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PULMONARY ARTERY ANEURYSM ASSOCIATED WITH SARCOIDOSIS IN A 75-YEAR-OLD WITH HEART FAILURE: THE CHALLENGES OF DIAGNOSIS AND MANAGEMENT

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ABSTRACT

Introduction: Pulmonary artery aneurysm (PAA) is a rare abnormality of pulmonary vasculature. It can be idiopathic or secondary to various pathologies, frequently with multiple factors leading to its formation. We report the case of a man with concomitant sarcoidosis and PAA.

Case description: A 75-year-old male with a diagnosis of pulmonary sarcoidosis was referred to the Cardiology department due to heart failure with reduced left ventricular ejection fraction (LVEF). The transthoracic echocardiogram revealed mildly reduced LVEF, aortic root and pulmonary artery (PA) dilatation, and no signs of pulmonary hypertension (PH). Cardiac magnetic resonance imaging was performed, revealing mild left ventricular dilation, LVEF of 40%, main PA dilation (43 mm) and a pattern of late gadolinium enhancement suggestive of cardiac sarcoidosis. At follow-up, a thoracic computed tomography (CT) angiography scan revealed ascending aorta ectasia and giant main PA aneurysm (60 mm). A right heart catheterisation was performed, and a mean PA pressure of 34 mmHg was obtained. Given the clinical context, the patient was considered to have PH due to lung disease and left heart disease, and PAA was possible due to vascular granulomatous involvement by sarcoidosis.

Conclusion: PAA is a rare finding and mostly occurs in the setting of PH. Sarcoidosis is a granulomatous disease that mostly affects the lungs, but the sarcoid involvement of great vessels has been described. In this clinical case, the probable cause for the PA fragility leading to aneurysm formation remains sarcoid vascular infiltration, regarding the discrepancy between the PA dimensions and mildly elevated PA pressure.

KEYWORDS

Pulmonary artery aneurysm, pulmonary hypertension, sarcoidosis

LEARNING POINTS

- Pulmonary artery aneurysm is a rare abnormality of pulmonary vasculature that can be idiopathic or a consequence of pulmonary hypertension, congenital heart disease, infection, vasculitis or collagenopathies.
- Due to the low incidence of this disease, there are no guidelines for its diagnosis, management or follow-up, and treatment is based on the underlying aetiology, aneurysm dimensions and occurrence of symptoms.
- Sarcoidosis is a multisystem disorder of unknown aetiology characterised by non-caseating granulomas that mostly involve the lungs, but can also affect skin, eyes, and lymph nodes. Sarcoid involvement of great vessels has been rarely described.

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INTRODUCTION

Pulmonary artery aneurysm (PAA) is a rare abnormality of pulmonary vasculature[1]. It is defined as a focal dilation of the pulmonary artery (PA) involving all three layers of the arterial wall with dilatation greater than 40 mm[1]. Regarding its aetiology, PAA can be differentiated into congenital causes, acquired causes or causes of idiopathic origin[2]. Sarcoidosis is a systemic disease of unknown aetiology that is characterised by the presence of non-caseating granulomas[3]. The real prevalence of this disease is unknown since many patients present non-specific symptoms or subclinical disease, making it under-recognised in clinical practice[3]. This disease can involve all organ systems to a varying extent and degree[4], and can exceptionally involve large vessels, leading to either stenosis or dilatation[1].

CASE DESCRIPTION

A 75-year-old man in good general condition and a past medical history of pulmonary sarcoidosis was diagnosed 20 years ago and prescribed with deflazacort. The patient was referred to the Cardiology department due to heart failure (HF) with reduced left ventricular ejection fraction (LVEF). The patient was in New York Heart Association (NYHA) functional class II and presented a normal physical examination. Blood analysis showed no relevant findings.

