

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

Michele Correale¹, Nicola Tarantino², Riccardo Ieva¹, Matteo Gravina², Grazia Casavecchia², Caterina Strazzella², Matteo Di Biase³ and Natale Daniele Brunetti²

¹Cardiology Department, Ospedali Riuniti University Hospital, Foggia, Italy; ²Department of Medical & Surgical Sciences, University of Foggia, Foggia, Italy; ³Clinica Santa Maria, Gruppo Villa Maria Care & Research, Bari, Italy

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

Date received: 20 March 2018; accepted: 30 May 2018

Pulmonary Circulation 2018; 8(3) 1–3

DOI: 10.1177/2045894018785047

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 thromboendarterectomy. 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.

Corresponding author:

Natale Daniele Brunetti, Department of Medical & Surgical Sciences, University of Foggia, Foggia, Italy.

Email: natale.brunetti@unifg.it



Creative Commons Non Commercial CC BY-NC: This article is distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 License (<http://www.creativecommons.org/licenses/by-nc/4.0/>) which permits non-commercial use, reproduction and distribution of the work without further permission provided the original work is attributed as specified on the SAGE and Open Access pages (<https://us.sagepub.com/en-us/nam/open-access-at-sage>).

© The Author(s) 2018.

Reprints and permissions:
sagepub.co.uk/journalsPermissions.nav
journals.sagepub.com/home/pul



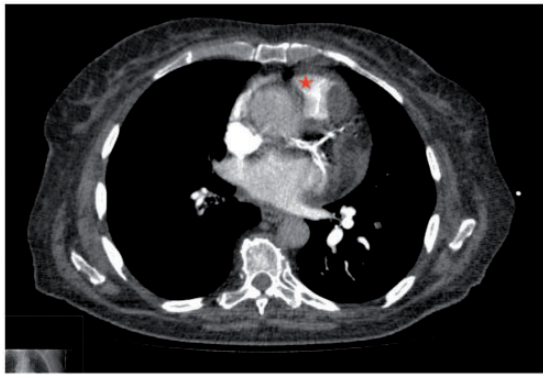


Fig. 1. CT scan before admission showing an irregular mass in the right ventricular outflow tract and main pulmonary artery, presumably non-thrombotic.

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).

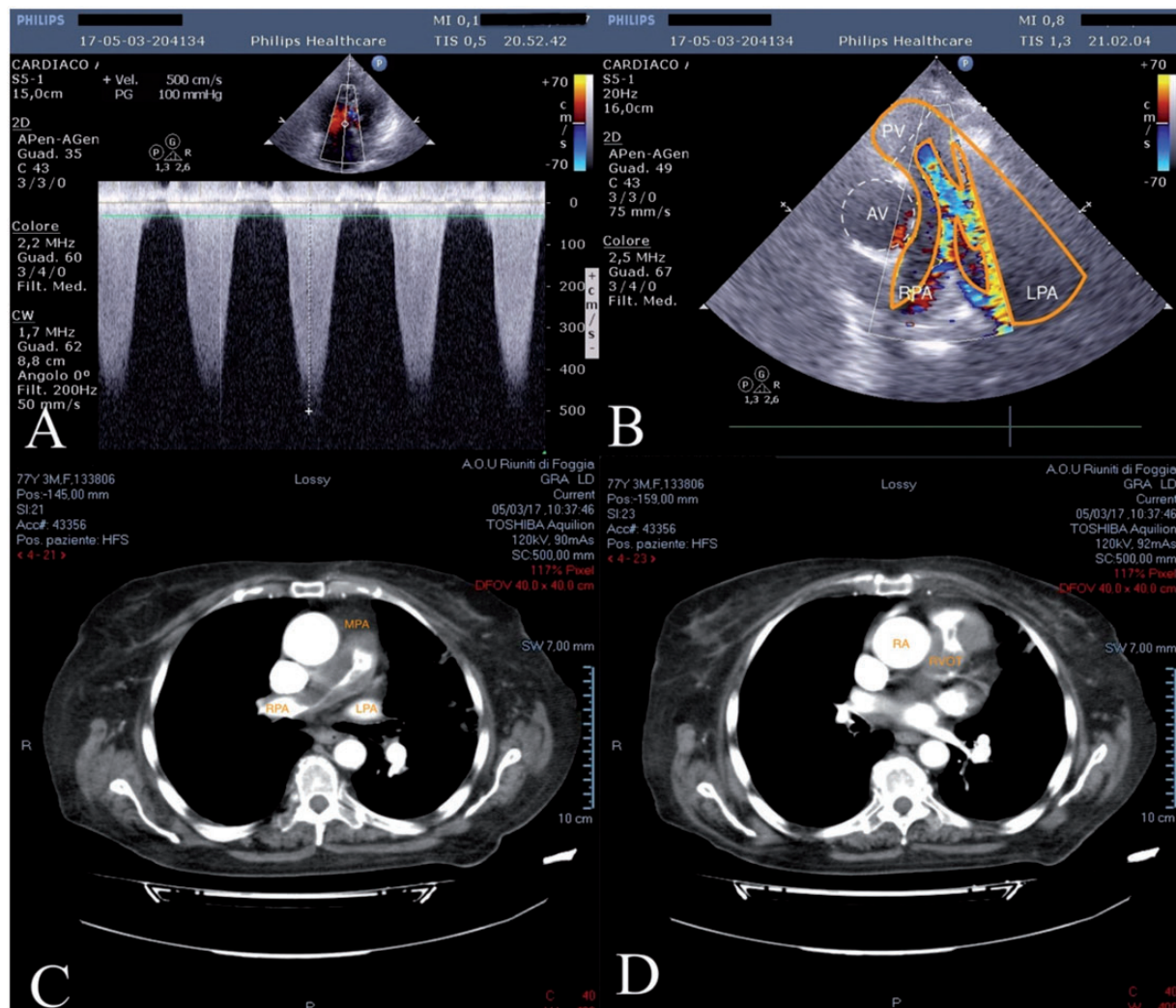


Fig. 2. (a) Right ventricle pressure gradient esteemed by tricuspid regurgitation velocity; (b) parasternal short-axis view showing endoluminal hypoechoic mass of the MPA and its branches determining blood flow acceleration; (c, d) chest angio-CT confirming the presence of a nearly occlusive hypodense 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.

Funding

This research received no specific grant from any funding agency in the public, commercial, or not-for-profit sectors.

ORCID iD

Natale Daniele Brunetti  <http://orcid.org/0000-0001-9610-7408>.

References

1. Correale M, Lacedonia D, D'Andrea G, et al. Chronic thromboembolic pulmonary hypertension. *Neth Heart J* 2015; 23: 193.
2. Correale M, Montrone D, Lacedonia D, et al. Multiprofessional and intrahospital experience for diagnosis and treatment of pulmonary arterial hypertension. *Monaldi Arch Chest Dis* 2012; 78: 205–209.
3. D'Amato N, Correale M and D'Agostino C. Aortic thrombus and acute pulmonary embolism in an individual heterozygous for the MTHFR C677T mutation. *Rev Esp Cardiol* 2010; 63: 1366.
4. De Gennaro L, Giannoccaro V, Lopriore V, et al. New onset right ventricular enlargement in recent dyspnea: Is echocardiography enough for a diagnosis of pulmonary thrombo-embolism? *Heart Lung* 2014; 43: 328–330.