

Left main coronary artery compression by a large pulmonary artery aneurysm in the absence of pulmonary hypertension: a case report

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Background

Pulmonary artery aneurysms (PAAs) are rare and they are infrequently diagnosed due to the non-specificity of their symptoms. However, their related complications, mainly described in patients with pulmonary hypertension (PH), are associated with significant morbidity and mortality.

Case summary

We report the case of a 64-year-old woman previously operated on for pulmonary valve stenosis, who presented with rapid worsening of dyspnoea and sudden onset of chest pain. Physical examination did not show heart failure symptoms, and an echocardiogram showed significant but not severe pulmonary regurgitation with preserved right and left ventricular function. Estimated pulmonary artery (PA) pressure was normal. As myocardial ischaemia was suspected the patient underwent a coronary computed tomography angiography that showed compression of the left main coronary artery by a large PAA. Early diagnosis led to surgery that solved her symptoms.

Discussion

Comprehensive medical evaluation of symptomatic patients with PA dilatation, even in the absence of PH, is key to rule out the possibility of serious complications as soon as possible.

Keywords

Case report • Pulmonary artery aneurysm • Congenital heart disease • Pulmonary valve stenosis • Left main coronary artery

Learning points

- Pulmonary artery aneurysms (PAAs) are an infrequent condition, mainly caused by pulmonary hypertension (PH) and congenital heart defects (CHDs). Although their overall prognosis is good, complications, mainly in the presence of PH, are associated with significant morbidity and mortality.
- Extrinsic compression of the left main coronary artery, for instance, is a rare but potentially fatal complication of PAAs. Physicians treating patients with CHDs should be aware of this infrequent complication, even in the absence of PH.

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Introduction

Pulmonary artery aneurysms (PAAs) are defined as a focal dilatation of all three pulmonary artery (PA) wall layers. Deterling and Clagett¹ reported an estimated prevalence of one in 13 696 postmortem examinations. Pulmonary hypertension (PH) and congenital heart defects (CHDs) are the main causative mechanisms. Less frequent causes are connective tissue abnormalities, inflammatory or infectious diseases, and iatrogenic and idiopathic PAAs.² Clinical manifestations of PAAs are diverse, and although their overall prognosis is good, complications are associated with significant morbidity and mortality.³ Extrinsic compression of the left main coronary artery (LMCA), for instance, has been associated with malignant ventricular arrhythmias and sudden death. We report a rare case of LMCA compression by a low pressure PAA, where early diagnosis avoided a possible fatal outcome.

Timeline

Time	Events
Initial evaluation	<p>Patient referred following rapid progression of exertional dyspnoea and recent onset of effort-related chest pain.</p> <p>Chest X-ray showed signs of dilated right heart and a prominent main pulmonary artery (PA).</p> <p>Transthoracic echocardiogram showed moderate right ventricle and atrial dilatation with normal right ventricular (RV) systolic function, significant pulmonary regurgitation, normal RV outflow gradient, normal estimated RV systolic pressure, and a normal-sized inferior vena cava. No RV restrictive physiology.</p>
1 month	<p>Cardiopulmonary stress testing showed frequent exercise-induced ventricular premature beats and poor functional capacity.</p> <p>Holter monitor showed a short run of non-sustained ventricular tachycardia.</p>
2 months	<p>Coronary computed tomography angiography (CCTA) confirmed extrinsic compression of the proximal segment of the left main coronary artery (LMCA) caused by an enlarged PA. Admission to hospital for surgical pulmonary artery aneurysm (PAA) repair along with pulmonary valve replacement.</p>
Follow-up (15 days)	<p>A second CCTA, showed the PAA had decreased in size and LMCA compression had disappeared.</p>
Follow-up (4 months)	<p>Exercise stress test showed functional capacity had improved, and no ventricular premature beats were observed.</p>
Outpatient clinic (9 years)	<p>The patient has remained asymptomatic and functional capacity is good.</p>

Case report

A 64-year-old woman who had been diagnosed with pulmonary valve stenosis (PVS) in childhood underwent open surgical commissurotomy and infundibulectomy at the age of 35. Significant residual pulmonary regurgitation (PR) was documented at follow-up. After remaining asymptomatic for years, she was referred to our centre following rapid progression of exertional dyspnoea and recent onset of effort-related chest pain.

Physical examination revealed good general condition, mild hypertension (147/81 mmHg), heart rate of 80 b.p.m., and oxygen saturation of 98%. There were no signs of heart failure and the cardiovascular examination showed a short, low-pitched diastolic murmur over the left upper sternal border.

