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Case Report
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Bilateral Pulmonary Artery Aneurysms with Pulmonary Arterial Hypertension Idiopathic

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Abstract

Background: Pulmonary artery aneurysm is a rare but fatal complication of Idiopathic pulmonary arterial hypertension. There are no clear guidelines about its optimal management.

Case Report: We report a 28-year-old male admitted to the hospital with hemoptysis. He had a suspicion of pulmonary arterial hypertension since the age of 14 years but he was lost to follow-up. The patient was diagnosed with pulmonary artery aneurysm due to idiopathic pulmonary arterial hypertension. The patient subsequently died one month later due to hemoptysis secondary to suspected rupture.

Conclusion: The diagnosis of idiopathic pulmonary hypertension is hard because of its non-specific nature. Early diagnosis and treatment can prevent complications.

Keywords: Idiopathic pulmonary arterial hypertension; Pulmonary artery aneurysm; Rupture

Introduction

Idiopathic pulmonary arterial hypertension is a rare but fatal disease that affects the pre-capillary pulmonary vasculature. The exact risk factors for idiopathic pulmonary hypertension are unknown. A pulmonary arterial mean pressure more than 25 mmHg at rest and more than 30 mmHg during exercise define pulmonary arterial hypertension. If left untreated, it can result in increasing back pressures and ultimately right heart failure and death [1]. Pulmonary artery aneurysm has been described as a dilatation of the pulmonary artery more than 29 mm [2]. Pulmonary artery aneurysm is a rare complication of idiopathic pulmonary arterial hypertension however, optimal management and timing of intervention for this rare entity is not well understood. Aneurysm formation and progression is associated with vessel wall stress and rupture carries an extremely high mortality rate (50% to 100%) [3]. We report a case of a 28-year-old male admitted to the hospital with hemoptysis and dyspnea III NYHA revealing idiopathic pulmonary arterial hypertension and pulmonary artery aneurysm.

Case Presentation

A 28 -year-old male patient was admitted to the hospital with fatigue, hemoptysis and dyspnea III NYHA. He had a suspicion of pulmonary arterial hypertension since the age of 14 years. At that time, there were no clinical symptoms besides the presence of recurrent respiratory infections and heart murmur. The patient was referred for hemodynamic and angiographic studies. However, he was lost to follow-up. During the current hospitalization, the physical examination revealed signs of right heart failure. Electrocardiography revealed signs of right ventricular hypertrophy with right atrial enlargement and right axis deviation. Chest X-ray revealed cardiomegaly, middle left arch convexity, and bilateral semilunar perihilar opacities due to enlargement of the main pulmonary artery and its branches (Figure 1). Echocardiography showed an enlarged Main Pulmonary Artery (MPA) with a diameter of 39 mm. The Right Pulmonary Artery (RPA), the left one, and the right ventricle were enlarged with a diameter of 40 mm, 42 mm and 42 mm respectively with symptoms of pulmonary hypertension. There was no sign of intracardiac or extracardiac shunting. Chest CT scan with contrast revealed Main pulmonary artery aneurysm (40 mm) with significant enlargement of the left (45 mm) and right pulmonary artery (43 mm) (Figure 2, 3). Right heart catheter demonstrated pulmonary artery pressure of 120/92 mmHg (mean 80 mmHg), right ventricular pressure of 120/10 mmHg, Pulmonary capillary pressure of 9 mmHg and a mean right atrial pressure of 10 mmHg. Vaso reactivity testing (nitric oxide) was positive. Potential primary causes of pulmonary arterial hypertension were eliminated, such as heart disease, pulmonary embolism, and the presence of any pulmonary disease. Additional laboratory studies excluded collagen and inflammatory disorders.

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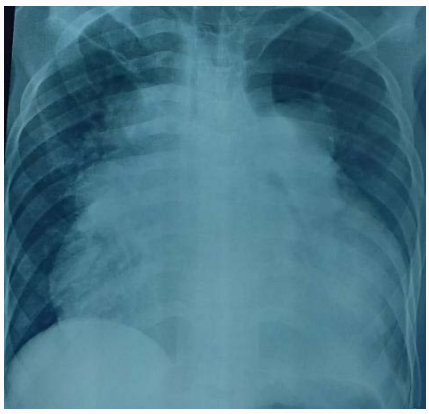

Figure 1: Chest X-ray showing cardiomegaly, middle left arch convexity, and bilateral semilunar perihilar opacities due to enlargement of the main pulmonary artery and its branches.

