Case Report

Multiple pulmonary artery aneurysms in a young female with patent ductus arteriosus on CT pulmonary angiography

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ABSTRACT

Aneurysms of lobar branches of pulmonary artery are extremely rare with few reported cases in literature. The exact etiopathogenesis of pulmonary artery aneurysms is not clearly understood and no clear guidelines on their management is available. Aneurysms of the secondary and tertiary branches appear to be even more uncommon than those of the trunk and primary branches.

Here we report an unusual case of young female with multiple pulmonary artery aneurysms in bilateral lungs with fusiform dilation of main pulmonary trunk and a small patent ductus arteriosus.

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Introduction

Aneurysms of the main pulmonary artery and its lobar branches are rare with very few cases reported in literature. Although rare, these aneurysms pose a significant risk of rupture and are potentially life threatening and warrant a prompt diagnosis and treatment. Any focal dilatation of the main pulmonary trunk measuring greater than 29 mm in diameter is considered abnormal and an interlobar artery diameter more than 17 mm is considered abnormal [1]. A true aneurysm contains all three layers of the vessel wall. The mortality rates associated with a pulmonary artery rupture has been reported from 50% to 100%, death occurs primarily due to aspiration of blood and asphyxia after intrapulmonary hemorrhage [3,4]. Sudden cardiac arrest can also occur secondary to a pulmonary artery dissection [2], hence prompt diagnosis and treatment is of utmost importance for patient survival and favorable prognosis.

Case Report

A 30-year-old female presented with complaints of dry cough for 2 weeks with breathlessness on minimal exertion. There

Abbreviations: PAA, Pulmonary artery aneurysms; PAPA, pulmonary artery pseudo-aneurysms; PAH, pulmonary artery hypertension.

* Competing Interest: The authors declare that they have no competing interests.

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was no history of any fever or loss of weight and appetite. The patient had prior history of taking anti tuberculosis medication 15 years ago and had completed the course.

On examination the patient was conscious, oriented, afebrile with no pedal edema, cyanosis or clubbing was present. The patient although had grade 1 dyspnea. The patient's ECG was normal and her resting SpO2 was 97% on room air. On auscultation breath sounds were heard equally in bilateral lung fields with no abnormal sounds heard. Sputum for acid fast bacilli was negative, cartridge based nucleic acid test for tuberculosis was negative. On routine hemogram patient's Hb was 10.5 g/dl and Total leukocyte count 6500 with 45% neutrophils.

To rule out heart disease an echo cardiogram was done which demonstrated a moderate patent ductus arteriosus with left to right shunt with moderate pulmonary artery hypertension and dilated left ventricle.

The routine chest x-ray (Figs. 1-3) of the patient was done which showed 2 well defined round to oval opacities in the right lower lung zone. There was enlargement of the left pulmonary bay. Right hilum was normal. Trachea was in midline. No evidence of any pleural effusion or pneumothorax. Bony thorax was normal.

Patient was advised multislice Ct pulmonary angiography which had the following findings.
Fig. 2–2.9. Multislice pulmonary angiography- Axial, coronal and sagittal reformatted images demonstrating the pulmonary artery aneurysms arising from the lobar branches of the right and left pulmonary arteries.

A well defined 3.8(AP) × 3.7(T) × 3.5(CC) cm sized rounded homogenously enhancing lesion (HU; Pre contrast = 40, post contrast = 120) following the blood pool, with a rim of peripheral calcification was noted in the right lower lung lobe, and was seen to communicate with the segmental branch of right pulmonary artery and the postaneurysmal segment showed fusiform dilatation measuring 9.3 mm in maximum external diameter.

Two other lesions showing similar attenuation of size 1.7 × 1.7 cm and 2.6 × 2.1 cm were noted in the right middle and left lower lung lobes respectively, and were seen to communicate with the segmental branches of right and left pulmonary arteries, respectively.

The main pulmonary trunk appeared dilated, measuring 42 mm in maximum diameter; right and left pulmonary arteries appear dilated measuring 25 and 23 mm in maximum external diameter.

On the virtual reconstruction images showed a patent ductus arteriosus of maximum width 9 mm extending from the distal arch of aorta to the pulmonary trunk, correlating with the echocardiographic findings.

Septal thickening and few fibrotic strands noted predominantly in bilateral basal, apical lung fields and in right middle lung lobe, possibly representing sequelae of old tuberculous lesions.

The above mentioned dilatations of the lobar branches of the left and right pulmonary arterial branches possibly represent pulmonary arterial aneurysms with a dilated main pulmonary trunk with a patent ductus arteriosus.

The patient’s current symptoms could be attributed to the moderate patent ductus arteriosus with resultant left to right shunting and pulmonary hypertension. The pulmonary artery aneurysms being incidental findings.

The patient refused further treatment and was discharged against medical advice.

Discussion

Pulmonary artery aneurysm (PAA) can either be acquired or congenital, the incidence of PAA was reported to be 1:14,000 with most occurring in the main pulmonary artery.
Congenital heart diseases are the most common anomalies usually associated with PAA. The exact pathophysiology is exactly unknown but involves altered flow dynamics and increased hemodynamic shear stress on the vessel walls usually caused by left- to – right shunts [5–7]. The most common congenital cardiac anomalies that are usually associated with PAA in decreasing order include patent ductus arteriosus, ventricular septal defect, atrial septal defect, hypoplastic aortic valve, and bicuspid aortic valve and pulmonary valve stenosis [5–7].

