ESC European Society of Cardiology

European Heart Journal – Case Reports (2021) **5**(7), 1–2
doi:10.1093/ehjcr/ytab255

An unusual case of pulmonary arterial hypertension

Juan F. Bulnes [1], Pablo Castro[1,2]*, Pablo Sepúlveda[1], and Hugo Verdejo[1,2]

[1]Division of Cardiovascular Diseases, Faculty of Medicine, Pontificia Universidad Católica de Chile, 362 Diagonal Paraguay st, 7th floor, Santiago 8320000, Chile; and [2]Advanced Center for Chronic Diseases (ACCDIS), 1007 Sergio Livingstone st, Santiago 8380492, Chile

Received 23 March 2021; first decision 27 April 2021; accepted 3 June 2021

A 51-year-old Hispanic female with no relevant medical history exhibited progressive dyspnoea on exertion without haemoptysis, fever, malaise, skin lesions, or arthritis. She denied having oral or genital aphthae, as well as ocular or neurological diseases. Physical examination showed jugular ingurgitation, a wide splitting of the second heart sound, with a loud pulmonary component. Lung auscultation was normal. She had no clubbing or peripheral oedema.

Transthoracic echocardiogram revealed severe pulmonary hypertension (PH), with an estimated systolic pulmonary artery pressure of 91 mmHg, and signs of chronic right ventricle pressure overload.

Computed tomography angiography revealed pulmonary arteries with severe tortuosity and multiple foci of dilatation and stenosis, showing a 'string of beads' appearance, without evidence of pulmonary embolism (*Figure 1*). There were no lesions either in the carotid or renal arteries, suggestive of fibromuscular dysplasia.

Complete blood count, C reactive protein, and sedimentation rate were normal. A serological evaluation for systemic vasculitis and connective tissue diseases was negative.

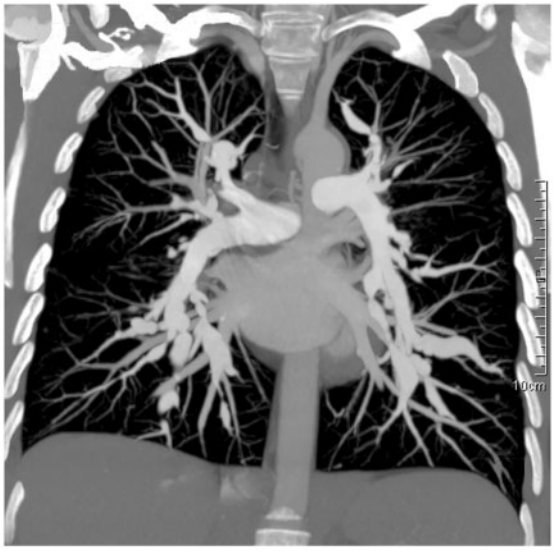

Figure 1 Computed tomography angiography showed pulmonary arteries with severe tortuosity and a 'string of beads' appearance.

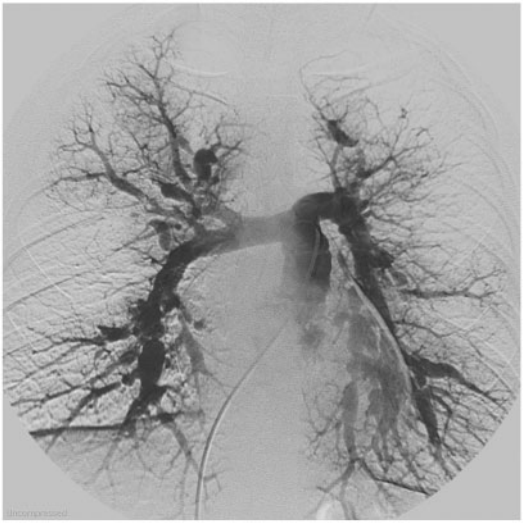

Figure 2 Pulmonary angiogram revealed multiple, bilateral aneurysms, scattered areas of narrowing, and severely compromised perfusion of both lungs.

* Corresponding author. Tel: +56 2 23543633, E-mail: pcastro@med.puc.cl
Handling Editor: Takeshi Kitai
Peer-reviewers: Aiste Monika Jakstaite and Elad Asher

Right heart catheterization showed precapillary PH (mean pulmonary artery pressure = 49 mmHg; pulmonary capillary wedge pressure = 9 mmHg; pulmonary vascular resistance = 11 Wood units), without intracardiac shunts. Pulmonary angiogram revealed multiple, bilateral aneurysms, scattered areas of narrowing with few intraluminal filling defects, and severely compromised perfusion of both lungs (*Figure 2*).

Specific therapy for pulmonary arterial hypertension was started, including Sildenafil, Ambrisentan, and inhaled Treprostinil.

Bilateral pulmonary artery aneurysms are very unusual findings, sometimes associated with congenital heart diseases with left-to-right shunting and systemic vasculitis, particularly Behçet disease.[1] In this case, a patient with idiopathic PH developed this severe form of vasculopathy. Although the mechanism remains largely unknown, there appears to be a link between the *vasa vasorum* in the adventitia and hyper-proliferative plexiform lesions seen in advanced idiopathic PH, which could lead to aneurysm formation.[2,3]

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 line with COPE guidance.

Conflict of interest: None declared.

Funding: This work was supported by FONDECYT (1181097 to P.C.) and FONDAP 15130011.

References

1. Kreibich M, Siepe M, Kroll J, Höhn R, Grohmann J, Beyersdorf F. Aneurysms of the pulmonary artery. *Circulation* 2015;**131**:310–316.
2. Humbert M, Guignabert C, Bonnet S, Dorfmüller P, Klinger JR, Nicolls MR et al. Pathology and pathobiology of pulmonary hypertension: state of the art and research perspectives. *Eur Respir J* 2019;**53**:1801887.
3. Oshima K, Crockett ES, Joshi SR, McLendon JM, Matsumoto Y, McMurtry IF et al. Aneurysm-type plexiform lesions form in supernumerary arteries in pulmonary arterial hypertension: potential therapeutic implications. *Am J Physiol Lung Cell Mol Physiol* 2019;**317**:L805–L815.