An electrocardiogram showed sinus rhythm, right bundle branch block, and frequent supraventricular premature complexes. A chest X-ray revealed signs of dilated right heart and a prominent PA (Figure 1).

Our differential diagnosis included progressive PR with right ventricular (RV) failure, RV restrictive physiology with diastolic dysfunction, exercise desaturation in the presence of a patent foramen ovale, progressive PH with RV failure, and LMCA or compression of the left mainstem bronchus by PA dilatation.

To evaluate these potential diagnoses, we first performed an echocardiogram that showed moderate right ventricle and atrial dilatation with normal RV systolic function (TAPSE of 19 mm and fractional area change of 43%), significant PR with diastolic retrograde flow from the main PA, pressure half-time of 209 ms and a PR index (ratio of PR duration/diastole duration) of 80%, normal RV outflow gradient (see [Supplementary material online, Videos S1 and S2](#)), mild tricuspid regurgitation with an estimated RV systolic pressure of 34 mmHg, and a normal-sized inferior vena cava. No evidence of RV restrictive physiology was observed.

We then performed cardiopulmonary exercise testing. The patient did not develop any symptoms and oxygen saturation remained constant throughout the test, but functional capacity was poor, achieving 1.2 METS. Age-predicted maximal heart rate attained was 65%, and percent-predicted peak oxygen consumption was 23%. During the test, she developed frequent exercise-induced ventricular premature beats. Consequently, we performed a 24-h Holter monitor, which showed frequent atrial and ventricular premature beats and a short run of non-sustained ventricular tachycardia (NSVT) (Figure 2).

We next performed a coronary computed tomography angiography (CCTA) to evaluate potential mechanical complications caused by PA dilatation. No compression of the left main bronchus was seen. Coronary artery anomalies and significant coronary artery disease were ruled out. However, the CCTA showed extrinsic compression of the proximal segment of LMCA caused by an enlarged PA (60 mm diameter in systole) (Figure 3A–C), and a takeoff angle of the LMCA of 26° (defined by a line parallel to the border of the left coronary sinus and a line parallel to the LMCA) (Figure 3D). A coronary angiogram confirmed the diagnosis and the right-sided heart catheterization showed normal PA pressure (see [Supplementary material online, Video S3](#)).

The patient underwent surgical PAA repair along with pulmonary valve replacement with a conduit constructed intraoperatively composed of a 25 mm bioprosthetic Carpentier-Edwards valve within a

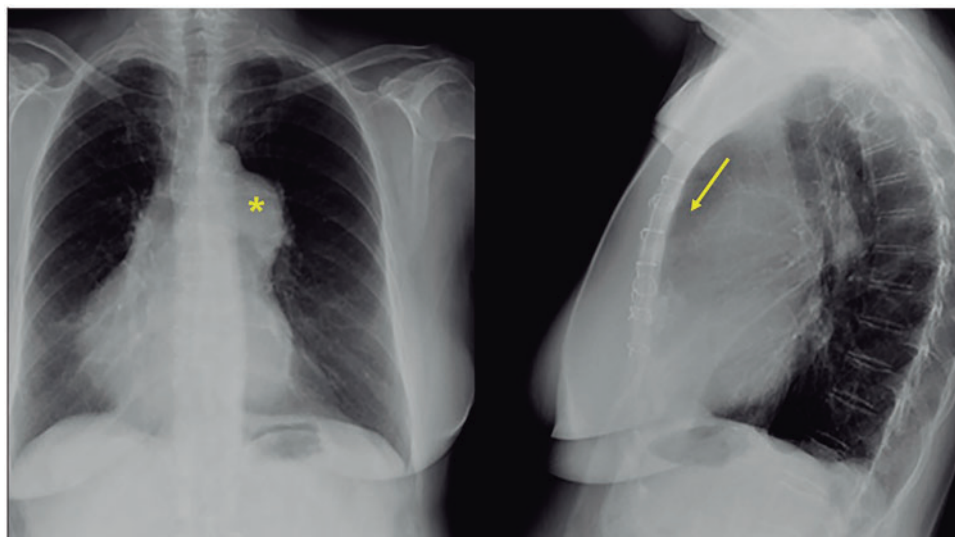


Figure 1 Posteroanterior chest X-ray showing a dextropositioned heart with mesocardia and enlargement of the right atrium and right ventricle with a dilated pulmonary trunk (asterisk). In the lateral view, filling of the retrosternal airspace (arrow) is suggestive of right ventricular dilatation.

