Pulmonary Hypertension due to Antiphospholipid Syndrome mimicking CTEPH

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Background: Antiphospholipid syndrome (APS) is characterized by endothelial activation and recurrent thrombosis associated with antiphospholipid antibodies (aPLs). Pulmonary arterial hypertension (PAH) is the second most common lung complication in APS patients. Especially, chronic thromboembolic pulmonary hypertension (CTEPH) is associated with thrombotic APS. However, concurrent development of APS-related non-thrombotic PAH is rarely reported. We report a case of group 1 PAH related to APS mimicking CTEPH.

Case presentation: A 56-year-old woman presented with dyspnea on exertion (NYHA 3) for 3 months. There was no history of calf pain, prolonged immobilization or any other co-morbidity. Chest CT, performed at another hospital, revealed PTE (Figure A). Despite the treatment for 2 weeks of rivaroxaban 15mg/day, dyspnea persisted. She was referred to PAH clinic for further evaluation. Echocardiogram and right heart catheterization (RHC) revealed PAH with right heart dysfunction: mPAP (mmHg) 41, PAWP (mmHg) 10, and PVR (WU) 5.3 (Figure B/C). Blood tests revealed persistently positivity for lupus anticoagulant, while the levels of protein C and S, ANA, and other aPLs were negative. 6-minutes walking distance (6MWD, meters) was 534. Initially, she was diagnosed as CTEPH with primary APS. The rivaroxaban was increased to 20mg/day combined with digoxin 0.25mg/day. After 11 months, chest CT showed disappearance of filling defects in pulmonary arteries (Figure A). RHC revealed slightly improvement of PAH but still remained. After 12 months, her dyspnea aggravated and RHC showed progressive PAH (Figure C). Stimultaneously with the diagnosis of PTE, serial RHC revealed non-thrombotic PAH related to primary APS. She received PAH-specific medication, macitetan 10mg/day. After 15 months, the size of right ventricle was normalized and PAH was markedly improved: mPAP 25 and PVR 2.51 (Figure B/C).

Conclusion: This case shows concurrent development of PTE and non-thrombotic PAH associated with primary APS mimicking CTEPH. A multidisciplinary approach and serial follow up of RHC are important to make an early diagnosis and provide optimal treatment to patients with APS-related PAH.