CASE REPORT

Extreme dilation of pulmonary artery: a literature review

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SUMMARY

Massive pulmonary dilation is a rare finding. These aneurysms can result from different aetiologies that can be congenital or acquired. We present a case of a 61-year-old Caucasian woman who presented with dyspnoea and cough and was discovered to have a massive pulmonary aneurysm. After eliminating most other possible causes, pulmonary hypertension and congenital pulmonic valve stenosis were deemed likely aetiological factors for our patient. These aneurysms are very rare and should prompt close attention and surveillance when present. It is essential to diagnose these findings in a timely fashion as severe complications such as arterial perforation and fatal haemorrhage may occur if left untreated. We present this case because mechanisms leading to pulmonary artery dilation are poorly understood. Furthermore the clinical symptoms and complications are not commonly known, meriting case presentation.

BACKGROUND

Pulmonary artery (PA) aneurysms are rare and their mechanisms and causes are not commonly known. However, some of the known aetiological factors are potentially treatable and should be thoroughly investigated.¹ Prompt recognition of these findings on routine radiographic imaging may help prevent complications such as coronary artery and airway compression.^{1 2} We present this case to discuss the literature regarding the pathogenesis, clinical presentation, complications and causes of PA aneurysm.

CASE PRESENTATION

This is a case of a 61-year-old Caucasian woman with a medical history significant for chronic obstructive pulmonary disease (COPD), pulmonary hypertension, a moderate-to-severe PA dilation of 4.6 cm and congenital mild pulmonic stenosis. She presented to the emergency room with shortness of breath, a cough, wheezing and bilateral leg swelling for the previous 2 days. She mentioned that her cough was dry and had been going on for 2 months. The patient had an echocardiogram in 2010 that showed maintained ejection fraction (EF). She had a subsequent transesophageal echocardiogram that ruled out atrial septal defect so this problem was never followed up on. Her physical examination was significant with a pulse rate of 101 and oxygen saturation at 80% on room air, which improved to 93% with 5 L of oxygen. A neck examination revealed significant jugular venous distension. Respiratory examination revealed bilateral basilar crackles but no wheezing. A holosystolic murmur

Figure 1 Chest X-ray.

was auscultated at her right lower sternal border. The patient's ECG was unremarkable for any ischaemic changes. Her blood work showed an elevated brain natriuretic peptide (BNP) level of 1201. Troponin levels were mildly elevated at 0.06 ng/mL. She was admitted with a diagnosis of acute congestive heart failure (CHF) and COPD exacerbation. She was administered intravenous furosemide and started on albuterol and ipratropium nebulisers. Oxygen therapy was continued. Subsequently, her jugular venous distension and pedal oedema improved and her BNP level came down to 366. However, the patient still continued to show symptoms of shortness of breath. A chest X-ray (figure 1) showed cardiomegaly and leftward mediastinal enlargement suggestive of PA dilation. A CT of the chest was performed (figure 2) and revealed no evidence of acute pulmonary embolism. However, there was a markedly dilated main PA of 6.7 cm (figure 3).

Figure 2 Chest CT (axial view).

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Figure 3 Chest CT (coronal view).

A transthoracic echocardiogram (TTE) was performed which showed a preserved EF of 51% but a severely elevated PA systolic pressure of 62 mm Hg. There was moderate pulmonic valve regurgitation with thickening and mild doming of the pulmonic valve but no significant stenosis (figure 4). The patient's right ventricle was enlarged and showed severely reduced global systolic function. The findings of the CT scan of predominant severe dilation of the main PA (6.7 cm) and the left PA were confirmed. Cardiothoracic surgery was consulted but due to comorbidities and the high risks of surgical intervention, only medical management was recommended. Thus, the patient followed up with the pulmonary hypertension clinic and was started on conservative management.

INVESTIGATIONS

- A chest X-ray is a simple non-invasive diagnostic modality that should be utilised in the first instance when pulmonary system pathology is suspected. Attention should be focused on any mediastinal enlargement.

Figure 4 Echocardiogram.

- A CT of the chest should also be performed to confirm the findings of the chest X-ray and determine the dimensions of the aneurysm.
- A TTE should be performed to measure the PA pressures and diagnose PA hypertension.
- Ideally, a right heart catheterisation for accurate PA pressures and a vasoreactivity test to guide therapy for pulmonary hypertension should both be performed. In our patient they were not performed due to the risk of aneurysm perforation.