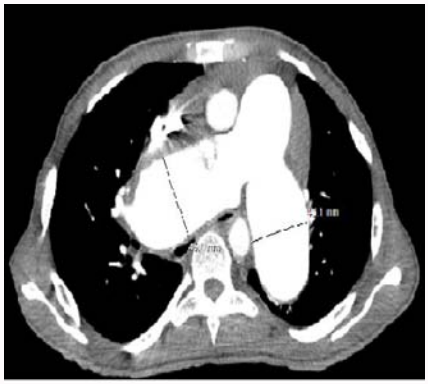

Figure 2: Main pulmonary artery dilatation with right and left pulmonary artery enlargement (Chest computed tomography scan with contrast protocol).

According to the gathered data, a diagnosis of Idiopathic pulmonary arterial hypertension was stated. Conservative treatment based on oral, high-dose calcium channel blockers was chosen. The treatment aimed to lower the pulmonary artery pressure while monitoring the aneurysm. The patient subsequently died one month later due to hemoptysis secondary to suspected rupture.

Discussion

No-specific presentation of pulmonary arterial hypertension leads to a delay in diagnosis. As the disease advances, other symptoms such as fatigue, near syncope, syncope, chest pain or angina, palpitations, and peripheral edema occur. The management is based on NYHA Classification with the goal improving symptoms and functional status. Calcium channel blocker is only used in the minority of patients with vasoreactive pulmonary arterial hypertension. Initiation of specific pulmonary hypertension therapy is recommended in patients without marked hemodynamic improvement after high doses of Calcium channel blockers [4,5]. Pulmonary artery aneurysm is a documented complication of idiopathic pulmonary arterial hypertension however, optimal management and timing of intervention for this rare entity is not understood. Patients with pulmonary artery aneurysm present with non-specific symptoms (chest pain, dyspnea and cough) and are referred with the suspicion of a pulmonary artery dilatation seen on chest X-ray [6]. Dyspnea, chest pain, hemoptysis, or the large size of the aneurysm can be risk factors of forthcoming rupture that is mostly fatal. The diagnosis requires trans-thoracic echocardiography and other imaging techniques including pulmonary angiography, magnetic resonance imaging or computed tomography. The complications of pulmonary artery aneurysm include dissection, rupture, intra-pulmonary erosion, compression of the trachea, bronchi pulmonary and superior vena cava or recurrent laryngeal nerve. Dissection and rupture are the most fatal complications. For patients with pulmonary artery aneurysm, there are no clear guidelines about its optimal management. Bilateral lung transplant can be done in most pulmonary hypertension patients, even in those with severe right ventricular dysfunction [7,8]. Another theoretical option would have been a graft repair of the aneurysm and pulmonary valve disease repair/replacement. Heart-lung transplantation is a good surgical option for difficult cases of idiopathic pulmonary arterial hypertension complicated by pulmonary artery aneurysm and right ventricular failure [9]. In the present case, bilateral pulmonary artery aneurysms and the right atrial and ventricular dilatation were secondary to idiopathic pulmonary hypertension. This case shows that the non-specific presentation of idiopathic pulmonary arterial hypertension leads to a delay in diagnosis.

Conclusion

Pulmonary artery aneurysm is a fatal complication of idiopathic pulmonary arterial hypertension. Most patients with idiopathic pulmonary arterial hypertension present with non-specific symptoms. Physicians should pay more attention to the presence of recurrent respiratory infections in children, which can be the first sign of the disease. This will prevent delay in diagnosis and the adverse complications of the disease.

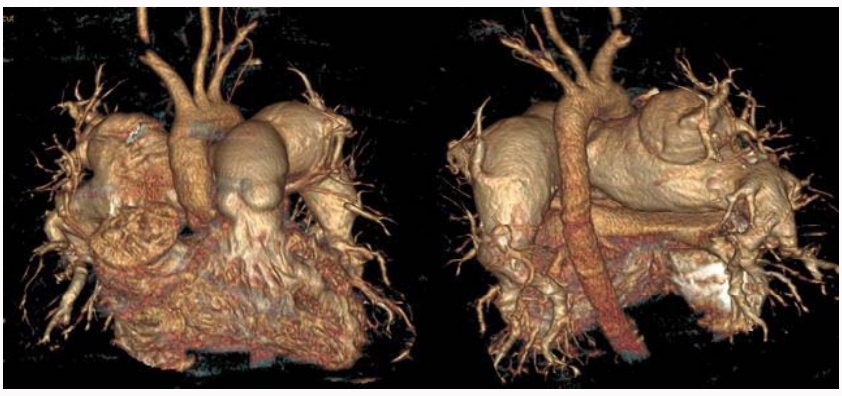

Figure 3: Main pulmonary artery dilatation with right and left pulmonary artery enlargement (computed tomography VRT reconstruction).

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