

FATAL PULMONARY ARTERY DISSECTION — COMPLICATION OF PATENT DUCTUS ARTERIOSUS

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ABSTRACT

Pulmonary artery (PA) dissection is a rare but fatal pathology. Diagnosis is rarely made during life as false lumen tends to rupture resulting in cardiac tamponade and sudden death. Congenital heart diseases like Patent Ductus Arteriosus (PDA) can lead to pulmonary hypertension which is the main cause of PA dissection. A 35 years old male presented to us with acute right sided chest pain and dyspnoea. Patient was diagnosed with some congenital heart disease in childhood but was not treated. CT Angiography confirmed main pulmonary artery dissection which was extending to right pulmonary artery. While in the ward, the patient deteriorated, went into shock, resuscitated, shifted to operation room. Median sternotomy was done, cardiopulmonary bypass established, patent ductus arteriosus closed and PA repair using T-graft done. But the patient did not come off cardiopulmonary bypass and died. Pulmonary artery dissection is an extremely rare condition but mostly fatal. The main cause is chronic pulmonary hypertension either primary or secondary to congenital heart diseases, though there are other causes as well like right heart endocarditis, amyloidosis, trauma, severe atherosclerosis etc. The progression and natural course is not known as there are very few cases reported in literature. Optimum management for Pulmonary artery dissection has to be defined because of its association with congenital heart diseases, so that prompt measures can be taken to save precious lives.

KEY WORDS: Pulmonary artery, Patent Ductus Arteriosus, Pulmonary Hypertension, Dissection.

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INTRODUCTION

The first dissecting aneurysm of pulmonary artery (PA) was reported by Walshe in 1862.¹ To date 64 cases of PA dissection have been reported.²⁻⁴ It is usually a complication of chronic pulmonary hypertension.⁵⁻⁸ The common underlying causes are congenital heart diseases, Idiopathic PA hypertension, and, chronic inflammation of PA, right heart endocarditis, amyloidosis and severe atherosclerosis.⁹ The patient was diagnosed with a Patent Ductus Arteriosus (PDA) in early childhood but was not treated. The case is rare as the dissection was not only extending from the main pulmonary artery (MPA) to the right pulmonary artery (RPA) but also to the segmental branches.

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CASE PRESENTATION

A 35 years old Afghan male diagnosed with some congenital heart disease in early childhood, presented to the Emergency room (ER) of Rehman Medical Institute with right sided chest pain and dyspnoea of recent onset. He took pain killers which were prescribed by the local General Practitioner (GP) and that relieved the chest pain to some extent. He also narrated that he had one episode of exertional dyspnoea (Grade II) 10 years back for which he sought medical advice. He was prescribed tab. Digoxin which he took for 6 months. The medical record is not available.

On examination the patient was in no distress, was not cyanosed with a blood pressure of 130/80, pulse rate of 80/minute and a respiratory rate of 16/minute. His oxygen saturation was normal at room air. Systemic examination was unremarkable. His initial investigation included complete blood count, renal function tests, Liver function tests, ECG and cardiac enzymes. All of these investigations were normal. Chest x-ray showed grossly enlarged cardiac

shadow. MPA and its branches were dilated. An area of consolidation is also seen in right middle zone with raised right hemidiaphragm. (Fig. 1)

Echocardiography revealed: dilated MPA (75mm), with RPA thrombosis/dissection. Right ventricular systolic dysfunction, pulmonary hypertension, small PDA, no pericardial effusion, Ejection fraction of 59%.

CT scan Chest showed marked dilation of MPA and its principal branches. MPA = 75mm, RPA



Figure 1: X-rays chest showing dilatation of main pulmonary artery and its branches.

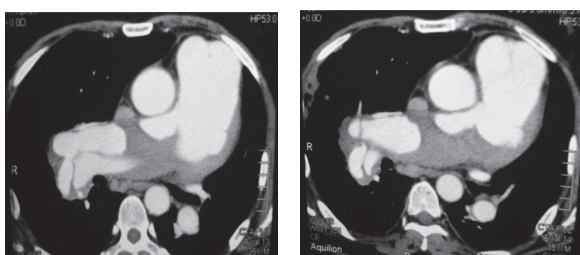


Figure 2: CT scan chest showing false lumen in Main Pulmonary Artery secondary to dissection.

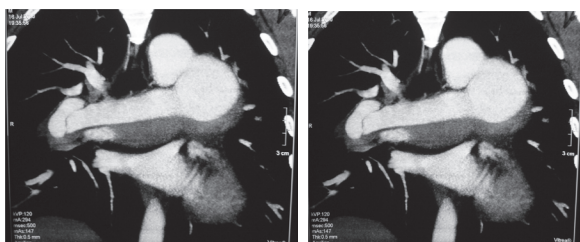


Figure 3: CT Angiography showing dissection of the main pulmonary artery and its branches.