DIFFERENTIAL DIAGNOSIS

- Aortic artery aneurysm
- PA tumour
- Cystic medionecrosis
- Trauma

TREATMENT

Usually, surgery is the preferred treatment for PA aneurysm, but in some cases a conservative approach is favoured due to the surgical risks. Determining the cause of the dilation is therefore essential and treatment should be tailored accordingly. In our case, the patient was found to not be a viable surgical candidate due to extensive COPD, severe pulmonary hypertension and right ventricle failure. She was treated for her comorbidities with inhalers and nasal oxygen.

OUTCOME AND FOLLOW-UP

The patient was not operated on due to the high risk of surgery resulting from her advanced COPD. Thus, she was referred to a pulmonary hypertension clinic. A right heart catheterisation was also not performed due to the risk of aneurysm perforation. Despite these restrictions, when treated with oxygen and short-acting and long-acting β -agonist inhalers, the patient's symptoms improved. However, due to presence of her aneurysm, the patient remained at a high risk for rupture.

DISCUSSION

When a PA is dilated extensively to surpass 40 mm—it is defined as an aneurysm.¹ PA aneurysms are a rare occurrence.

The mechanisms of PA aneurysms are poorly understood and their causes, clinical symptoms and complications are not widely known. It has been reported that PA dilations can occur from different aetiologies. They can be congenital due to improperly divided truncus arteriosus, left to right atrial and ventricular septal defects and from arterial malformations in cases such as Marfan's syndrome. Congenital anomalies such as stenosis, absent valve, bicuspid or quadricuspid pulmonary valves may also cause PA dilation. Pulmonary dilation may also be acquired from primary pulmonary hypertension, mitral valve stenosis, pulmonary embolism, trauma and rupture of the aortic aneurysm into the pulmonary artery.² In our patient, after excluding most of the aforementioned causes, congenital pulmonic valve stenosis was thought to be the likely dominant aetiological factor. Pulmonary valvular stenosis has been shown to cause dilation on the basis of turbulent blood flow at the distal end of the pulmonic stenosis.³ This mechanism of post-stenotic dilation is not clear. However, it is postulated that due to the stenosis there is a drop in pressure and increase in velocity of the blood across the obstructive lesion. This has an effect on the downstream vessel causing it to dilate (Venturi Forces). Animal experiments and experiments involving elastic rubber tubes have shown that pulmonary dilation can be reproduced downstream to pulmonic valvular stenotic lesions.^{4 5} Any cause leading to PA dilation in massive proportions may eventually lead to poor PA blood flow and increased collateral circulation. In our patient the gradient across the pulmonic valve was normal so pulmonary hypertension was deemed the dominant cause for right ventricular dysfunction.⁶ Owing to these cardiopulmonary effects, clinical patients with these aneurysms may present with cough, decreased tolerance to activity and shortness of breath. The patient may also present with generalised weakness, chest pain and haemoptysis.⁷ Owing to the localised dilation of the pulmonary vessel, the surrounding structures may get compressed and both the left main coronary artery compression and left main bronchus compression have been shown to occur in rare cases.⁸ However, these aneurysms may also be picked up incidentally on routine imaging and emphasise the importance of paying due diligence to radiographic findings. It is essential to diagnose these findings in a timely fashion, as it may lead to severe complications such as arterial perforation and fatal haemorrhage. Pulmonary emboli and heart failure are also feared consequences.⁹

Invasive testing to get accurate measurements of valve gradients and PA pressures is essential to guide therapy. Surgery as a

treatment is usually recommended due to the risk of rupture. However, a conservative approach may have to be followed, as in our case, when there is a high risk from the surgery.¹⁰

Learning points

- Congenital anomalies such as pulmonic valve stenosis should be monitored as they may lead to complications of post-stenotic pulmonary dilation.
- Prompt recognition of this finding on routine radiographic imaging might help prevent complications such as main coronary artery and airway compression.
- Treatable causes of pulmonary artery dilation should be identified and treated when possible.

Competing interests None.

Patient consent Obtained.

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