

Pulmonary artery aneurysm associated with severe degenerative aortic stenosis

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ABSTRACT

We report a pulmonary artery (PA) aneurysm associated with severe aortic stenosis and an aortic root dilatation occurring in a 59-year-old woman who presented with dyspnoea and chest pain. PA aneurysms are rare, and there are no definitive guidelines on its management. There are contentious opinions on whether such aneurysms should be managed conservatively or surgically. Our patient had associated aortic stenosis and underwent a successful aortic valve replacement and PA aneurysm repair. This case illustrates that concomitant PA repair with other cardiothoracic surgery can be performed safely, even in patients with moderate surgical risks. We also discuss the natural history, prognosis and management of PA aneurysms.

Keywords: aortic stenosis, aortic valve replacement, pulmonary artery aneurysm

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INTRODUCTION

Pulmonary artery (PA) aneurysm is rare. It has been associated with structural cardiac and vascular abnormality, pulmonary hypertension, infection and vasculitis.^(1,2) The presentation is usually nonspecific, although haemoptysis and PA dissection have been reported. Isolated idiopathic PA has been thought to be due to an inherent weakness of the arterial wall, and such a giant aneurysm runs the risks of dissection and rupture.^(3,4) There are no definitive guidelines on the management of this type of aneurysm due to the paucity of data regarding the long-term outcome following surgical or medical interventions. We report a case of PA aneurysm associated with calcific aortic stenosis and ascending aortic root dilatation, treated successfully with surgical therapy.

CASE REPORT

A 58-year-old hypertensive Malay woman with type 2 diabetes mellitus presented with a six-month history of progressive effort-related dyspnoea. This was associated with occasional chest pain on severe exertion. Her

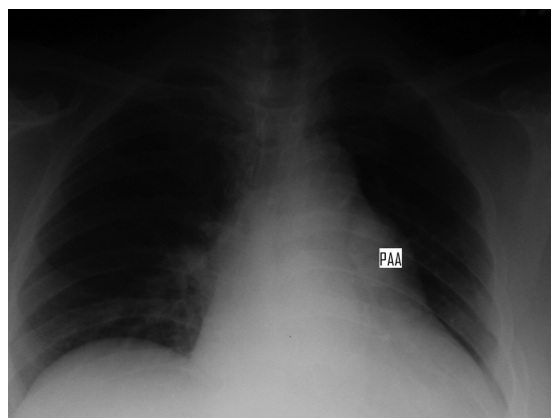


Fig. 1 Posteroanterior chest radiograph shows main pulmonary artery enlargements.



Fig. 2 Pulmonary angiogram shows a large isolated main pulmonary artery aneurysm.

medication included ticlopidine 250 mg bd, atenolol 50 mg od, perindopril 4 mg od, chlorothiazide 250 mg od, metformin 500 mg tds and lovastatin 20 mg. Clinically, she was obese, with a body mass index of 37 kg/m². Her blood pressure was 150/90 mmHg and the heart rate 70/min with a sinus rhythm. There was a harsh ejection systolic murmur over the aortic region, radiating to the neck. The apex beat was not displaced. The jugular venous pressure was normal, and there was no right ventricular heave. There were no stigmata of connective tissue diseases.

Laboratory investigations did not show any significant abnormalities. Her full blood count, serum urea and electrolyte tests, and liver function test were normal. Her cholesterol level was 3.5 mmol/L (low-density lipoprotein

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Fig. 3 CT of the pulmonary artery shows a large main pulmonary artery aneurysm measuring 4.7 cm.

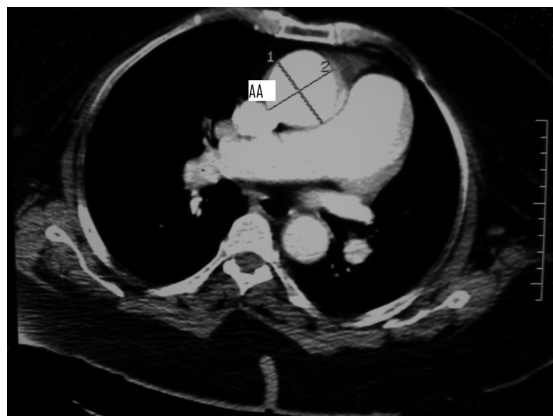


Fig. 4 CT angiogram shows a dilated ascending aorta measuring 4.5 cm in diameter.

2.0 mmol/L; triglyceride 1.4 mmol/L), random glucose 7.1 mmol/L, erythrocyte sedimentation rate 20 mm/1st hour and her autoantibodies ANA, dsDNA and VDRL were negative. Electrocardiography showed a normal sinus rhythm with left ventricular hypertrophy on Sokolow Lyon voltage criteria. Chest radiograph revealed cardiomegaly with dilated pulmonary arteries (Fig. 1). Two-dimensional echocardiography showed severe calcific trileaflet aortic valve stenosis (measured peak pressure gradient 90 mmHg; mean 60 mmHg; aortic valve area 1.0 cm²), concentric left ventricular hypertrophy (interventricular septum 1.7 cm), good left ventricular systolic function with an ejection fraction of 65% (based on Simpson's rule) and a significant diastolic dysfunction (mitral valve E/A ratio 0.8; IVRT 100 ms; DT 280 ms). There was a large main PA measuring approximately 4.5 cm. The right heart chambers appeared mildly dilated with an estimated pulmonary systolic pressure of 40 mmHg. The ascending aorta was also dilated, measuring approximately 4 cm.