Among other acquired causes connective tissue disorders like Marfan’s syndrome, Ehlers-Danlos syndrome and cystic medial necrosis are also common but usually affect the aorta and its branches.

Bacterial or fungal infections can cause mycotic pseudoaneurysms and less commonly aneurysms due to their ability to destroy or alter vessel walls. In patients with advanced syphilis, aneurysm formation most commonly occurs in the large pulmonary arteries with destruction occurring at the level of the vasa vasorum [8].

Patients with long standing tuberculosis (TB) are at high risk of pseudoaneurysm formation in the intraparenchymal pulmonary arteries adjacent to tuberculous cavitary lesions [9]. These aneurysms are known as Rasmussen aneurysms which are usually seen more commonly in the upper lung lobes and are peripheral PAA s which occur due to the erosion of the wall of a peripheral pulmonary artery branch may lead to a bout of massive hemoptyisis. Septic emboli form a bacterial endocarditis or pneumonia can also spread hematogenously and cause mycotic PAPAs.

Pulmonary artery hypertension (PAH) has been widely implicated as a cause of PAA [10]. The mechanism of aneurysm formation is due to increased arterial wall stress.

Pulmonary thromboembolic disease has been implicated as a cause of PAA or PAPA, by either of the 2 mechanisms - one the thrombus formed can cause direct intimal injury leasing to the formation of a PAPA or Chronic thromboembolic disease can cause wall thickening, mural webs and calcifications which can lead to weakening of the wall and aneurysm formation.
In vasculitic disorders like Behcet’s disease and Hughes-Stovins syndrome also PAA s are common [12,13]. The obliterator endarteritis of the vaso vasorum of the pulmonary artery can lead to transmural necrosis and aneurysm formation [11,13].

Atherosclerotic disease and chronic lung disease can also lead to thinning of vessel wall and aneurysm formation leading to PAA s [6].

Iatrogenic and traumatic causes have increased in incidence as a source of PAA and PAPA. Numerous cases have been reported citing causes such as Swan-Ganz catheter placement, chest tube placement, catheter angiography, cardiothoracic surgery and lung biopsies [2,10,24–26]. Idiopathic PAA is a rare diagnosis that is made in absence of any such definitive causative factor.

In order to better characterize and classify PAAs, 4 pathological criteria have been defined: (I) simple dilatation of the pulmonary trunk, with or without involvement of the rest of the arterial tree; (II) absence of abnormal intra- or extracardiac shunts; (III) absence of chronic cardiac or pulmonary disease, either clinically or at autopsy; (IV) absence of arterial disease, such as syphilis or more than minimal atheromatosis or arteriolar sclerosis [14].

Diagnosis

The mainstay of imaging for both detection and follow-up of PAA and PAPA remains computed tomography angiography (CTA). Multidetector row CTA performed with bolus tracking over the descending aorta, along with coronal and sagittal multiplanar and maximum-intensity projection (MIP) reformatted images, has been reported as an imaging protocol for hemothysis [15]. The advantage of CTA is that it allows for the assessment of presence, size, location, and characteristics including saccular or fusiform aneurysm type [16]. Equally as important, it can provide information regarding the underlying etiology of an aneurysm in that additional clues may be found in the lungs, heart, or mediastinal structures [16].

Three-dimensional and advanced postimaging processing software can prove to be extremely valuable in both detection as well as treatment planning.

Dual-energy CT is an advanced CT technique that allows for better material differentiation, can also overcome some artifacts thus improving CT pulmonary angiogram scan quality at relatively reduced contrast and radiation dose [17].

Role of MR angiography - Magnetic resonance imaging, while not as commonly used as CTA for evaluating the pulmonary arteries, is a viable alternative where CTA cannot be employed (ie, allergy to iodinated contrast material, renal insufficiency, metallic heart valve implants). Fast spin echo and gradient echo imaging are useful in the morphologic evaluation of pulmonary vasculature from the main pulmonary trunk to the subsegmental level.

Management

After diagnosis the treatment must be targeted to the underlying cause or causes as well as choosing the least invasive procedure to treat the aneurysms with a favorable outcome.

Neither a consensus to treat PAA based on size criteria nor treatment guidelines delineating the roles of medical or procedural disciplines currently exist, hence a multidisciplinary approach to the problem might be beneficial.

Several surgical techniques are available like aneurysmorhaphy, lobectomy, bilobectomy, aneurysmectomy, and pneumonectomy [23], however surgical resection carries a high risk of intrapulmonary hemorrhage in case of pulmonary hypertension.

Endovascular treatment [18] can be offered as the first line therapy when feasible with the advantage of less morbidity and mortality feasible. Endovascular therapy may best serve saccular PAA or PAPA, both in the central and peripheral pulmonary arteries [18–22].

Coil embolization of arterial aneurysms or pseudoaneurysms throughout the body is a widely accepted minimally invasive therapy. However bronco-pulmonary shunting is to be ruled before such a procedure.

Vascular plugs, stent-assisted coil embolization, glue (cyanoacrylate), and transcatheter balloon embolization are the other techniques used.

Conclusion

To summarize, pulmonary artery aneurysms are rare entities that warrant a prompt diagnosis and treatment despite non specific clinical features. CT pulmonary angiography remains the gold standard investigation of choice. Despite not having a large scale consensus on their management, by a multidisciplinary team of intensivist, pulmonologist, interventional radiologist, thoracic surgeon should plan an approach that would prove beneficial with minimal procedure related morbidity and mortality and improving survival.

Consent for study

Written and informed consent was obtained from the patient for publication of this case report and any accompanying images.

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