Pulmonary Arterial Aneurysm with Thrombus Formation and Atrial Septal Defect with Pulmonary Hypertension

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Abstract

Introduction: Pulmonary arterial aneurysm (PAA) is a rare condition with varying symptomatology. Patients are often referred due to incidental finding of intrathoracic mass on chest radiograph. Common causes include structural cardiac and vascular abnormality, pulmonary hypertension, infection, vasculitis, connective tissue disease, and trauma.

Case presentation: A 37-year-old female came in due to shortness of breath associated with easy fatigability, exertional dyspnea, orthopnea, and occasional palpitations. Workup revealed 2D echocardiography findings of atrial septal defect (ASD) with left to right shunt; dilated right atrium and right ventricle; severe pulmonary hypertension; dilated main pulmonary artery. Contrast-enhanced computed tomography (CT) angiography showed PAA in the right main trunk, left and right pulmonary artery with thrombus formation. Cardiac catheterization and hemodynamic study were intended prior to definitive surgical treatment however patient did not opt to proceed. She was managed with warfarin, bisoprolol, digoxin, and trimetazidine. In spite of symptom relief, the patient still needs regular follow-up for reassessment to monitor disease progression.

Discussion: Pulmonary arterial aneurysm (PAA) is a rare complication of ASD, a common congenital heart disease. In this case, it presented as a mediastinal mass with non-specific symptoms diagnosed after routine chest x-ray, verified through echocardiogram. Contrast-enhanced CT angiography ruled out aortic aneurysm and provided additional information of the extent of thrombus formation. However, pulmonary angiography still remains the diagnostic gold standard. Due to the rarity of the disease, optimal treatment of PAA is still uncertain. It is reasonable, however, to treat conservatively for asymptomatic patient with no significant pulmonary hypertension. A more aggressive surgical approach has been advocated for patients with complications.

Conclusion: It is important to emphasize that PAA can present with non-specific manifestations thus high index of suspicion is critical for the diagnosis. It is also vital for early detection of the aneurysm and correction of the etiology to prevent disease morbidity and mortality.

Keywords: pulmonary artery aneurysm, thrombus, atrial septal defect

Introduction

Pulmonary artery aneurysm (PAA) is a rare condition, which is defined as a pulmonary artery diameter greater than four centimeters.[1,2] This can develop due to congenital or acquired heart conditions, infections, collagen vascular diseases, connective tissue disorders, trauma, vasculopathic changes and may be idiopathic.[3] In more than 50% of all cases, PAA is associated with congenital heart disease.[1,2,4] Likewise, pulmonary arterial hypertension is an important cause of PAA formation.[5] In this case, the patient had uncorrected atrial septal defect (ASD) with pulmonary hypertension as the underlying cause of PAA. Pulmonary arterial aneurysm generally affects younger age group with an equal sex incidence.[5] Eighty-nine percent of all PAAs are located in the main pulmonary artery.[5] Also, aneurysms more commonly develop in the left pulmonary artery.[6] The diagnosis often requires radiological imaging, as the clinical presentations are nonspecific.[3,7] Since this is a rare condition, clinical cases are limited and there are no standardized management.[2] It is rational however to manage conservatively in cases of PAA without significant pulmonary hypertension. But definite operative treatment is recommended for patients with complications or risk of rupture.[8]

Case Presentation

A 37-year-old female, Filipino was admitted due to shortness of breath. She was diagnosed 14 years ago to have ASD with pulmonary hypertension, based on a two-dimensional (2D) echocardiography requested in the course of a consult for cough, paroxysmal nocturnal dyspnea and orthopnea. She refused the recommended surgery then. She was maintained with unrecalled medications but lost to