= 45mm, Left pulmonary artery (LPA) = 35mm. A thin line of non-enhancing portion of lumen is noted along left lateral aspect of MPA and along superior aspect of RPA, representing false lumen secondary to dissection, upper lobe consolidation, and mild cardiomegaly. (Fig. 2) CT Angiography of Pulmonary Artery: Pulmonary artery dilatation, thrombosis and dissection in the RPA. Segmental infarction of upper lobe of right lung, PDA. (Fig. 3)

He was managed using beta blockers, nitrates and aspirin. Suddenly the patient deteriorated with systolic blood pressure of 86mmHg, and diastolic of 60mmHg with O₂ saturation of 67-72% at room air. Resuscitation was started and was immediately shifted to I.C.U. Arterial line was put in and invasive monitoring started. Patient was intubated and put on ventilator. His condition deteriorated further and Cardiopulmonary resuscitation (CPR) was started. He was shifted to operation room for emergency repair. Median sternotomy was done. Aortic and bi-caval venous cannulation done. Cardiopulmonary bypass (CPB) established, systemic hypothermia of 28C° achieved, aorta cross clamped and intermittent, antegrade cold blood cardioplegia into aortic root.

Operative findings: Severely dilated MPA, RPA and LPA. Main pulmonary artery was opened. Big PDA was found. Dissection in the MPA extending in to the origin of LPA and in to the whole length of RPA and its segmental branches was seen which was, all full of blood clots. Clots were sucked, MPA, RPA and LPA were washed. Patent ductus arteriosus was closed from within using pledgeted 5/0 prolene sutures and T-shaped interposition graft between MPA and RPA and LPA sewn in place. Patient rewarmed, but he could not come off CPB and died because of global hypoxia and myocardial damage.

DISCUSSION

Pulmonary dissection though rare but fatal complication of chronic pulmonary hypertension. The false lumen in PA dissection tends to rupture, unlike aortic dissection, rather than developing a re-entry site, resulting in rupture and cardiac tamponade and sudden death. Most of the cases are diagnosed post mortem. Dissection of PA always occurs at the site of pulmonary dilatation or aneurysm. Although the pathophysiological cause of dissection in pulmonary artery is not clear. Pulmonary hypertension and resultant mucoid degeneration of media and fragmentation of elastic fibers strongly predispose to this condition.¹⁰ Symptoms of pulmonary artery dissection are non specific, and symptoms include dyspnoea occurring in 82%, chest pain in 67% and

central cyanosis in 52%.⁹ Non-invasive imaging methods used to detect pulmonary artery dissection are transthoracic echocardiogram^{11,12}, CT scanning,⁹ MRI¹³ and pulmonary arteriography.¹⁴ CT particularly multidetector CT may be used if echocardiography failed to detect an intimal flap.^{15,16}

The commonest cause of PA dissection is congenital heart diseases¹⁷, in addition to other rare causes like Right heart Endocarditis, chronic inflammation of pulmonary arteries, amyloidosis, trauma secondary to cardiac intervention like PA angiography and severe atherosclerosis have been implicated. There are instances where there was no cause of pulmonary dissection. Over the past two centuries only 64 cases of PA dissection have been reported of whom 09 were diagnosed during life.¹⁷ We are reporting a case of PA dissection due to Pulmonary artery hypertension secondary to PDA.

Among 64 previously reported cases of PA dissection, 34 had underlying cardiac disease, most commonly congenital heart diseases including PDA and 07 had rheumatic mitral stenosis. Ten patients with idiopathic PA hypertension with dissection have been reported. Two of these developed dissection post lung transplant or heart lung transplantation.¹⁸ There were 03 cases that have been attributed to cardiac intervention^{19,16,10}. One was due to Marfan's syndrome.²⁰ Over all sex distribution appear to be equal with wide age range from 26-85 years. Peak incidence of dissection was in 3rd and 6th decades. In younger patients congenital heart diseases were the most common underlying cause, whereas in older patients, diseases were associated with PA dissection.¹⁷ The main PA trunk is the site of dissection in almost 80% cases, usually without the involvement of the branches. Occasionally an isolated dissection of the right or left PA's and intrapulmonary branches can occur^{5,6,20,21} In small proportion of patients PA dissection may occur at the site of localized aneurysm formation.

The optimum management of patient with PA dissection has not yet been defined because of low number of cases in literature. Senbakkavaci et al reported surgical treatment of PA dissection using T shaped prosthetic graft replacement of MPA as emergency surgery to prevent lethal bleeding. We used the same technique of interposition T-graft. Out of the reported patients those have presented emergently and were diagnosed in timely manner, 03 patients had successful intervention.^{9,21}

CONCLUSION

Congenital heart diseases like PDA which lead to pulmonary hypertension can result in dissection

and rupture subsequently. So the congenital defects like PDA should not be ignored. Diagnosis of PA dissection should be suspected in any patient with pulmonary hypertension who present with chest pain and timely diagnosis and intervention can save precious lives.

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CONFLICT OF INTEREST

Authors declare no conflict of interest.

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None declared.