

Case Report

Idiopathic Pulmonary Artery Aneurysm

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Abstract

Idiopathic pulmonary artery aneurysm (PAA) is a rare lesion. Clinical experience with this condition is limited and current knowledge is mainly derived from autopsy findings. We report a patient who came to us with complaints of chest pain, breathlessness on exertion and pedal oedema and was diagnosed to have PAA.

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Key words: Pulmonary artery aneurysm, Parahilar opacity.

Introduction

First described by Bristowe in 1860 in a case at necropsy,¹ and subsequently described by Deterling and Claggett in 1947,² pulmonary artery aneurysm (PAA) is an infrequent disease of the pulmonary vasculature, most frequently found post-mortem.³ The possible causes are myriad and diverse in pathophysiology. Patients with post-stenotic dilatation of the main pulmonary artery usually present fairly late with insidious cardio-respiratory symptoms. Diagnosis requires radiological imaging assistance as the clinical findings are non-specific.⁴ We describe a patient who was diagnosed to have PAA, a diagnosis rarely made ante-mortem.

Case Report

A 52-year-old man presented to our outpatient department with complaints of chest pain, breathlessness and swelling on feet and face for the last one month. The chest pain was mild in nature but aggravating with activity. Breathlessness was progressive, aggravating on exertion and accompanied with orthopnoea and paroxysmal nocturnal dyspnoea. Swelling which started from feet was progressive and later on there was swelling on the face also. There was no history of smoking. He had no other significant preceding medical history.

He was dyspnoeic and physical examination revealed anaemia, raised jugular venous pressure (JVP). His heart rate was 100 beats per minute, irregular in rhythm; blood pressure was 160/100 mmHg, respirations 16 breaths per minute. A loud systolic murmur was heard along the lateral border of sternum in the second intercostal space on the left side. Auscultation of the lungs revealed bilateral basal crepitations. Laboratory evaluation revealed haemoglobin 9 g/dL; total and differential leucocyte

counts and serum lipid profile were within normal limits. Blood culture for pyogenic organisms did not yield any result. Venereal Disease Research Laboratory (VDRL) test for syphilis was also negative. The chest radiograph revealed a left parahilar opacity (Figure 1). Electrocardiogram (ECG) was suggestive of right bundle branch block (RBBB) with right axis deviation and right ventricular hypertrophy.

Two-dimensional (2D) transthoracic echocardiography demonstrated dilatation of left ventricle (LV), right atrium and right ventricle with LV systolic dysfunction (ejection fraction of 28%) with moderate mitral regurgitation, moderate tricuspid regurgitation, moderate aortic regurgitation and normal pulmonary valve function, global LV hypokinesia, inter-ventricular and posterior wall discordance seen with normal pericardium. There were no signs of intracardiac shunt, pulmonic stenosis. Contrast-enhanced computed tomography (CECT) of thorax showed a

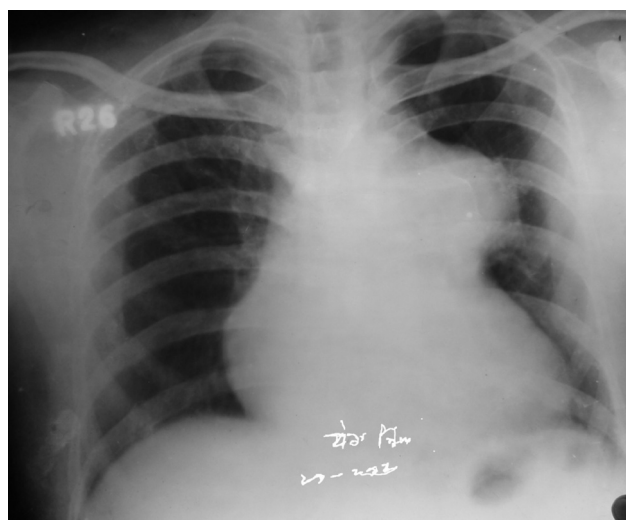


Figure 1. Chest radiograph (postero-anterior view) showing a parahilar opacity on left side.

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grossly dilated (maximum diameter 5.95 cm) pulmonary artery (Figure 2) with dilated right atrium. There was no evidence of any calcification and thrombosis. Inferior vena cava (IVC) and hepatic veins were prominent with retrograde filling of IVC. The rest of the mediastinal structures were within normal limits.

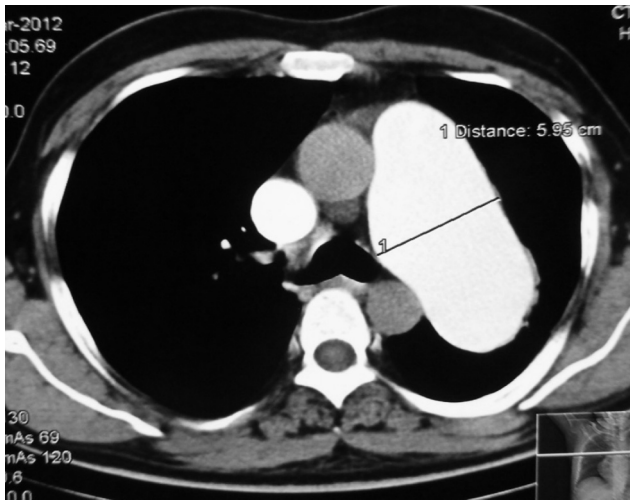


Figure 2. Contrast enhanced computed tomography of chest showing dilated pulmonary artery.

