Received: 5 February 2024 | Accepted: 13 March 2024

DOI: 10.1002/rcr2.1330

CASE REPORT

Respirology Case Reports | WILEY

# Successful embolization of a clinically significant pulmonary arteriovenous malformation

**Pedro Fernandes** | **João P. Silva** | **Pedro Patrão** | **António Reis** | **Teresa Belo**

Pulmonology and Imagiology, ULS Dão-Lafões, Viseu, Portugal

**Correspondence**
Pedro Fernandes, Pulmonology and Imagiology, ULS Dão-Lafões, Rua Tenente Cabeleira Filipe, 23, 2430-306, Marinha Grande, Portugal.
Email: pdsf94@gmail.com

**Associate Editor:** Johnny Chan

## Abstract

The authors present the clinical case of a 59-year-old female patient with a history of peripheral desaturation, which was detected in the perioperative period 4 years earlier. She reported exertional dyspnea, quantified as grade 2 on the Modified Medical Research Council (mMRC) Dyspnea Scale (walks slower than people of the same age because of dyspnea or has to stop for breath when walking at her own pace), and morning cough with mucoid sputum and denied platypnea, epistaxis, telangiectasias and hemoptysis. A computed chest tomography scan revealed a contrast-enhanced lesion on the right upper lobe with an afferent and two efferent vessels compatible with pulmonary arteriovenous malformation. The transesophageal echocardiogram revealed an important right–left shunt compatible with arteriovenous fistula in the pulmonary circulation. An angiography confirmed the diagnosis and a selective embolization of the afferent artery was performed with resolution of symptoms.

**KEYWORDS**
desaturation, dyspnea, pulmonary arteriovenous malformation, right–left shunt

## INTRODUCTION

Pulmonary arteriovenous malformations (PAVMs) are communications between pulmonary arteries and veins.[1] They occur more frequently in females, between the ages of 30 and 40 years-old and the aetiology is variable, being more frequent in hereditary hemorrhagic telangiectasia (HHT) and also in cases of liver cirrhosis, penetrating chest trauma, mitral stenosis and in some cases idiopathic.[1,2] Clinically it usually presents with platypnea (dyspnea in an upright position, which improves with recumbency), orthodeoxia (low oxygen saturation in the upright posture and improvement when lying down), epistaxis, hemoptysis and telangiectasia and is associated with multiple important embolic and infectious complications such as brain abscess, stroke and hemorrhagic events.[1,3,4] This condition is commonly diagnosed when an imagological image shows a lung nodule with an afferent and efferent vessel. Transthoracic contrast echocardiography should be performed to identify and quantify the shunt. If the shunt is intracardiac, the contrast is visible in the left ventricle in less than one cardiac cycle. In intrapulmonary right-to-left shunt, contrast is visible in the left ventricle between the next three to eight cycles. The number of bubbles of contrast visible in the left ventricle determines the grade of the shunt. Computed chest tomography scan (chest-CT) should then be used to determine the size of the afferent vessel to define if it is possible to perform a selective embolization of the vessel. Embolization of the afferent vessel is highly effective and is associated with few complications, but the relapse rate is approximately 15%. The prognostic depends on the aetiology, but is associated with a mortality of 10%, with two thirds of deaths occurring from cerebrovascular complications, reinforcing the importance of treatment.

## CASE REPORT

The authors present the case of a non-smoker 59-year-old female patient with history of peripheral desaturation in rest that was detected in the perioperative period of a cholecystectomy. She reported exertional dyspnea, quantified as grade 2 on the Modified Medical Research Council (mMRC) Dyspnea Scale (walks slower than people of the same age