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follow-up. One month prior to admission, she complained of occasional chest pain associated with rapid beating of her heart, easy fatigability, exertional dyspnea, orthopnea, paroxysmal nocturnal dyspnea, and insomnia. She was admitted at a local hospital and workup revealed chest radiographic finding of a mediastinal mass. Computed tomography (CT) scan of the chest revealed dilated pulmonary artery and its branches; mass lesion in the left parahilar area was not ruled out. The patient was given trimetazidine, digoxin, and carvedilol, which provided relief. She was then discharged improved and advised to seek consult at a tertiary hospital for second opinion. However, a few days after discharge, the patient developed shortness of breath associated with chest pain, which prompted consult and was subsequently admitted. On examination, she was conscious, coherent, normotensive, in mild respiratory distress with tachypnea of 24 cycles/min, regular pulse rate/cardiac rate of 69 beats/min. Cardiovascular system revealed the presence of soft, grade 2/6, mid-systolic murmur at the second left intercostal space with fixed S2 splitting, loud P2, and sternal heave. PMI located at fifth left ICS. No cyanosis, ascites, or bipedal edema. Blood work up revealed normal complete blood count, serum electrolytes, creatinine and cardiac enzymes.

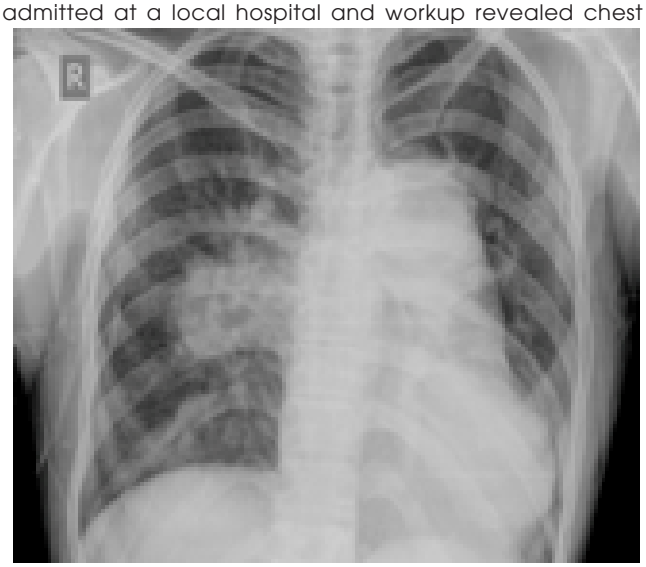

Figure 1. Chest radiograph (posterior-anterior view) showing a parahilar opacity on the right

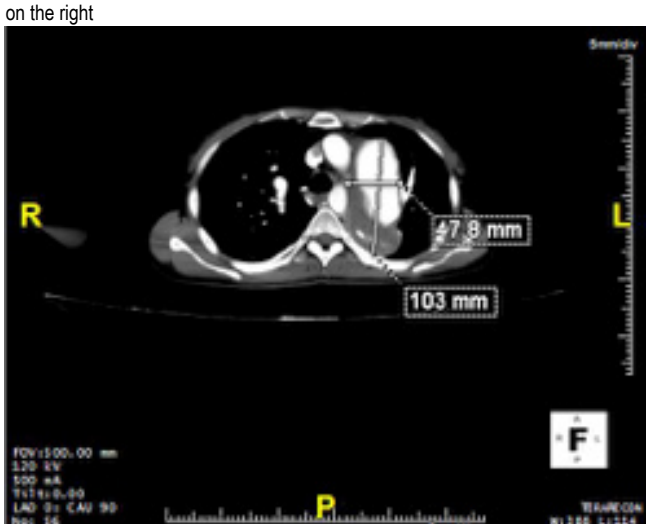

Figure 2. Axial CT scan of the chest at the level of the main pulmonary artery

Electrocardiogram (ECG) revealed sinus rhythm, right ventricular hypertrophy with diffuse wall ischemia. Chest radiograph showed parahilar opacity on the right (Figure 1). 2D echocardiography confirmed an ASD with left-to-right shunt, Qp:Qs of 2:1, normal left atrial size, small left ventricular size with flattening of interventricular septum during systole and diastole; dilated RA and RV with ejection fraction of left and right ventricle of >55%; severe pulmonary hypertension by tricuspid regurgitation jet; severe tricuspid regurgitation; dilated main pulmonary trunk (Figure 3). Aortogram was done to rule out aortic aneurysm. Chest CT scan showed widened main pulmonary trunk measuring 4.78 cm in widest diameter with thrombus formation on the left pulmonary artery (Figure 2). CT angiography also revealed aneurysmal dilatation of pulmonary trunk measuring 5.46 cm in its transverse diameter, 4.33 cm in the right pulmonary artery and 4.94 cm in the left pulmonary artery with thrombus (Figure 4).

