Case Report

A rare case report of pulmonary artery dissecting aneurysm

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Abstract

Pulmonary artery dissection is a rare event and usually occurs as a complication of chronic severe primary or secondary pulmonary arterial hypertension, trauma, vascular abnormality or chronic inflammation however Idiopathic pulmonary artery dissection is extremely rare and usually not diagnosed during life as it leads to sudden death. We report a rare case of dissecting pulmonary aneurysm in a very young patient without obvious cause. This case was unusual that the patient was hemodynamically stable despite symptoms of chest pain and mild dyspnea since 15 days. We describe Chest radiograph, Echocardiography and CT findings in a patient with dissecting pulmonary aneurysm.
Keywords: Aneurysm, Dissection, Pulmonary arterial hypertension.

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INTRODUCTION

Pulmonary artery aneurysm with dissection is rare and usually occurs secondary to pulmonary hypertension, pulmonary artery trauma or vascular anomaly however Idiopathic dissecting pulmonary aneurysm is extremely rare and to best of our knowledge and literature search, there have been not more than 100 cases reported, which Includes idiopathic as well as non-idiopathic dissecting pulmonary aneurysm. PA dissection usually manifests as cardiogenic shock or sudden death and is therefore typically diagnosed at postmortem rather than during life[1,2]. There are sporadic reports of patients (less than 10 cases of idiopathic and non-idiopathic) dissecting pulmonary aneurysm in the literature who diagnosed during life. We report a case of Idiopathic Dissecting Pulmonary Aneurysm diagnosed by CT scan in a patient who was hemodynamically stable and no obvious cause for it.

CASE SUMMARY

Twenty two years old man, thin built, who presented with vague chest pain, shortness of breath since 15 days, No past history of any cardiac intervention, No other significant past history. On clinical examination – afebrile, pulse – 80 / Min., BP – 110/70 mm Hg, RR – 18 / minute, no cyanosis, no pallor, no s/o CHF, no marphanoid features. Investigation - Normal CBC, ESR, KFT, LFT, Serological test for syphilis, HBsAg and HIV were negative, LE cell phenomenon- negative. Cardiac enzymes to exclude coronary syndrome were normal. Chest X ray PA view (Fig. 1) revealed a large homogenous opacity seen in left hilar and parahilar region occupying left mid and lower zones without air-bronchogram. Transthoracic echocardiography (Figure 2a and b) - a paraternal long-axis view of the left heart showed -dilated Aortic root -3.9 cm, a short-axis view of the heart showed dilated main pulmonary artery - 4.7 cm, right pulmonary artery - 2.4 cm, left pulmonary artery - 1.7 cm and there was a rent in MPA of posterior-lateral wall with large aneurysm with blood flowing in and out of aneurysm (Figure 2a and b) Doppler imaging showed mild pulmonary regurgitation and an estimated pulmonary artery diastolic pressure was only 6 mm Hg. Apical 4 chamber view of the left heart showed normal size right atrium and right ventricle, LV dominance, No e/o of TR, good biventricular function. There was e/o moderate pericardial effusion, more on posterior side. Echo did not show any other congenital, rheumatic or other valvar heart disease; there was no evidence of

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vegetation. HRCT Chest and CT Pulmonary Angiography (Fig.3)– revealed a very large pulmonary artery aneurysm measuring up to 8cms involving the main pulmonary artery. There was long segment linear hypodensity within the main pulmonary artery involving trunk of main pulmonary artery s/o intimal flap extending up to 4cms within the aneurysm. The flap was not extending in to either of main branches of pulmonary artery, however the proximal pulmonary arteries were dilated with pruning of the peripheral branches. The ascending aorta, aortic arch and descending thoracic aorta were mildly dilated measuring up to 3.5 cms. There was moderate pericardial effusion.

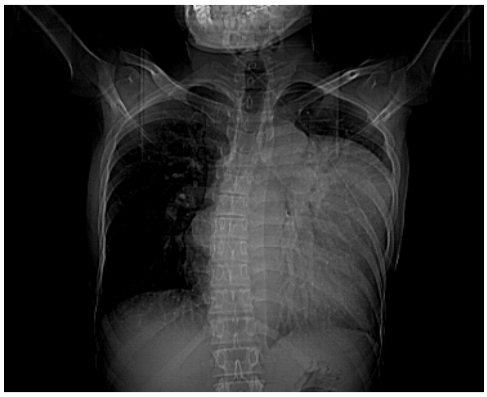

Figure 1: Frontal chest radiograph shows large homogenous mass seen in left parahilar region without air bronchogram.