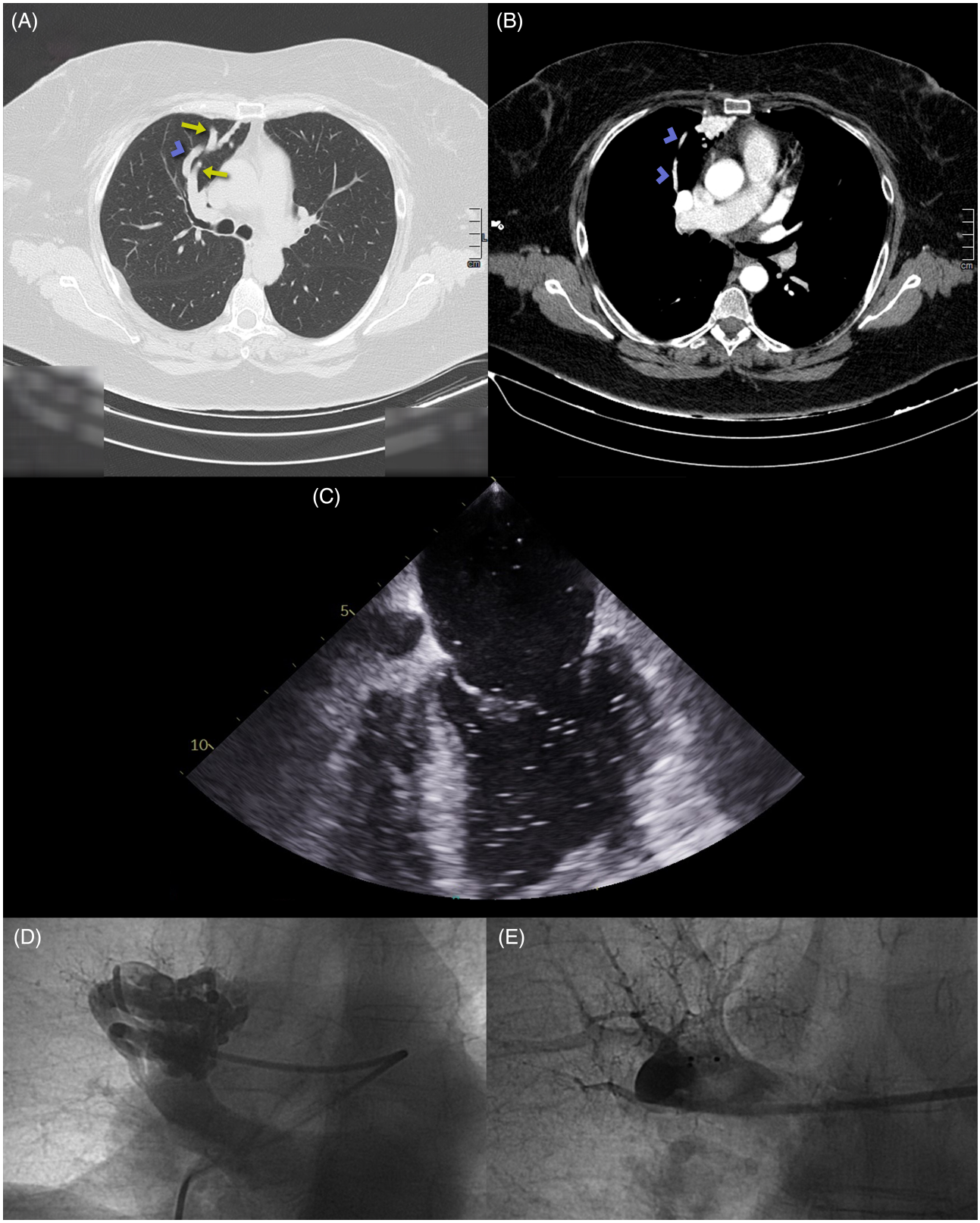


**FIGURE 1** Computed chest tomography with a 3 × 4cm contrast-enhanced lesion on the right upper lobe (A), apparently with an afferent with 8 mm diameter and an efferent vessel (B), compatible with pulmonary arteriovenous malformation. (C) Transesophageal echocardiography with contrast showing bubbles in the left ventricle. Aortography with pulmonary arteriovenous malformation with reflux of contrast to the heart (D) followed by selective embolization of the afferent artery with two AVPlugs (E).

because of dyspnea or has to stop for breath when walking at own pace) and denied platypnea, epistaxis, telangiectasias or hemoptysis.[3,5] The patient was obese (BMI 38 Kg/m2) and further physical examination revealed pulmonary auscultation with a global decrease in breath sounds and cardiac auscultation was normal. Peripheral oxygen saturation was 87% on both upright position and supine. There was no sign of telangiectasia and there was no family history of HHT or history of chest trauma.

Arterial blood gas in room air presented partial respiratory failure, with a partial pressure of oxygen of 56 mmHg and 86% oxygen saturation. Analysis revealed a normal red cell count, with no sign of anaemia, and a normal liver and kidney function. An ultrasonography of the liver showed no signs of cirrhosis.

A chest-CT scan was performed (Figure 1A,B) and revealed a 3 × 4 cm contrast-enhanced lesion on the right upper lobe, apparently with an afferent vessel with 8 mm diameter and two efferent vessels draining to the right superior pulmonary vein, compatible with pulmonary arteriovenous malformation.

The transesophageal echocardiogram (Figure 1C) revealed an important right–left shunt suggestive of extracardiac shunt, due to the bubbles from the contrast appearing on the left ventricle after five cardiac cycles, thus being compatible with the clinical suspicion of arteriovenous shunt in the pulmonary circulation. The left ventricle ejection fraction was 61%, the cardiac walls had normal thickness and there were no signs of pulmonary hypertension. The grading of the shunt was not performed.