Patient was managed as a case of congenital heart disease, atrial septal defect, right ventricular and right atrial

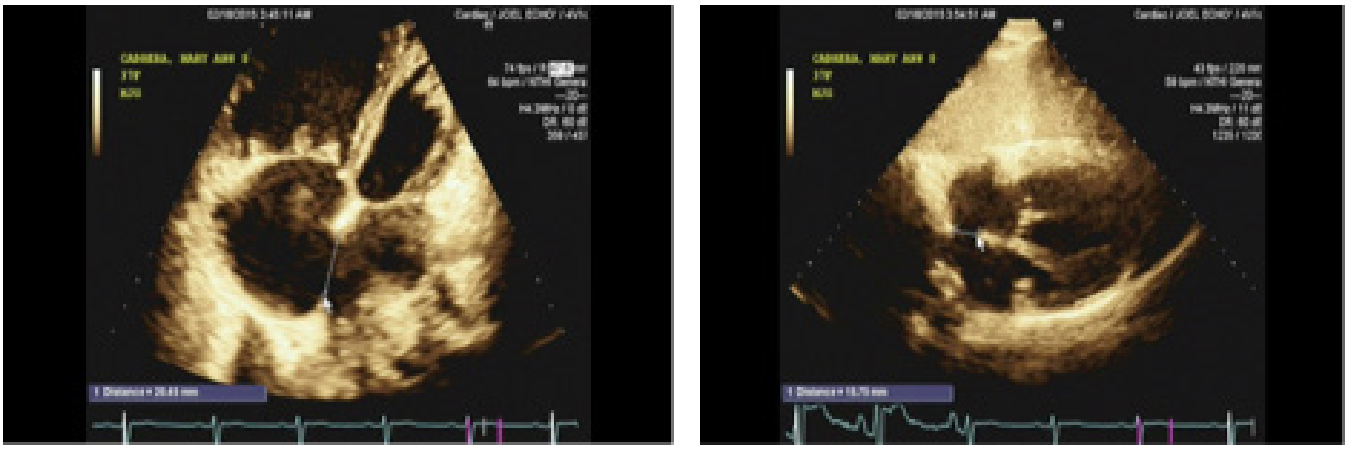

Figure 3a and 3b. Echocardiographic view of atrial septal defect

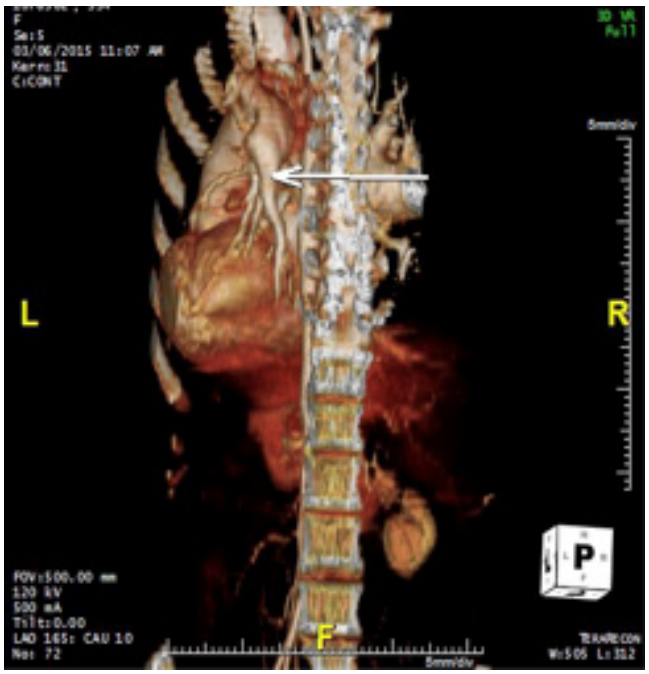

Figure 4. CT angiography showing aneurysmal dilatation of pulmonary artery

enlargement, tricuspid regurgitation, sinus rhythm, Functional Class II; Pulmonary artery aneurysm with thrombus formation; pulmonary hypertension.

