CHRONIC THROMBOEMBOLIC PULMONARY ARTERIAL HYPERTENSION SECONDARY TO PRIMARY APS – A CASE REPORT

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BACKGROUND

Chronic thromboembolic pulmonary arterial hypertension (PAH) is a condition that occurs due to obstruction of the pulmonary artery due to unresolved thromboembolic disease (PAH group 4). Its incidence is unknown, but it is estimated to occur in 1–5% of survivors of acute pulmonary embolism. As for its pathogenesis, the most popular theory refers to the presence of a hypercoagulable state (with positive anti-phospholipid antibodies in 10–20% of patients).

CASE REPORT

Female, 30 years old, with a report of recurrent deep vein thrombosis (DVT) in the right lower limb (first episode 10 years ago), using warfarin anticoagulation (with INR in the therapeutic range), with dyspnea on mild exertion for a long time, at least 6 months, with no further investigation to date. In addition, she reported a past of two obstetric losses with a gestational age < 20 weeks. During physical examination, mild cognitive deficit and asymmetric edema in the right lower limb is notable. Imaging exams were performed: Doppler ultrasonography of the right lower limb with evidence of common femoral vein rethrombosis; contrast-enhanced chest CT showing signs of bilateral chronic thromboembolism; transthoracic echocardiogram with 66% ejection fraction, significant PAH (PASP 72) and right ventricle with reduced systolic function. Laboratory tests were also performed, with thrombocytopenia, ANA 1/160 (dense fine speckled pattern with positive metaphase plate) being seen; anticardiolipin IgG 92 (VR: positive if > 20); anticardiolipin IgM 3.1 (VR: positive if > 20), both repeated after 12 weeks, corroborating the diagnosis of antiphospholipid antibody syndrome. Due to recurrent episodes of thrombosis despite the use of anticoagulation, hydroxychloroquine and simvastatin were started, and warfarin was maintained. During hospitalization, she acquired COVID-19 (mild condition), with hospital discharge and outpatient follow-up at a tertiary pulmonology service.

CONCLUSION

There is no evidence for routine surveillance for PAH after an acute thromboembolic event. However, patients with pulmonary embolism with a report of new or persistent dyspnea or exercise intolerance after three months of oral anticoagulation use should be followed up with imaging tests, as the natural history of chronic thromboembolic pulmonary hypertension typically progresses to failure right heart and death.

KEYWORDS

Thrombosis, Pulmonary hypertension, Antiphospholipid antibody syndrome.