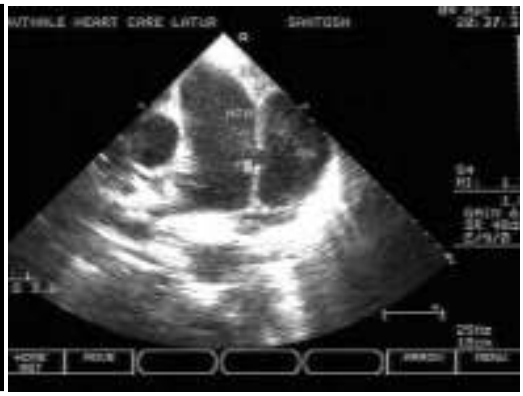

Figure 2: 2D echocardiography is suggestive of dilated pulmonary artery with rent in it and blood flowing to and fro

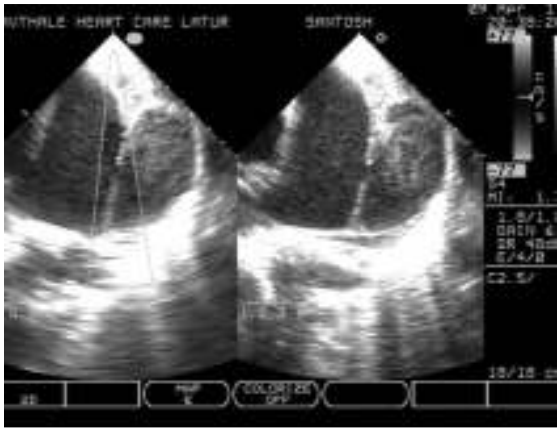

Figure 3(a)

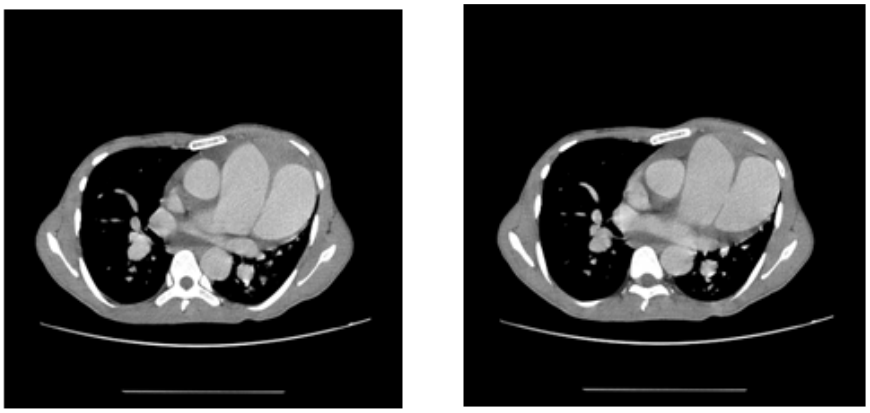

Figure 3 a and b: CT chest with contrast is suggestive of grossly dilated main pulmonary artery with long segment linear hypodensity within the trunk of MPA suggestive of intimal dissection flap (arrow)