Due to the low prevalence of the cases identified ante-mortem, there has been no clear guideline in optimal management. Definitively, the underlying etiology of this disease process must be corrected in which case the closure of ASD and repair of the aneurysm. The patient then was prepared for right heart catheterization and hemodynamic study to assess significant pulmonary hypertension prior to surgery, however, concurrent with the resolution of her symptoms with conservative management of bisoprolol (beta-blocker) 2.5mg tab per orem (PO) once a day (OD), warfarin (vitamin K antagonist) 2.5mg tab OD, digoxin (Na-K ATPase inhibitor) 0.25mg tab PO OD, and trimetazidine (anti-angina medications) 35mg tab twice a day (BID), family opted not to proceed with the surgical plans. Nonetheless, it was emphasized to them the need for further follow-up every three to six months to monitor the aneurysmal dilatation and disease progression until a definite surgical procedure is done.

Discussion

Aneurysm of the pulmonary artery is a rare disease in adults. The precise incidence of the disease is unknown however they represent less than one percent of all thoracic artery aneurysms.[9] Its estimated incidence is one in 14,000 autopsies and only eight cases were detected in a series of more than 100,000 necropsies reported in 1947.[9] Predisposing underlying conditions include collagen vascular disease, infection or Behcet's disease, vasculitides, pulmonary valve stenosis with post-stenotic dilatation or congenital heart disease i.e. left-to-right shunt.[1,5,8,14]

In this case, a differential diagnosis of the etiologic cause includes Behcet's syndrome however, this is less likely since the presentation of recurrent oral and genital ulcerations as well as ocular involvement were not manifested. Another consideration could be Marfan's syndrome, a connective tissue disease clinically presents with a triad of (1) skeletal changes that include long, thin extremities, frequently associated loose joints, (2) reduced vision is the result of dislocations of the lenses, (3) and aortic aneurysms, such as pulmonary artery aneurysm.[10] However, in our case, there was no arachnodactyly, Walker sign nor Steinberg sign and the arm-length was not significantly longer than the height of the patient.

In more than 50% of the cases, pulmonary arterial aneurysms are associated with congenital heart disease, most frequently patent ductus arteriosus, followed in decreasing order by ventricular septal defect, atrial septal defect, hypoplastic aortic valve, and bicuspid aortic valve.[9,11] In our case, the patient had long-standing uncorrected atrial septal defect complicated with pulmonary arterial hypertension as the etiology of PAA.

Atrial septal defect (ASD) are typically asymptomatic in infancy and early childhood, and elective defect closure is usually performed at ages of four to six years.[12] Severe pulmonary hypertension complicating left-to-right shunting at the atrial level is seen in significant proportion of untreated patients in later adulthood.[13] In moderate to large atrial septal defects, if not-operated, clinical parameters tend to deteriorate over time, thus prompt closure of such defect seems to be the preferred treatment option.[13] Nonetheless, pulmonary arterial hypertension resolves following successful surgical procedures, unless it is too far advanced.[14]

Secondary pulmonary artery hypertension (SPAH), which is the case of the patient, is defined as a pulmonary artery systolic pressure higher than 30 mm Hg or a pulmonary artery mean pressure higher than 20 mm Hg secondary to either a pulmonary or a cardiac disorder.[14] One of the three pathophysiologic mechanisms of the pulmonary hypertension is pressure-volume overload caused by left-to-right intracardiac shunts, left ventricular dysfunction or valvular disease.[14] Furthermore, perturbation from sheer stress resulting from shunt flow may induce apoptosis, remodeling, and aneurysmal transformation of the vessel wall.[5] In our case, the presence of left-to-right shunt with Qp:Qs ratio of 2:1 increases the pressure-volume on the right-sided chambers coupled with severe tricuspid regurgitation further aggravating the wall stress on the right ventricle resulting to pulmonary hypertension. Nevertheless, the patient had normal left ventricular function with ejection fraction of

>55% which excludes it as the etiology of the pulmonary hypertension.

