombolytics are generally contraindicated in view of the frequent occurrence of hemoptysis and increased risk of pulmonary hemorrhage and have been used in few case reports where the pulmonary thrombosis is life-threatening.^[9] Antiplatelets are also contraindicated.^[10]

Surgery is usually recommended in patients with acute rupture, leading to massive hemoptysis, expanding aneurysm, and severe ischemia. Procedure usually includes resection with reconstruction with prosthetic tube or venous graft.^[11] However, surgical mortality is high, and there is a 25% chance of postoperative recurrence.^[12] Transcatheter arterial embolization is another option for those at high surgical risk and involves the use of steel coils, Ethibloc, and cyanoacrylate.^[13] The overall prognosis of this disease is dismal with a high incidence of mortality and pulmonary hemorrhage.

In our case, the presence of pulmonary aneurysms with thrombi and HLA B51, with lack of uveitis and ulcers pointed toward the diagnosis of HSS. We initially started her on immunosuppression with steroids and cyclophosphamide, on which she had good response with improvement of SpO₂ and reduction of pulmonary hypertension. Anticoagulation was withheld in apprehension of rupture and hemorrhage, on the background of past history of hemoptysis. Once the active inflammation is controlled, high-risk surgical endarterectomy of the right pulmonary artery followed by excision of the left lower lobe may provide symptomatic relief. However, the risk of recurrence still remains and prognosis largely depends on control of active disease with anti-inflammatory drugs.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient (s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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