A Holter monitor revealed conduction impairment (first-degree atrioventricular block and complete left bundle branch block). A chest X-ray revealed mediastinal enlargement *(Fig. 1)*. A transthoracic echocardiogram (TTE) revealed mild left ventricle dilatation, mildly reduced LVEF, mild mitral valve regurgitation, mild aortic valve regurgitation, aortic root and ascendant aorta (AA) dilatation (38 and 39 mm, respectively), PA dilatation and no echocardiographic signs of pulmonary hypertension (PH). To study HF aetiology, cardiac magnetic resonance imaging was performed and revealed mildly increased left ventricular volume, mild to moderate reduced LVEF, PA trunk dilation (43 mm), right PA dilatation (29 mm) and a pattern of late gadolinium enhancement suggestive of cardiac sarcoidosis.

The cause of HF was identified as cardiac sarcoidosis in the form of dilated cardiomyopathy. The patient was regularly evaluated in the Cardiology department, maintaining clinical stability. On follow-up, a thoracic computed tomography (CT) scan was requested 2 years later for evaluation of pulmonary parenchyma and dimensions of the main thoracic vessels. It was verified that the AA and PA diameters had increased to 46 and 54 mm, respectively. A new TTE was performed, revealing preserved biventricular systolic function, PA trunk aneurysm with a 54 mm diameter, no major valvular morphological changes, and no echocardiographic signs of PH *(Fig. 2)*. A CT thoracic angiogram was performed and confirmed the previous findings. It revealed AA dilation (42 mm), PA trunk aneurysm (60 mm), and right and left PA dilatation (31 mm and 30 mm, respectively) *(Fig. 3, 4 and 5)*. An 18F-fluorodeoxyglucose (FDG) positron emission tomography showed no significantly increased

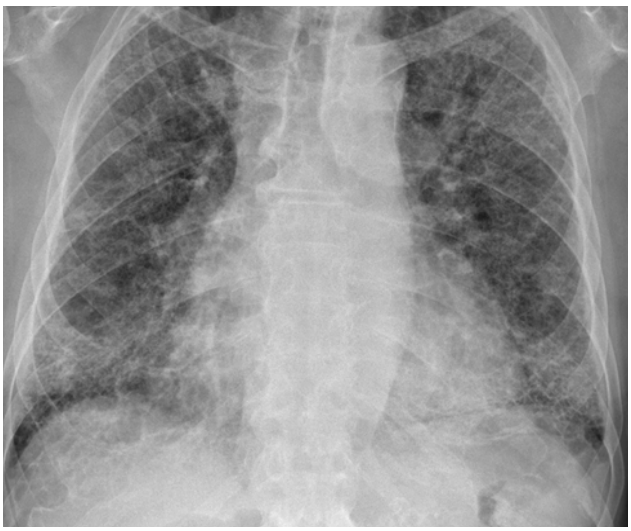

Figure 1. Posteroanterior chest radiograph showing hilar enlargement suggestive of main thoracic vessel dilation.

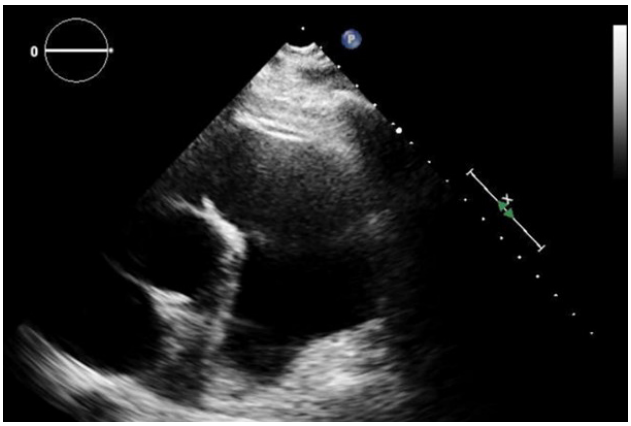

Figure 2. Transthoracic echocardiography parasternal short-axis view showing pulmonary artery trunk aneurysm (54mm).