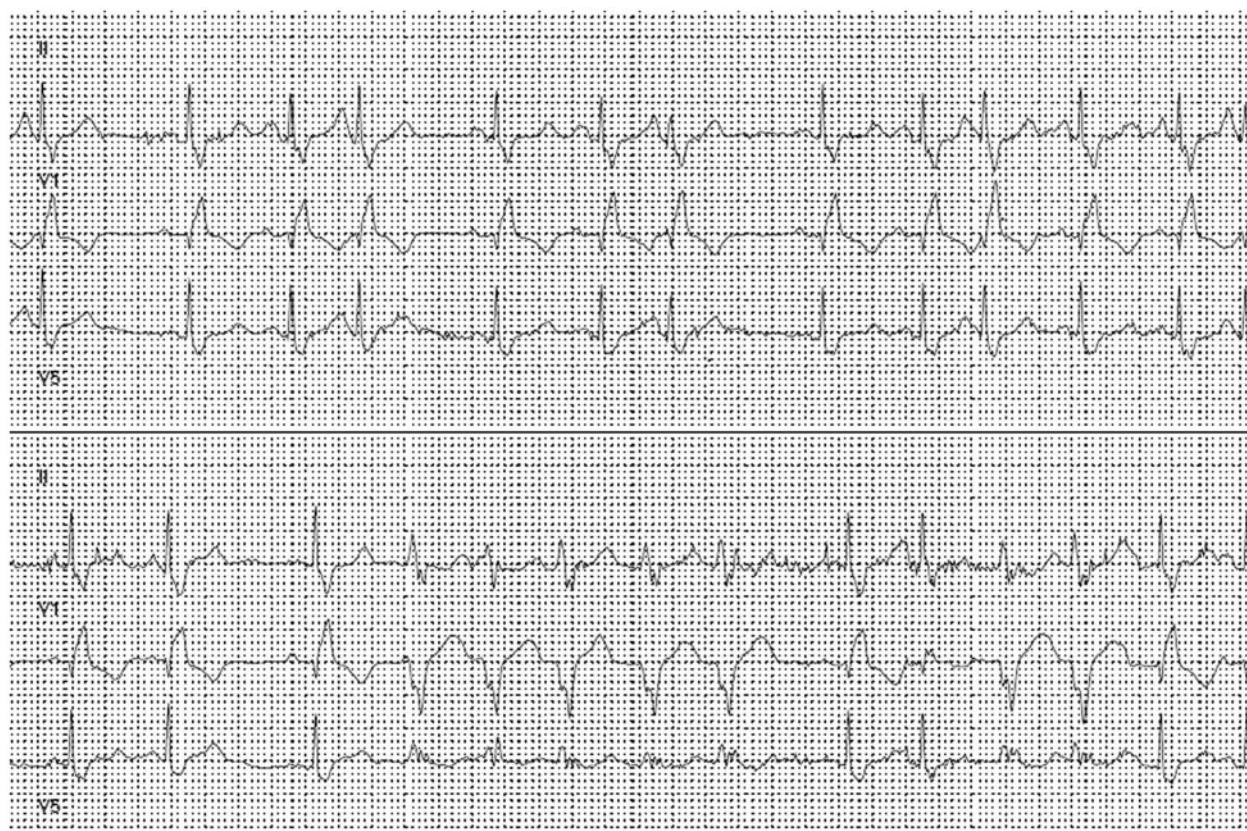


Figure 2 A 24-h Holter electrocardiogram with atrial and ventricular premature beats and a non-sustained ventricular tachycardia.

