Pulmonary Circulation

Don't stop at first glance: pulmonary artery angiosarcoma mimicking chronic thromboembolic pulmonary hypertension

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Abstract

We report the case of an incomplete diagnosis of chronic thromboembolic pulmonary hypertension, with relevant prognostic implications, missing the presence of a primary pulmonary artery angiosarcoma. After the late neoplasm diagnosis, the patient, treated for months with riociguat, was considered inoperable and died soon after.

This case highlights the need to manage patients with suspected pulmonary arterial hypertension by expert referral centers with specific and multi-professional expertise (heart and thoracic imaging) in order to avoid incomplete or delayed diagnoses.

Keywords

pulmonary hypertension, pulmonary arterial hypertension, echocardiography, angiosarcoma

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A 77-year-old woman with chronic thromboembolic pulmonary hypertension (CTEPH), diagnosed a few months before (Fig. 1), was referred for epoprostenol therapy because of a poor response to riociguat. The woman previously refused surgical thromboendoarterectomy. The admission echocardiography showed right chamber dilation and an extremely high atrioventricular gradient (100 mmHg (Fig. 2a)). An assessment of the right heart revealed an acceleration at the main pulmonary artery of 3 m/s associated with a hypoechoic, irregular mass, extending from the right ventricular outflow tract to the pulmonary artery bifurcation (Fig. 2b), never previously described. All procedures performed were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

However, a prior computed tomography (CT) scan had already shown an irregular mass in the right ventricular outflow tract and main pulmonary artery. The tissue was heterogeneously hypo-isodense, in continuity with the arterial and ventricular wall, suggesting a non-thrombotic nature (Fig. 1). A new angio-CT scan confirmed the presence of a low-density subocclusive structure, presumably a primary pulmonary artery angiosarcoma (Fig. 2c and d). After cardiac surgery work-up, the patient was considered inoperable on the basis of the extension of the tumor; the patient died ten days later.

This case highlights the need to manage patients with suspect pulmonary arterial hypertension (PAH) or CTEPH in adequate referral centers with specific and multi-professional expertise (heart and thoracic imaging), as already reported elsewhere.[1–3]

The echocardiographic exam is a fast and easily available exam, but a systematic approach, not limited to the single disease focused on at that moment, is paramount.

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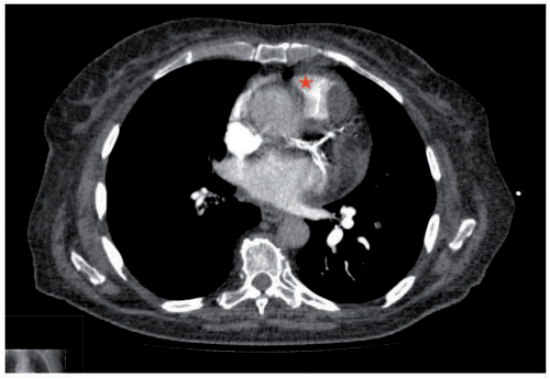

Fig. 1. CT scan before admission showing an irregular mass in the right ventricular outflow tract and main pulmonary artery, presumably non-thrombotic.

Echocardiographic exam should be performed by experienced cardiologists in the diagnosis of PAH or CTEPH, in order to exclude cardiovascular and non-cardiovascular co-morbidities (congenital heart diseases, Tetralogy of Fallot, neoplasms). Even a diagnosis of pulmonary thromboembolism exclusively based on echocardiography may be occasionally misleading without a careful diagnostic work-up.[4] The involvement of the right ventricular outflow tract and pulmonary valve by extraluminal infiltration, and small or no change in size after appropriate anticoagulation at the ultrasound follow-up, are in favor of diagnoses other than pulmonary embolism. The presence of tumors should be also considered in order to avoid any delay in required treatments (chemotherapy, surgical debulking, or percutaneous stenting).

