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A giant pulmonary artery aneurysm in a patient with severe idiopathic pulmonary arterial hypertension

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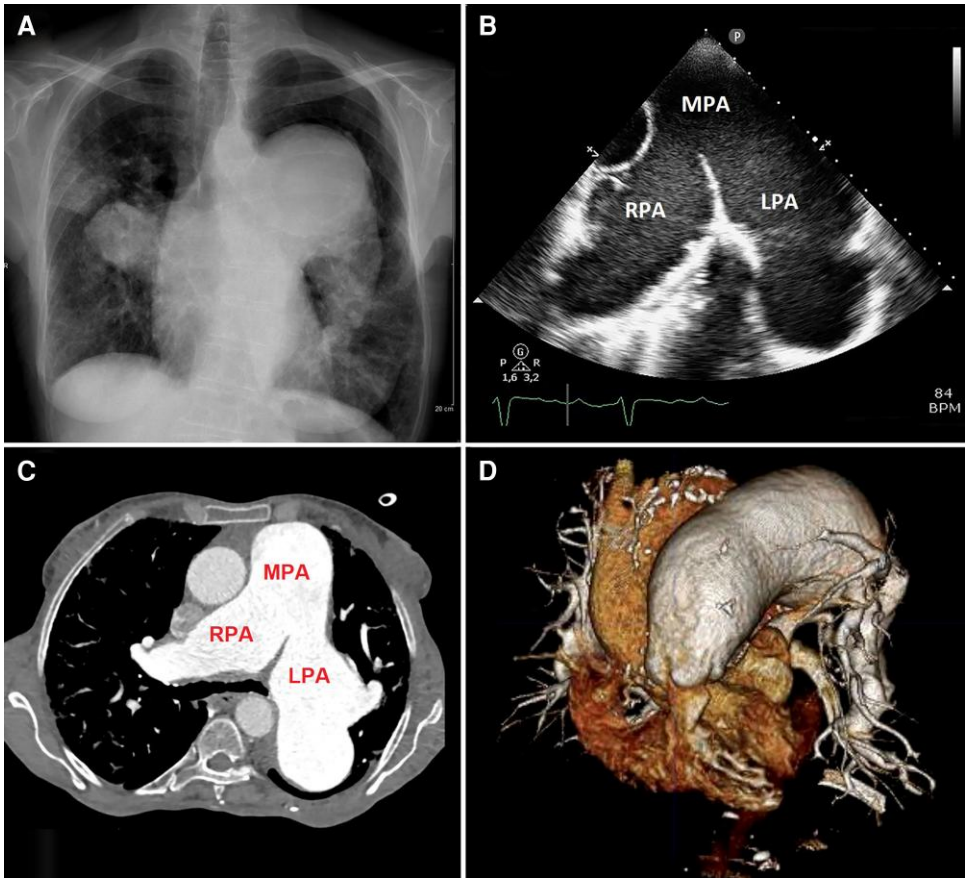

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A 81-year-old woman with a history of atrial fibrillation and pulmonary hypertension was referred to our Outpatient Clinic for evaluation of chronic dyspnoea. Physical examination revealed a diffuse low-pitch systolic murmur on the right hemithorax. Electrocardiography showed right ventricular (RV) hypertrophy consistent with chronic RV pressure overload. Chest X-ray documented mediastinal widening, with huge dilatation of the main pulmonary artery (MPA) and its main branches (*Panel A*). On transthoracic echocardiography, the RV was moderately dilated (RV to left ventricular basal diameter ratio = 1.3), with mildly reduced RV systolic function (TAPSE <18 mm) and severe pulmonary hypertension (tricuspid regurgitation velocity >3.4 m/s). On the basal short-axis view, a giant aneurysmal dilatation of the MPA (7.7 cm maximal transverse diameter) and both left and right branches (5.9 and 5.1 cm maximal transverse diameter, respectively) was observed (*Panel B*; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery). Spontaneous echo-contrast was observed at the level of PA bifurcation, without evidence of thrombosis (see Supplementary material online, *Movie S1*); RV outflow tract velocity time integral was low (8.5 cm), indicating slow blood flow within the PA aneurysm (PAA). Contrast-enhanced computed tomography (CT) scan of the chest confirmed the giant PAA, without thrombosis or dissection (*Panel C*; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery. *Panel D*: three-dimensional reconstruction of CT showing the giant left-sided PAA). As no specific causes could be identified, we assumed the PAA was secondary to severe, long-standing idiopathic pulmonary hypertension. Because of patient's advanced age and absence of PAA-related complications, conservative treatment with anticoagulation, macitentan, tadalafil, and inhaled treprostinil was chosen.

Ethics statement

This study was conducted in accordance with the World Medical Association Declaration of Helsinki. The Local Ethics Committee decided that ethics approval was not required in a single case image. No personal identifying information was included in this manuscript.

Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports*.

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Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in accordance with COPE guidelines.

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Data availability

The data underlying this article will be shared on reasonable request to the corresponding author.