capture of FDG in the PA. The treponema pallidum particle agglutination test was negative, and the interferon gamma release assay was also negative. Pulmonary function tests revealed a reduced diffusing capacity of the lungs for carbon monoxide (<20%; normal is >75% of predicted). The patient was initially prescribed with methotrexate and posteriorly azathioprine, but these drugs were withdraw due to intolerance. A right heart catheterisation revealed a mean pulmonary arterial pressure of 34 mmHg and no reliable measure of pulmonary arterial wedge pressure, due to inadequacy of wedging the PA catheter. Taking in account the previous findings, the diagnosis of PH due to lung disease and left heart disease, and PAA possibly due to vascular granulomatous involvement by sarcoidosis, was made. A conservative approach has been chosen regarding the high surgical risk of the patient. The patient kept his previous medication and has been stable in HF NYHA functional class II, without symptoms suggestive of complications.

DISCUSSION

PAA is defined as an excessive dilatation involving all three layers of the pulmonary artery wall or any of its main branches[5]. A cut-off point to differentiate PAA and PA

ectasia is defined as a PA diameter superior to 40 mm[5]. PAA is a rare finding and is infrequently diagnosed[2,5]. As such, there are no specific guidelines about its management[1]. PAA can be classified as congenital (particularly due to patent ductus arteriosus, ventricular septal defect and atrial septal defect[6]), idiopathic or secondary to a specific underlying disease such as PH, infection (syphilis, tuberculosis), connective tissue disorders (Marfan's syndrome, Ehlers-Danlos syndrome) and vasculitis (Behçet's disease)[2,5]. Sarcoidosis is a multisystem disorder characterised by non-caseating granulomas in involved tissues[7]. This disease mostly affects lungs in 90% of cases, but other organs such as skin, eyes and lymph nodes can also be involved[7]. The evolvement of great vessels, such as PA, has been rarely described[8]. Frequently, the clinical manifestations of PAA are non-specific[2] and include dyspnoea, chest pain, hoarseness, palpitation and syncopal episodes[2].

Conversely, patients can also be asymptomatic even in the presence of large PAAs with diameters up to 70 mm[2]. The best management of a PAA remains uncertain. Since it is an infrequent disease with various possible underlying causes and limited experience in its management, there are no clear guidelines for the best therapeutic approach[2]. First, it is of major importance to assess accompanying diseases or presence of anatomical anomalies, because for PAA secondary to a specific disease such as tuberculosis, syphilis, Behçet's disease or vasculitis, conservative treatment should be attempted first[5]. It is considered that non-giant idiopathic PAAs can be treated conservatively with β-blockers and diuretics, but a very close monitoring regarding the presence of symptoms and PAA dimension is required.

This can be assessed by CT scan or TTE. Surgery is recommended to reduce symptoms and diminish the risks of fatal complications when the PAA presents a diameter >55 mm (especially in cases of diameter >70 mm), PAAs associated with PH or anatomical anomalies, PAAs presenting a rapid growth (defined as ≥5 mm increase in the diameter of the aneurysm in 6 months) or if the PAA causes compression of a critical structure or causes symptoms[5,6]. The surgical approach can be aneurysm repair and replacement with allogeneic/synthetic grafts[5]. Concerning the present clinical case, giving the low likelihood of other diseases with vascular involvement (such as syphilis or Behçet's syndrome), and considering the clinical context the most probable cause for the PA fragility leading to aneurysm remains sarcoid vascular infiltration.

This is supported by a finding of mildly elevated pulmonary artery pressure that does not entirely justify or provide a substantial contribution to the PAA dimensions. As previously described, our patient presented an indication for surgical treatment. However, this was withdrawn due to the high surgical risk (EuroSCORE II 7.17%), considering that the patient presented a stage IV sarcoidosis with extensive pulmonary fibrosis and unfavourable respiratory evolution, with a need of 24h oxygen therapy during follow-up. Besides, the presence of PH (even if mild), was also associated with an increased surgical risk. A conservative approach was further supported by the PAA dimensions remaining stable during follow-up. In conclusion, sarcoidosis can exceptionally involve large vessels, leading to either stenosis or dilatation.