Further investigations did not show any disorder that could lead to the aneurysmal dilatation of the pulmonary artery. Thus, a diagnosis of idiopathic aneurysm of the main pulmonary artery, ischaemic heart disease was made.

Patient refused to go to a higher centre for opinion regarding surgery. As per the recommendation of cardiologist, he was started on beta-blockers and diuretics. Gradually there was a marked improvement in his general condition; breathlessness and pedal oedema significantly decreased. He is doing well on regular follow-up.

Discussion

In contrast to aneurysms in the vessels of the systemic circulation, true PAAs are rare. Predisposing conditions include congenital and acquired heart disease, e.g. left-to-right shunting or pulmonary valve stenosis with post-stenotic dilatation. Other causes include infection (tuberculosis, syphilis, osteomyelitis, pneumonia), systemic vasculitis (Hughes-Stovin's disease, Behcet's disease), Collagen vascular diseases, connective tissue disorders (Marfan's syndrome, Ehler's-Danlos syndrome), trauma (direct or blunt chest injury), mucoid vasculopathic changes and idiopathic PAA.⁴ Idiopathic PAAs are rare anomalies with poorly understood pathogenesis. The precise incidence of the disease is unknown; and the approximate incidence is estimated to be 1 in 14,000 autopsies.² Whether PAA are more common among patients of a particular gender has not been ascertained.⁵ A true aneurysm is defined by dilatation

of all three layers of the vessel wall. The lesion involves the pulmonary trunk and may also extend to the main branches and the peripheral pulmonary arteries. It seems that intrinsic weaknesses of the arterial wall in combination with increased haemodynamic stress are responsible for its formation.³ Clinical presentation of a PAA varies. It can be asymptomatic, being a chance finding on a chest radiograph, whereas dyspnoea, haemoptysis and chest pain are the most frequently encountered symptoms in patients of PAA. If the patient has an incompetent pulmonary valve, then right heart failure may be the initial manifestation.⁶ These aneurysms can grow to an impressive size before identification as evident in our case where the diameter was observed to be 5.95 cm.

Certain criteria have been established for the specific diagnosis of idiopathic dilatation of the pulmonary artery. According to these criteria, idiopathic PAA is defined as diameter of pulmonary trunk of greater than 30 mm in the absence of a cardiac or pulmonary cause for pulmonary artery dilatation and in the presence of normal pulmonary artery pressure.⁷

As in the case under report, PAA usually presents as a parahilar opacity on the chest radiograph, which is likely to be misinterpreted and misdiagnosed as hilar or mediastinal mass. Further radiologic evaluation is necessary to make the correct diagnosis. Although pulmonary angiography is the gold standard for establishing the diagnosis, but newer non-invasive imaging methods, such as CECT angiography and magnetic resonance imaging (MRI) have simplified the diagnosis.⁸ Owing to its high spatial resolution, CECT is considered to be the primary technique for diagnosing PAA since it offers a unique opportunity to evaluate the presence, size, shape and exact location of the aneurysm, and concomitant cardiovascular abnormalities. The aneurysms appear as saccular or fusiform areas of dilatation of various sizes.⁹ MRI is also a very useful non-invasive imaging modality, especially in the possible detection of possible intimal flap, it may also show the arterial wall thickening in connective tissue disease, and provide information regarding blood flow direction in cases of post-stenotic dilatation due to disease involving the pulmonary valve. In our case the diagnosis was established on CECT.

The natural history of proximal PAA is poorly understood because of the limited number of cases diagnosed ante-mortem; occurrence of dissection has often been documented.¹⁰ However, not all aneurysms progress to the rupture stage. Patients of PAA without significant pulmonary hypertension have been reported to have had a uncomplicated course of 1-7 years.¹¹

The management of PAA is not clearly established.¹² In the current surgical era, some authors recommend surgical intervention when the pathology is diagnosed.² However, the role of surgery in main PAA is still not well defined.¹³ Several different surgical procedures are reported as treatments for PAAs, such

as, aneurysmorrhaphy, aneurysmectomy and repair with dacron graft, or autologous pericardial replacement.¹⁴ However, conservative treatment is still advocated by some, when there is no left-to-right intracardiac shunt or significant pulmonary artery hypertension² and in idiopathic cases.¹⁵ Furthermore, the management of such cases requires individualisation, according to the primary cause, whereas long-term clinical and radiological follow-up is necessary, taking into consideration the potentially fatal complications.¹⁶

Physicians should consider the diagnosis of PAA in patients presenting with chest pain and dyspnoea on exertion along with a parahilar opacity on a chest radiograph and obtain a correct diagnosis by either angiography or other non-invasive diagnostic modalities. It is essential to recognise, diagnose and treat this entity at the earliest because of its tendency to rupture leading to increased mortality.

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