Thrombosis of aneurysmal pulmonary arteries in patent ductus arteriosus with Eisenmenger syndrome

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ABSTRACT

A 45-year-old lady with patent ductus arteriosus with Eisenmenger’s syndrome had presented with hemoptysis. Computed tomography revealed aneurysmally dilated pulmonary arteries with a large calcified organized thrombus.

Keywords: Eisenmenger syndrome, patent ductus arteriosus, pulmonary thrombus

INTRODUCTION

Eisenmenger syndrome (ES) can have variable presentation and manifestations. Thrombosis of aneurysmal pulmonary arteries is reported in patients with patent ductus arteriosus who develop ES. We present the case of a 45 year old lady with the aforesaid condition who presented with hemoptysis.

A 45-year-old lady, presented with multiple episodes of hemoptysis of 3 days’ duration, associated with exertional dyspnea and nonproductive cough.

She was diagnosed to have large patent ductus arteriosus (PDA) with Eisenmenger syndrome (ES) at the age of 15 years when she had presented with progressive exertional dyspnea and cyanosis. Transthoracic echocardiogram had shown bidirectional shunting across the PDA with a right to left shunting on saline contrast echocardiography. Cardiac catheterization study showed significant systemic desaturation, severe pulmonary arterial hypertension, and pulmonary vascular disease (pulmonary vascular resistance 32 Wood units, and systemic vascular resistance 15 Wood units). She was kept on medical follow-up with pulmonary vasodilators and iron supplementation. She underwent three sessions of phlebotomy for symptomatic polycythemia in the last 2 years.

On examination, she had cyanosis (SpO2 64%) with grade III pandigital clubbing and elevated jugular venous pulse with prominent C-V waves. Cardiovascular examination showed wide split S2 with a loud P2, and 3/6 pansystolic murmur at left lower sternal border and 2/6 ejection systolic murmur in left upper sternal border. The rest of the systemic examination was unremarkable. Investigations showed a hemoglobin of 17.3 g/dl, with normal platelet count (160 × 10^3/mm^3) and normal liver and renal function tests.

Electrocardiogram showed sinus rhythm with right axis deviation, bialtrial enlargement, and right ventricular hypertrophy. Chest X-ray showed cardiomegaly with dilated main and right pulmonary artery and prominent main pulmonary arteries. Pulmonary angiography showed a large calcified thrombus in the upper lobe pulmonary arteries.

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arteries [Figure 1a]. Echocardiogram showed fair left ventricular function (LVEF 52%), large PDA (74 mm) with bidirectional shunting and severe pulmonary arterial hypertension (estimated right ventricular systolic pressure = 112 + right atrial mean pressure, severe pulmonary regurgitation with a peak velocity of 66 mm Hg), and a doubtful mass in the pulmonary artery compressing the left atrium [Figure 1b].

Contrast-enhanced computerized tomography of the chest was done which showed dilated main (42 mm) and left (26 mm) pulmonary arteries. The right pulmonary artery (RPA) was aneurysmally dilated (110 mm × 58 mm × 63 mm) with calcification and near-complete occlusion by thrombus. The dilated RPA was seen compressing the left atrium, superior vena cava, and right pulmonary veins [Figure 1c]. Multiple aortopulmonary collaterals were noted arising from bilateral subclavian arteries, aortic arch, descending thoracic aorta, and abdominal aorta. Bilateral symmetric ground-glass opacities were also noted, suggestive of chronic pulmonary arterial hypertension.

In view of poor prognosis and high surgical risk, she was discharged on pulmonary vasodilators and iron supplementation.

**DISCUSSION**

ES is the end result of congenital acyanotic heart diseases with left-to-right shunts, where due to the development of pulmonary vascular disease, right-to-left shunting ensues. It is associated with both the thromboses due to endothelial damage, procoagulant activation, and impaired fibrinolysis, as well as bleeding tendencies because of abnormal platelet function, thrombocytopenia, and clotting factor deficiency.[1]

In fact, the original case report published by Dr. Eisenmenger had pulmonary artery thrombosis described at autopsy. Wood’s series also reported the prevalence of pulmonary thrombus in about 25% of cases.[2]

Silversides et al. first analyzed 34 patients with a mean age 42 ± 10 years with ES, out of which PDA accounted for only 9%, while ventricular septal defect accounted for the majority (65%). Pulmonary artery thrombosis was seen in 7 (21%) of patients and was more commonly associated with female sex and lower oxygen saturation (<80%).[3]

Out of the 54 patients with ES studied by Broberg et al., PDA was present in only 7 (12.96%). The mean age was 38.4 years, and 67.2% were female. Pulmonary artery thrombus was seen in 20% of cases and was significantly associated with older age, history of hemoptysis, presence of atrial septal defect, increased pro-brain natriuretic peptide, dilated pulmonary arteries on imaging, and left ventricular systolic dysfunction. Surprisingly, none of the cases with PDA had evidence of thrombus, and no correlation with sex, hematocrit, platelet count, or prothrombotic factors could be established.[4]

Anticoagulation has a limited role in the treatment of thrombosis in patients with ES, in view of bleeding tendencies and difficulty to accurately monitoring international normalized ratio.[5] It can also lead to hemoptysis in about one-fifth of patients and can cause life-threatening bleed in about 8%. Hence, while prophylactic anticoagulation is routinely advised for those with primary pulmonary hypertension, data in patients with ES are conflicting and require more research.[6]

As depicted in our index case, pulmonary thrombosis should always be kept as a differential in patients with ES who present with hemoptysis and mandates exclusion by adequate imaging techniques. However, the prognosis is not favorable in these patients and the role of anticoagulation is debatable.

**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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