Coronary angiography revealed a single vessel mid-left anterior descending artery discrete stenosis of 70%. The pullback gradient across the aortic valve was 50 mmHg. The aortogram showed a post-stenotic dilatation of the ascending aorta measuring 4.5 cm. Right heart catheterisation did not show any intracardiac shunt. There was no pulmonary stenosis. The PA pressure was measured at 55/26 mmHg. Pulmonary angiogram confirmed a large isolated main PA aneurysm measured at 4.8 cm (Fig. 2). High-resolution computed tomography (CT) measurements were as follows: main PA 4.7 cm; right PA 2.7 cm; left PA 2.5 cm; ascending aorta 4.5 cm × 4.5 cm; aortic arch 2.8 cm; and descending aorta 3 cm × 3 cm. There was no evidence of pulmonary embolism or thrombosis (Figs. 3 & 4).

The patient underwent successful surgery. She had a bioprosthesis aortic valve replacement, a left internal

mammary artery graft to the left anterior descending artery. The main PA was repaired with a 30-mm Dacron graft replacement, following the resection of the aneurysm. The pulmonary valve was competent and not replaced. The ascending aorta was left alone, as it was felt that the mortality and morbidity of the surgery would be significantly increased if concomitant repair was performed. The histological specimen of the resected aneurysm showed medial degeneration and fragmentation of the elastic fibres. There was no loss of smooth muscle or inflammation. On follow-up, the patient was well. Her breathlessness had improved, and she was in New York Heart Association Class I. Echocardiography showed a well-functioning aortic valve and intact grafts.

DISCUSSION

The size of a PA aneurysm was more than 4 cm in diameter. An autopsy report in 1947 showed that proximal PA aneurysm was found in only eight of 109,571 cases (0.0073%), although the precise incidence is unknown, due to the lack of prospective studies.⁽³⁾ The aetiologies have included infection (syphilis, in the past), pulmonary hypertension, medial degeneration of the PA, arteriosclerosis, valvular heart disease, vasculitis or related connective tissue disease.^(1,4) Pulmonary hypertension was found in 66% of such cases.⁽⁵⁾ Hughes-Stovin syndrome, an association of multiple PA aneurysms, pulmonary embolism and vascular thrombosis, has been associated with such giant aneurysms.⁽⁶⁾ This syndrome, initially thought to be an incomplete Behçet's disease, is a cause of vascular aneurysms and needs to be managed with aggressive immunosuppression.^(6,7) Dennison et al also demonstrated a case of simultaneous aortic and pulmonary aneurysm due to giant cell arteritis.⁽⁸⁾ PA aneurysms are usually visible on chest radiographs as an enlarging, round or fusiform mass, although CT or

magnetic resonance (MR) imaging would be required for further delineation and measurement. PA aneurysm itself produces no symptoms unless there are complications, such as a compression on the bronchus, dissection/rupture or thrombus formation.

There is no consensus on the management of PA aneurysm. It has been postulated that low pressure PA, in the absence of congenital left-to-right shunts or Eisenmenger's syndrome, has been associated with a very low risk of rupture, despite its large size. Patients are more likely to present with symptoms of pulmonary regurgitation. Small case series have reported survival of patients with a median age of 52 years without evidence of media dissection or intimal tear, and hence careful medical observation might be appropriate for these patients.^(9,10) Deterling and Clagett, however, have reported progression to rupture in the setting of severe pulmonary hypertension or congenital left-to-right shunting.⁽³⁾ Laplace's law dictates that wall tension is directly proportional to the intravascular pressure and radius of the vessel, and is inversely related to the wall thickness. Hence, progressive enlargement of the aneurysms on follow-up indicates higher wall stress and an increased risk of rupture, necessitating surgical correction.^(3,9)

There was no evidence of any inheritable connective tissue diseases or vasculitis in our patient. We postulate that the aetiology of the aneurysm could be due to the intrinsic weakness of the arterial wall, compounded by the increased haemodynamic shear stresses due to pulmonary hypertension related to her calcific aortic stenosis. The same principle applies to the cause of the ascending aorta dilatation. The abnormality could lie in the process of extracellular matrix remodelling in the aortic wall, including inadequate synthesis, degradation and transportation of extracellular matrix proteins.⁽³⁾ Surgical techniques to repair pulmonary trunk aneurysm have included aneurysmorrhaphy, Dacron graft replacement and pulmonary allograft replacement.⁽¹¹⁾ Our patient's PA was associated with moderate pulmonary hypertension (PA pressure 55 mmHg). Concomitant repair of the aneurysm appeared to be a strategically sound approach when the patient required heart surgery for severe calcific aortic stenosis. The ascending aortic root was not replaced as consensus was root replacement is advised only if

dilatation is more than 4.5 cm in diameter during aortic valve surgery.⁽¹²⁾

Although PA aneurysm is rare, it can occur in association with other structural cardiac and vascular abnormalities. Patients undergoing cardiac (especially valvular) surgery, should initially be screened carefully with a routine chest radiograph. Subsequent detailed examination with transthoracic echocardiogram and CT of the PA would be essential, if any such abnormality exists. Isolated low-pressure PA aneurysm can be managed conservatively, but surgical therapy should be contemplated in the presence of severe pulmonary hypertension, left-to-right cardiac shunt or valvular heart disease.

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