Clinical presentation of a PAA varies depending on the etiology. A high index of suspicion is required for the diagnosis of pulmonary artery aneurysm since most of the clinical features are nonspecific.[2,3,5,7] Patients with PAA can be asymptomatic presenting as an incidental finding on chest radiograph or more frequently, presents with dyspnea, hemoptysis and chest pain due to underlying pulmonary hypertension.[2,5,16] In our case, the patient's symptoms were due to severe pulmonary hypertension worsened by the presence of thrombus.

Chest radiography can provide initial evidence of pulmonary artery aneurysm presenting either as hilar opacity or mediastinal mass. The 2D transthoracic echocardiography with Doppler analysis is used to confirm the diagnosis of PAA with or without pulmonary arterial hypertension and to exclude possible cardiac disease.[14] Pulmonary angiography still remains as the gold standard in the diagnosis of PAA.[7,15] However, newer non-invasive imaging studies such as contrast-enchanced CT angiography and magnetic resonance imaging (MRI) have simplified the diagnosis.[7] Due to high spatial resolution of contrast-enhanced CT angiography, it is considered as the primary technique for the diagnosis of PAA. It also offers additional information on the exact location of the aneurysm, size, shape and concomitant cardiovascular disease.[7] In our case, CT angiography with contrast was done which refined the diagnosis.

Complications are considered life-threatening events, such as dissection, stenosis of bronchi or other surrounding structures, and pulmonary thrombosis.[16] Thrombosis is a common complication in pulmonary hypertension of any etiology and may worsen the patient's functional class which is diminished as a result of pulmonary hypertension.[1] Our patient neither had dissection, compression of surrounding structures nor bronchial stenosis however, there was evidence of pulmonary thrombosis.

In general, the optimal treatment of PAA is still uncertain. Owing to infrequency of the disease and limited knowledge, there is no clear guideline for best therapeutic approach.[1,5] Furthermore, there has been no accepted standard diameter of PAA for surgical intervention, but based on data extrapolated from the natural history of aortic aneurysms, it was suggested that a diameter of six centimeters or greater is deemed for surgical intervention to avoid rupture.[15] In addition, aggressive surgical approach has been advocated for patients with pulmonary hypertension because of the risk of impending dissection and rupture of the aneurysm.[17,18] However, it is reasonable to treat conservatively for asymptomatic patient with no significant evidence of pulmonary hypertension but needs to be re-evaluated regularly.[5] In our patient, even with resolution of symptoms with medical management, the presence of pulmonary arterial hypertension and thrombus formation ideally should have been managed with surgical closure of ASD and pulmonary aneurysmal repair. Though, in terms of the surgical outcome, since no large series of PAA have been published, mortality and morbidity data cannot be provided.[5] Even so, a definite surgical intervention is imperative to prevent worsening of pulmonary hypertension and to avoid further morbidity and mortality.

Conclusion

Atrial septal defect (ASD) is a common adult congenital heart disease that is typically asymptomatic in early childhood. If not operated, it can progress to pulmonary hypertension that can further develop to a rare complication of PAA. Depending on the etiology, it can present with non-specific clinical manifestations. PAA can be asymptomatic presenting as a mediastinal mass that can be identified on chest radiograph but should be confirmed through a transthoracic echocardiogram. Pulmonary angiography still remains the gold standard for the diagnosis but contrast-enhanced CT angiography can be considered as the primary technique, which can also provide information of the extent of the aneurysm with or without thrombus formation. Even though there are no clear guidelines for optimal management, conservative management is reasonable if there is no presence of significant pulmonary hypertension. it is also crucial to timely detect the aneurysm and to correct the underlying etiology to prevent further complications.

Disclosure: None to declare

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