DISCUSSION

Aortic aneurysm with dissection is a common and well recognized entity; however, pulmonary artery (PA) dissection is a rare event and usually occurs as a complication of chronic severe pulmonary arterial hypertension, either primary or secondary to underlying congenital heart disease (i.e. patent ductus arteriosus, VSD with eisenmenger`s complex), rheumatic heart disease (i.e. Mitral stenosis with severe PH), vascular abnormality (i.e. cystic medial necrosis, Behçet disease, Marfan syndrome, and Takayasu arteritis), infective endocarditis (Bacterial or mycotic aneurysm) or chronic inflammatory disease like syphilis, tuberculosis, or have undergone cardiac intervention, trauma (i.e. due to improper placement of a catheter), Heart Lung transplant , other rare causes includes amyloidosis, severe atherosclerosis. However Idiopathic pulmonary artery aneurysm leading to dissection is an extremely rare disease. Over the past two centuries, only less than 100 cases of PA dissection have been reported in the literature[3-5] of whom less than 10 were diagnosed during life[4]. Among the previously reported cases of PA dissection, 34 patients had underlying cardiac disease most commonly congenital heart defects including patent ductus arteriosus and seven patients had rheumatic mitral stenosis. Nine patients with idiopathic PA hypertension with dissection have been reported. Two of these patients developed a dissection post lung[6] or heart lung transplantation[7]. In four patients, dissection was associated with pulmonary thrombosis. There are three cases that have been related to a cardiac intervention[6-7]. Although Marfan's syndrome is often associated with aortic dissection, so far there is only one reported case of PA dissection in a patient with Marfan's syndrome[8]. Interestingly, two patients were reported to have aortic and PA dissection, which were not associated with Marfan's syndrome[9]. There are three previous cases that have been reported as idiopathic dissection and were hypothesized to be secondary to some inflammatory or non-specified cause[10]. The overall sex distribution among all of the reported patients appears to be equal with a wide age range from 26 days to 85 years. Peak incidence of dissection was in the third and sixth decades. In younger patients, congenital heart abnormalities were the most common underlying causes, whereas in older patients, other diseases were associated with PA dissection. Pathophysiologic cause of dissection in pulmonary artery aneurysm is not clear, pulmonary hypertension and resultant mucoid degeneration of the media and fragmentation of the elastic fibers strongly predispose predisposes to intimal tears. Dissection of the pulmonary artery always occurs at the site of a pulmonary aneurysm or dilation. It is assumed that pulmonary artery dissection occurs at the point where pulmonary artery tissue becomes too fragile to support the tension of the pulmonary artery wall. Tissue fragility is caused by infectious and connective tissue diseases, and high wall tension is caused by high internal pressure and a large-radius aneurysm. Whether medial degeneration causes the dissection, predisposes to intimal tears, or results from chronically raised intravascular pressure remains controversial. The main pulmonary trunk is the site of dissection in about 80% of patients, usually without involvement of the branches. Occasionally, an isolated dissection of the right or left pulmonary arteries and intrapulmonary branches can occur[8,10]. In a small proportion of patients, PA dissection may occur at the site of localized aneurysm formation. PA dissection usually manifests as cardiogenic shock or sudden death and is therefore typically diagnosed at postmortem rather than during life, there are sporadic reports of patients with PA dissection in the literature who present with acute chest pain during life and diagnosed antemortemly. The symptoms of pulmonary artery dissection are nonspecific, with 82% of patients having exertional dyspnea, 67% having retrosternal chest pain, and 52% having central cyanosis[2]. Most patients present with cardiac shock or sudden death; hence, the diagnosis is rarely made in living patients. In contrast to aortic dissection, the false lumen in pulmonary artery dissection tends to rupture rather than extend distally and develops a reentry site. Rupture occurs most commonly in the pericardium and rarely in the lungs, mediastinum, or pleural cavity, leading to sudden death. Diagnosis is made by noninvasive imaging methods such as chest radiograph, echocardiography and computed tomography. Chest radiography only suggest mediastinal or chest mass. It may be indicative of vascular etiology of the mass. Echocardiography helps in estimating pulmonary artery pressure and excluding other cardiac abnormalities[11]. Transesophageal echocardiography may display the intimal flap, intimal tears, and intraluminal thrombi. Computed tomography of chest with contrast gives accurate evaluation of the diameter and extent of the aneurysm and can exclude the presence of a pulmonary embolism. CT shows the intimal flap within the aneurysm and its extension in to its branches. CT also excludes any other abnormality in the lung parenchyma and mediastinum[12]. Cardiac MRI helps in confirming the intimal dissection of main pulmonary artery and to rule out congenital heart disease. Over the past few decades, PA dissection has been diagnosed during life more frequently[13]. This may reflect the technological advances and increased use of sophisticated modalities like Echocardiography and CT scan to make diagnosis of patient having chest pain, dyspnea or haemodynamic

instability. The optimum treatment of patients with pulmonary artery dissection has not been defined because of the low number of cases in the literature. Because pulmonary artery dissection is highly life threatening, patients with this condition should undergo emergency surgery to prevent sudden death. In past, successful surgical repair has been performed on very few patient, there are very few reports of patients who were managed conservatively with diuretics and vasodilators as they were not willing to undergo surgery although it is not known for how long these patients were followed up and their long term prognosis without surgical repair.

CONCLUSION

Idiopathic Dissecting pulmonary artery aneurysm is extremely rare and fatal event. Diagnosis of it during life is occasionally done as it leads to cardiogenic shock and sudden death. In recent few years with the advent of increasing use of CT scan in patients having chest pain and dyspnea, dissecting pulmonary artery aneurysm frequently diagnosed during life. Our patient is very young and dose not have any predisposing factor particularly congenital heart disease and is extremely unusual that the patient was hemodynamically stable despite symptomatic since last 15 days. The exact etiology, predisposing factors, progression, natural course and optimal treatment of idiopathic PA dissection is not known.

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