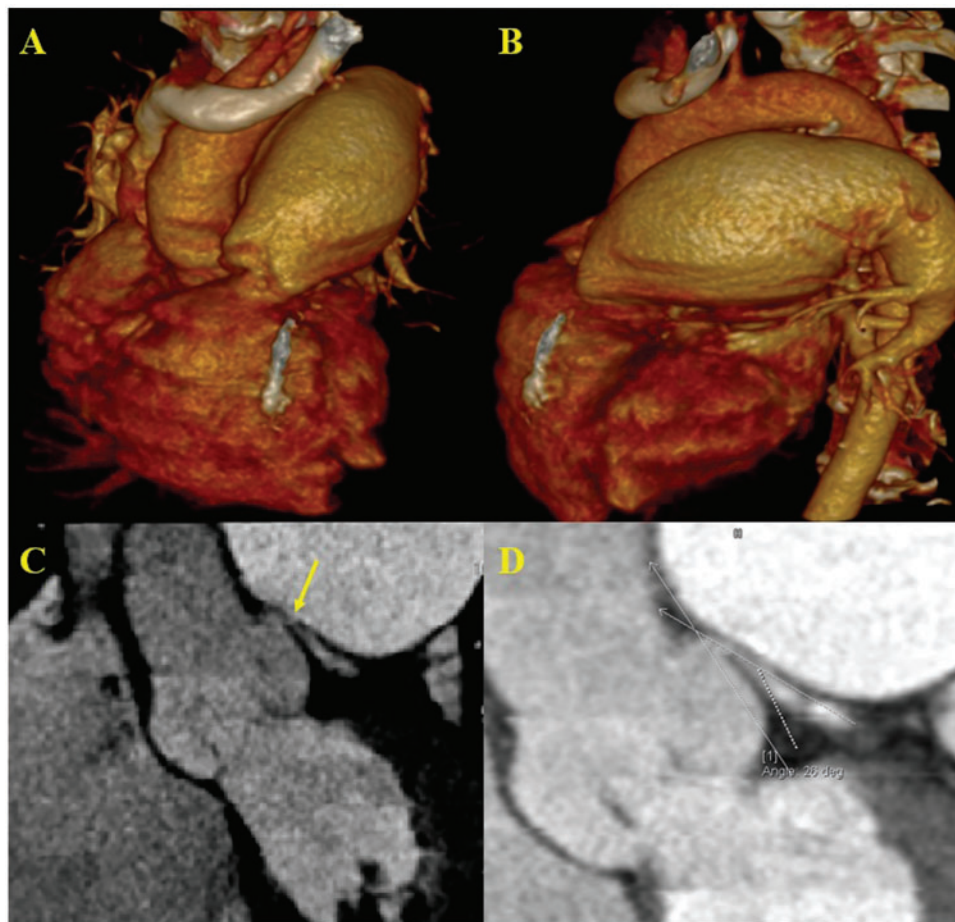


Figure 3 Coronary computed tomography angiography showing severe enlargement of the main pulmonary artery (A and B) and extrinsic compression of the proximal segment of the left main coronary artery (arrow) with a takeoff angle of 26° (C and D).

Dacron tube graft. As complete decompression of LMCA was visually documented intraoperatively, coronary revascularization surgery was unnecessary. Intraoperative findings showed a thickened, trileaflet pulmonary valve, and the anatomopathological examination of the PA wall showed changes suggestive of cystic medial necrosis.

A second CCTA, performed 2 weeks after surgery, showed the PAA had decreased in size (Figure 4A and B) and LMCA compression had disappeared (Figure 4C). An exercise stress test performed 4 months after surgery showed functional capacity had improved, and no ventricular premature beats were observed. Over a 9-year follow-up, the patient has remained asymptomatic and functional capacity is good.

Discussion

Pulmonary valve stenosis has been associated with post-stenotic dilatation of PA independently of the severity of the stenosis.⁴ Dilatation may be due to intrinsic weakness of the arterial wall in association with haemodynamic shear stress, especially if valvular regurgitation is present. In this setting, cystic medial necrosis is a common finding in

samples of the PAA vascular wall, as in our case. Such findings are similar to those described in patients with bicuspid aortic valve and aortic root dilatation.⁵

Thrombosis, dissection, rupture and compression of adjacent structures are potentially fatal PAAs complications. Very few cases of low pressure PAA complications have been reported to date.⁶

Extrinsic compression of the LMCA is an unusual PAA complication that may present with ventricular ischaemia, malignant arrhythmias, or sudden death.^{7,8} A severe PA dilatation has been described as a predisposing risk factor for LMCA compression in patients with PH.⁹ In our case, however, coronary flow insufficiency occurred in the setting of a low pressure PAA, and to our knowledge, only two previous cases have been reported in this context.^{10,11} In the absence of PH, LMCA obstruction can take place due to its anatomical relation with PA. In a study that evaluated angiographic determinants of LMCA extrinsic compression, Kajita et al. found that invasive angiography revealed inferior displacement of the LMCA caused by a dilated PA, with a mean LMCA takeoff angle of 23° compared with an angle of 70° in the control group. The authors suggested that the origin of the LMCA from the right sinus of Valsalva or a rightward-positioned left coronary sinus indicated a higher risk of compression.¹²