A pulmonary angiography (Figure 1D) confirmed the diagnosis. Selective embolization of the afferent artery was performed by interventional radiology through puncture of the right femoral vein and insertion of a 7Fr introducer sheath and a 5Fr catheter followed by embolization with two amplatzer Plug type II (8 mm each). Resolution of desaturation and dyspnea on exertion was achieved. Given the important relapse rate of the treatment, a Chest-CT was performed and revealed no evidence of recurrence of the PAVM. A six-minute walk test was performed as well with no desaturation. The arterial blood gas in room air revealed a partial pressure of oxygen of 81 mmHg and 96% oxygen saturation. She will maintain regular clinical follow-up and a undergo a Chest-CT every 3 years.

## DISCUSSION

Although the patient didn't present any typical symptoms such as platypnea, epistaxis, telangiectasias or hemoptysis, the chest-CT scan was compatible with the diagnosis of pulmonary arteriovenous malformation. The patient presents only one of the Curacao criteria [6], given the fact that the patient didn't present epistaxis nor telangiectasis and denied family history of HHT, making this diagnosis unlikely. If the diagnosis of HHT is suspected, other sites of possible arterial malformations such as the gastrointestinal tract, brain, skin and liver should be screened. Cirrhosis was ruled out by the normal liver function and by normal abdominal echography. In patients with lung nodules with afferent and efferent vessels visualized on a Chest-CT scan, the diagnosis of PAVM should be suspected and the clinical manifestations should be inquired. Some vascular tumours can share some imagological findings on the Chest-CT scan, but do not present the right-to-left shunt. Taking these facts into account, the patient presented with an idiopathic pulmonary arteriovenous malformation. The transesophageal echocardiogram with the evidence of the right–left shunt suggestive of being extracardiac allowed confirmed the diagnosis. Since the afferent vessel had a diameter bigger than 2 mm (8 mm on the CT) an angiography and embolization was possible, which allowed a successful treatment and prevention of the complications associated with poor outcomes. The patient remains closely monitored due to the relapse risk and will undergo a Chest-CT scan every 3 years. If a new PAVMs appears, a new embolization should be performed, if possible. In some cases, surgical local lung resection could be performed, despite not being the preferred treatment option due to the associated loss of lung parenchyma. The presence of multiple small PAVMs not allowing embolization and hypoxemia should lead to referral for lung transplant consultation.

The patient presented an idiopathic pulmonary arteriovenous malformation, considering that fulfilled only one Curaçao criterion for THH, and didn't have liver or heart disease and didn't have a chest trauma history. The diagnosis was achieved with a transesophageal echocardiogram. The Chest-CT scan revealed an afferent vessel big enough for selective embolization. The patient underwent successful a selective embolic therapy with resolution of symptoms and hypoxemia and prevention of complications associated with poor outcome.

The sex-minute walk test showed no functional limitation, although there is no test previous to treatment for comparison.

### AUTHOR CONTRIBUTIONS

**Pedro Fernandes**: Main author. **Pedro Patrão**: Image selection; overview. **João P. Silva and António Reis**: Review and overview. **Teresa Belo**: Reviewer and language correction.

### ACKNOWLEDGMENTS

This case was presented as an abstract at the 38th Congresso de pneumologia, Algarve, Portugal, 2022.

### DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available on request from the corresponding author. The data are not publicly available due to privacy or ethical restrictions.

### ETHICS STATEMENT

The authors declare that appropriate written informed consent was obtained for the publication of this manuscript and accompanying images.

## ORCID

*Pedro Fernandes* https://orcid.org/0000-0002-3720-2648
*Teresa Belo* https://orcid.org/0009-0005-8687-7624

## REFERENCES

1. McWilliams J. Pulmonary arteriovenous malformations: Clinical features and diagnostic evaluation in adults.
2. Treatable cause of positional dyspnea. Cureus. 2020;12:9052. https://doi.org/10.7759/cureus.9052
3. Fernandes P, Paulo Silva J, Argel M, Guerra S, Ferro R, Nunes R. Pulmonary arteriovenous malformation: case report. Pulmonology. 2022;28:58.
4. Bausewein C, Farquhar M, Booth S, Gysels M, Higginson IJ. Measurement of breathlessness in advanced disease: a systematic review. Respir Med. 2006;101:399–410. https://doi.org/10.1016/j.rmed.2006.07.003
5. McDonald J, Bayrak-Toydemir P, DeMille D, Wooderchak-Donahue W, Whitehead K. Curaçao diagnostic criteria for hereditary hemorrhagic telangiectasia is highly predictive of a pathogenic variant in ENG or ACVRL1 (HHT1 and HHT2). Genet Med. 2020;22:1201–5. https://doi.org/10.1038/s41436-020-0775-8

**How to cite this article:** Fernandes P, Silva JP, Patrão P, Reis A, Belo T. Successful embolization of a clinically significant pulmonary arteriovenous malformation. Respirology Case Reports. 2024;12(4):e01330. https://doi.org/10.1002/rcr2.1330