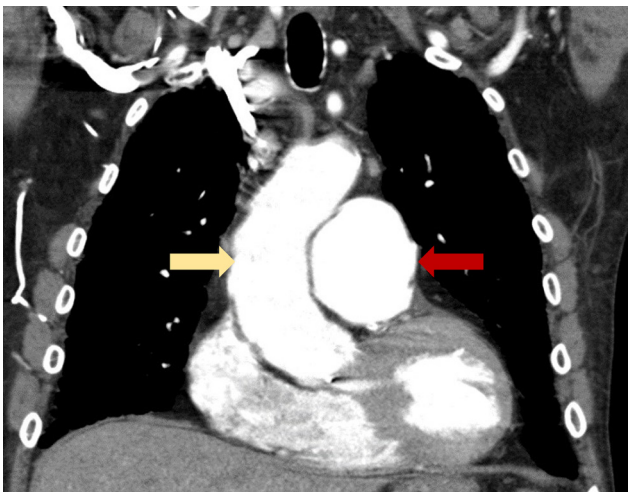

Figure 3. Thoracic computed tomography angiography (coronal view) showing Aao (yellow arrow) and PA aneurysm (red arrow).

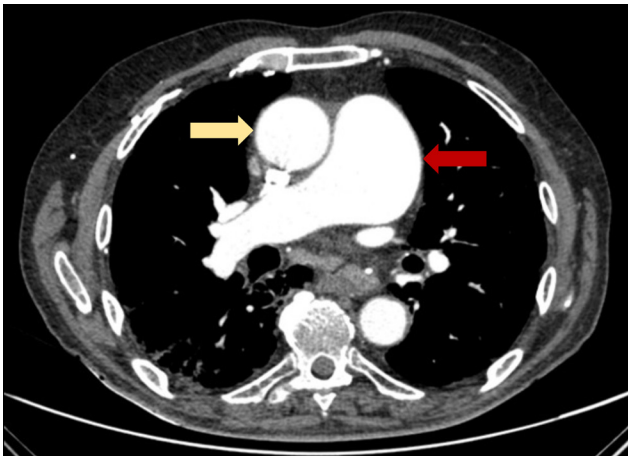

Figure 4. Thoracic computed tomography angiography (axial view) showing Aao (yellow arrow) and main PA aneurysm (red arrow).

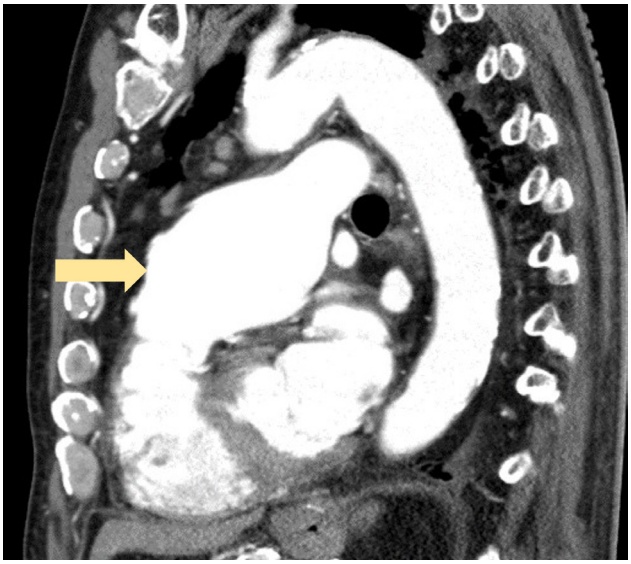

Figure 5. Thoracic computed tomography angiography (sagittal view) main PA aneurysm (yellow arrow).

In the present clinical case, the probable cause for the PA fragility leading to aneurysm formation remains sarcoid vascular infiltration, regarding the discrepancy between the PA dimensions and mildly elevated PA pressure.

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