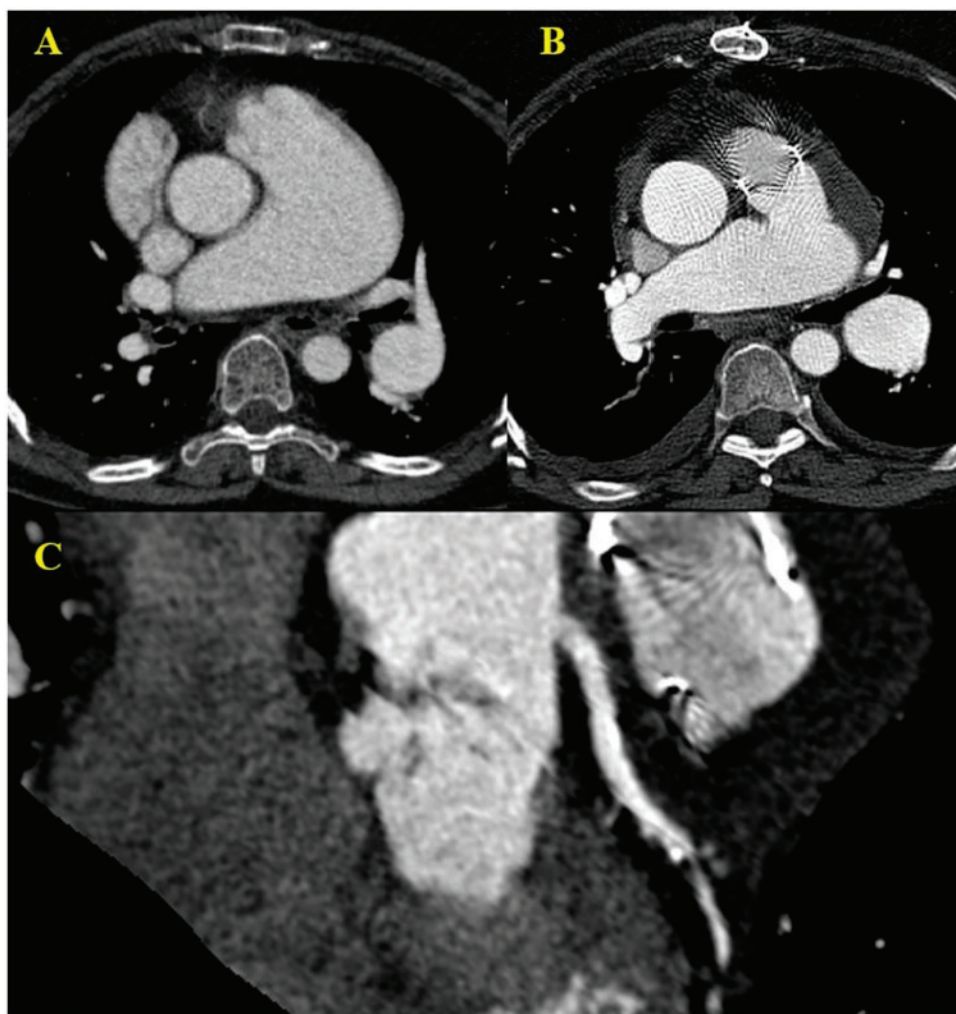


Figure 4 Coronary computed tomography angiography showing a decrease in the size of the pulmonary artery (A and B) and resolution of the left main coronary artery compression after surgery (C).

Our case has some unique aspects. First, although the patient clinically presented with chest pain suggestive of angina, no objective evidence of myocardial ischaemia was seen in the stress test. Second, we attributed the NSVT with left bundle branch block morphology to left ventricular septum ischaemia rather than to a RV origin. And third, the uncommon presence of LMCA compression was due to a low pressure PAA.

There are currently no guidelines on the management of PAAs. However, surgery remains the cornerstone of therapy for PA trunk lesions. A recent review suggests surgical intervention should be considered if the absolute size of a PAA is 55 mm or greater, if the rate of dilatation is greater than 5 mm in 6 months, if symptoms appear, if valvulopathies or shunts that require surgery coexist, and if PH, compression of adjacent structures, thrombosis, dissection, or rupture are documented.¹³

Coronary revascularization may not be necessary if compression of LMCA resolves after aneurysm surgery, as in our case. In high-risk

patients, percutaneous coronary revascularization has also been described as an option.¹⁴

In conclusion PAAs are a rarely diagnosed entity that may lead to life-threatening complications. We report an unusual case of extrinsic compression of the LMCA by a low pressure PAA due to PVS. Physicians treating patients with CHD should be aware of this infrequent but potentially fatal complication.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as [Supplementary data](#).

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and

associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: none declared.

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