A Healthy Young Lady with 'Unprovoked' Persistent Bilateral Pulmonary Embolism - Why?

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Abstract

Pulmonary embolism (PE) was reported in about 9% patients with antiphospholipid syndrome (APS). Seronegative APS is an entity which demonstrates clinical manifestations highly suggestive of APS but persistently negative APS antibodies. A 31-year-old lady presented with a two-month history of exertional dyspnoea. She had two consecutive miscarriages at 12 and 14 weeks, previously. Physical examination revealed a thin lady who was tachycardic, tachypneic, hypoxic but normotensive. There was a loud P2 without signs of heart failure. Investigations revealed a type 1 respiratory failure, sinus tachycardia with right ventricular strain pattern, cardiomegaly with normal lung fields, and dilated right atrium and right ventricle with increased in pulmonary arterial pressure of 70mmHg from echocardiography. CT pulmonary angiography (CTPA) confirmed the presence of PE over bilateral pulmonary arteries. She continued to have exertional dyspnoea and was readmitted 9 months later with worsening dyspnoea. APS antibodies performed during both admissions were negative. Seronegative APS was diagnosed. Interestingly, two CTPAs performed at 6 months and 9 months after initial presentation revealed persistent bilateral pulmonary embolism. The provoking factors for PE should be sought because 'unprovoked' PE especially in young individuals need further attention. APS, including seronegative APS, should be considered. Persistence of symptoms of PE also warrants further attention as chronic thromboembolic pulmonary hypertension (CTEPH) may be the cause. Currently, there are available medical and surgical treatment of CTEPH, therefore establishing its diagnosis is important and it is best performed in pulmonary hypertension expert centre.

Keywords: Pulmonary embolism; seronegative anti-phospholipid antibodies, chronic thromboembolic pulmonary hypertension (CTEPH)

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