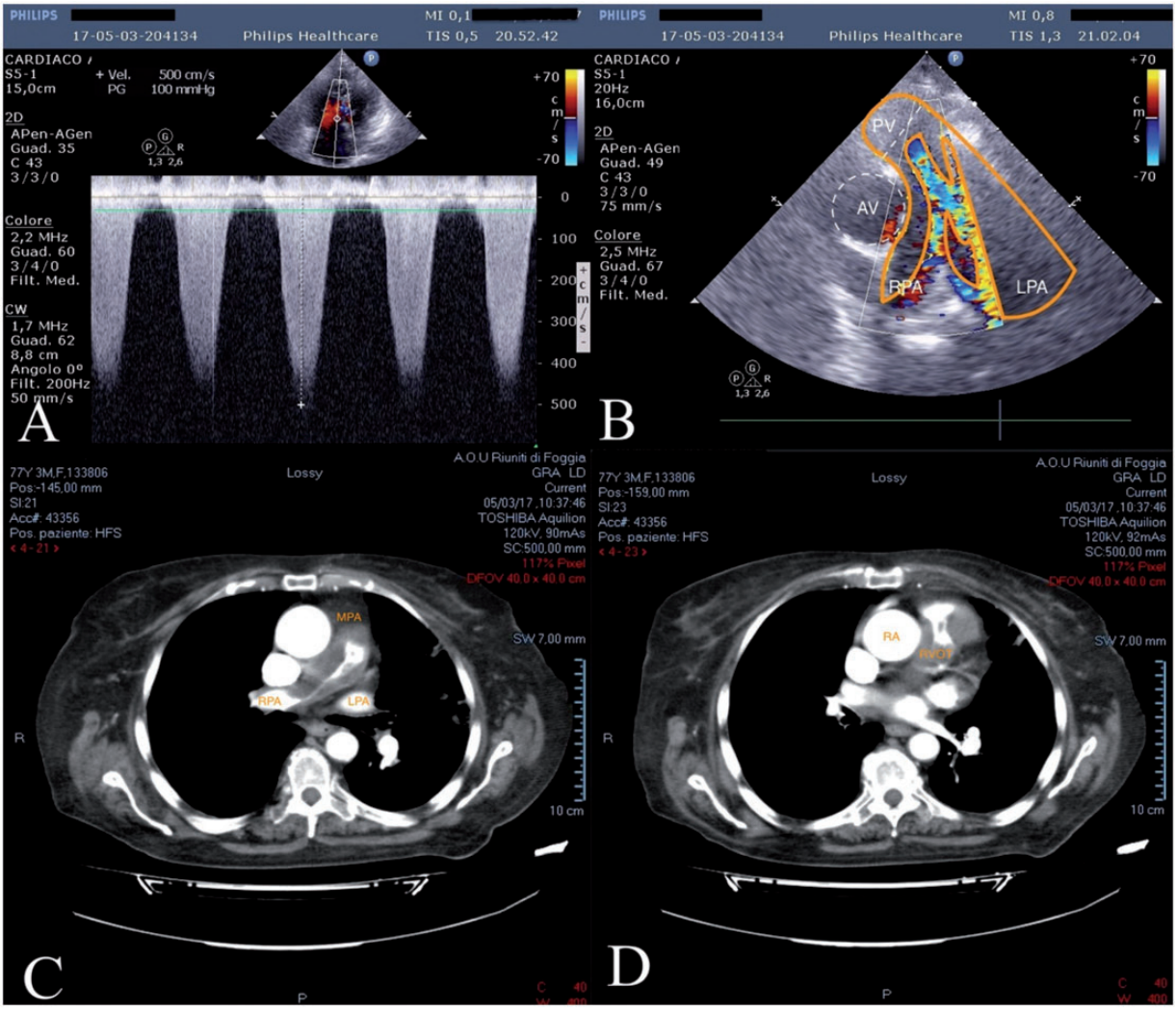

Fig. 2. (a) Right ventricle pressure gradient esteemed by tricuspid regurgitation velocity; (b) parasternal short-axis view showing endoluminal hypoechoic mass of the MPA and its branches determining blood flow acceleration; (c, d) chest angio-CT confirming the presence of a nearly occlusive hypodense formation of MPA extending from the RVOT. AV, aortic valve – in dashed circle; MPA, main pulmonary artery; LPA, left pulmonary artery; PV, pulmonary valve – in dashed line; RPA, right pulmonary artery; RVOT, right ventricular outflow tract.

Conflict of interest

The author(s) declare that there is no conflict of interest.

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