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# Idiopathic Low Pressure Pulmonary Artery Aneurysm Presenting as Pulmonary Embolism

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**Abstract:**

Aneurysms of the pulmonary artery are very rare with an estimated incidence of less than 1 in 14,000 cases based on autopsy results. Most cases are associated with other congenital malformations, primary/ secondary pulmonary hypertension, vasculitis, infectious agents, collagen vascular disorders, cystic medial necrosis or local trauma. Idiopathic pulmonary artery aneurysm is an extremely rare entity with very few cases reported in the literature. Our reported case of idiopathic pulmonary aneurysm complicated by pulmonary embolism highlights the need for a consensus management of such a case.

**Key words:** Pulmonary aneurysm, Idiopathic, Pulmonary embolism.

## Introduction

Pulmonary Artery Aneurysm (PAA) is a rare condition with an estimated incidence of less than 1 in 14,000 cases based on autopsy results [1]. Idiopathic pulmonary artery aneurysm is a rare clinical entity, and therefore the natural course and clinical management are not well established. The clinical manifestation of PAA depends on its size and location. Non specific symptoms associated with PAA include hemoptysis, exertional dyspnea, fever, cough and chest pain. Low-pressure PAA might be a source of recurrent emboli because of stasis and endothelial dysfunction [2]. We report the case of a 77 year old female who presented with sudden onset dyspnea due to pulmonary artery embolism.

## Case Report

An ambulatory 77 year female resident of Delhi sustained fracture neck of right femur due to fall. She presented to us with sudden onset breathlessness after two days. Our patient was previously healthy female with no history of unconsciousness, swelling over leg, chest pain, palpitations, haemoptysis, cough, fever, oral or genital ulcers, cyanosis or recurrent respiratory tract infections. There was no history suggestive of tuberculosis, hypertension, diabetes and hyper-coagulation state. Examination revealed, a conscious oriented female with tachycardia and tachypnea. There was no evidence of cyanosis, clubbing, genital or oral ulcers, petechae, skin rash or marfanoid features. She had normal BMI and there was no evidence of deep vein thrombosis. Rest of the general physical and systemic examinations were unremarkable.

Complete blood count including platelet count, fasting blood sugar, coagulation profile, liver and kidney function tests and urine examination were within normal limits. Arterial blood gas analysis showed $PaO_2$ of 62 mm Hg,


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$pCO_2$ 45 mm Hg with oxygen saturation of 92%. Chest X Ray was normal and ECG showed sinus tachycardia. Cardiac enzyme (CK-MB) and troponin T were within normal range. D-dimer was highly raised with value 1778 microgram/L. 2-D Echocardiography revealed normal pulmonary artery pressure of 16 mm Hg at rest with normal sized atria and ventricles, no evidence of clot and normal valves. CT pulmonary angiography showed aneurysmal dilatation of left and right branches of main pulmonary artery measuring 32 mm and 30 mm respectively while the main pulmonary trunk was normal. There was right sided subsegmental upper lobe resolving infarcts of lung without any evidence of pulmonary artery hypertension. Doppler ultrasound of bilateral lower limb veins, pelvic veins and inferior vena cava were normal. Ultrasound of the abdomen and pelvis did not reveal any abnormality with no evidence of mass lesion or cysts in kidney and liver. No evidence of fat globules was seen on urine and sputum examination along with normal reports of ANA, c-ANCA, p-ANCA, VDRL, Mantoux test and ESR. Fundus and slit lamp examination of the eye were normal.

After initiation of therapy with anticoagulants, our patient showed signs of improvement and was discharged with the advice for surgical intervention for repair of PAA.

## Discussion

Pulmonary artery aneurysm is a rare entity. PAA can be congenital or acquired. Most cases are associated with other congenital malformations, primary/secondary pulmonary hypertension, vasculitides, infective etiology, collagen vascular disorders, cystic medial necrosis or local trauma. Idiopathic pulmonary artery aneurysm is very rare entity, and therefore the natural course and clinical management are not well established.

The clinical manifestation of PAA depends on its size and location. Generally, the symptoms are non specific including hemoptysis, exertional dyspnea, fever or cough, and chest pain. Chest radiographs are generally unremarkable but may reveal pulmonary artery dilatation. The diagnostic modalities include echocardiography (transthoracic and transesophageal), and MR or CT angiography. Pulmonary angiography is the gold standard method to diagnose PAA. However, CT angiography is an alternate non-invasive investigation which is highly sensitive and specific [3]. She had highly elevated D-dimer, normal 2-D echocardiography which in the background of her clinical presentation required further investigation for embolism. CT pulmonary angiography showed aneurysmal dilation of left and right branches of main pulmonary artery measuring 32 mm and 30 mm respectively while the main pulmonary trunk was normal. There was right sided sub segmental upper lobe resolving infarcts of lung without any evidence of pulmonary artery hypertension. However, search for cause of pulmonary embolism was negative. So it was assumed that the embolus might have arisen from the pulmonary aneurysm itself. On reviewing the published data, there is some data regarding the association between low-pressure PAA and the generation of emboli. Low-pressure PAA might be a source of recurrent emboli because of stasis and endothelial dysfunction [2]. Low pressure aneurysm of pulmonary artery remains asymptomatic and is incidentally detected on autopsy or rarely present with embolic event such as in the present case.

Idiopathic PAA is a benign condition, so the diagnosis is often made on autopsy. PAA may be associated with primary/secondary pulmonary hypertension, which poses a high risk of dissection and rupture, while low-pressure PAA's seem to be more benign but occasionally leads to embolisation [4]. It seems that intrinsic weaknesses of the arterial wall in combination with stasis of blood and increased hemodynamic stress are responsible for the formation of PAA. The treatment of PAA is still not clear. Few authors recommend a conservative approach in

asymptomatic or idiopathic PAA patients [5] while others recommend a surgical repair if its size is greater than 6 cm. If the patient is symptomatic, surgery is recommended irrespective of the size due to increased risk of rupture or dissection and embolisation in these patients.

## Conclusion

The finding of low pressure idiopathic pulmonary artery aneurysm complicated by embolisation is extremely rare in clinical practice. Our individual experience with such case is an effort to enrich the literature and aid clinicians in getting acquainted with this very rare condition.

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