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Exploring Pulmonary Artery Aneurysms and Pseudoaneurysms: A Comprehensive Review

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A B S T R A C T

Pulmonary artery aneurysms represent an unusual yet intriguing pathology under the spectrum of cardiovascular diseases. The etiology of Pulmonary artery aneurysms encloses a diversified range of conditions including congenital cardiovascular defects, infectious agents such as tuberculosis, systemic vasculitis, connective tissue disorders like Marfan syndrome, and trauma. Pulmonary artery aneurysms give rise to diagnostic challenges due to their asymptomatic nature which delay the complications. Early detection is very crucial as clinically patients with pulmonary artery aneurysms remain asymptomatic or present with vague symptoms including chest pain, dyspnea, or hemoptysis. The management of this disease is very controversial and treatment strategies differ based on the aneurysm size, location, and etiology, ranging from surgical interventions like aneurysm resection or endovascular stenting to conservative management. In this review, we will highlight the etiologies, clinical manifestations, and diagnostic approaches including imaging features of pulmonary artery aneurysms. Moreover, we will also discuss the need for recent advancements in the management of Pulmonary artery aneurysms to optimize patient outcomes and enhance our understanding of this incomprehensible cardiovascular disease.

Keywords: Pulmonary artery, aneurysm, pseudoaneurysm, congenital, cardiology, surgery.

1. INTRODUCTION:

A pulmonary artery aneurysm is an unusual but potentially critical condition that is characterized by focal dilatation of a segment of the pulmonary artery that can lead to significant life-threatening complications including pulmonary embolism, thrombosis, and rupture. Although a true aneurysm is defined as dilatation of the artery involving all three layers of vasculature wall – tunica intima, tunica media, and tunica adventitia, on the other hand, pseudoaneurysm doesn't involve all three layers which in turn makes it more liable to rupture. A pulmonary artery pseudoaneurysm (PAPA) is a rare and potentially life-threatening disease characterized by a saccular outpouching of a pulmonary artery representing a contained rupture of that artery(1). Although their incidence is low the mortality rate associated with the rupture of pulmonary artery aneurysm (PAA) or (PAPA) has been reported from 50% to 100%; death is secondary to aspiration or asphyxia after intrapulmonary hemorrhage (2, 3). PAA can also lead to dissection of the pulmonary artery and sudden cardiac death (1, 4). Hence prompt diagnosis and treatment are crucial for the survival of patients and in improving outcomes of the patients with PAA.

2. ETIOLOGIES:

- **Congenital:**

Historically congenital causes compromised approximately half of the reported cases of PAA[5]. Currently, the routine use of computed tomography(CT) of the chest has changed that landscape. At present congenital causes compromised 25% of the total cases reported. Congenital causes are further subclassified into three groups- Hereditary haemorrhagic telangiectasia (HHT), Connective tissue disorders, and Congenital Heart Diseases.

- Hereditary haemorrhagic Telangiectasia (HHT): It is an autosomal dominant disorder designated by abnormal blood vessel formations resulting in arteriovenous malformations including pulmonary artery aneurysms.
- Congenital Heart Diseases: This includes structural defects like atrial septal defect (ASD), ventricular septal defect (VSD), or patent ductus arteriosus (PDA). These defects can cause increased blood pressure and flow in pulmonary circulation for example Eisenmenger's syndrome potentially leading to the formation of an aneurysm.
- Connective tissue disorders: For example Marfan syndrome and Ehlers-Danlos syndrome.

Marfan syndrome is a genetic disorder principally affecting connective tissues including blood vessels, predisposing individuals to arterial wall weakness and subsequently aneurysm formation. Similarly, Ehlers-Danlos syndrome is another connective tissue disorder characterized by arterial fragility and aneurysm formation.

- **Acquired:**

1. **Pulmonary hypertension**: It is caused by chronic elevation of pulmonary arterial pressure due to various etiologies such as chronic obstructive pulmonary disease (COPD), idiopathic pulmonary arterial hypertension(PAH), or left heart disease which in turn may lead to pulmonary artery dilation and aneurysm formation.
2. **Autoimmune diseases and vasculitis**: In this category, Behcet's disease and Hughes-Stovin syndrome(HSS) contribute to the greatest number of cases. They are autoimmune conditions causing vasculitis and the formation of aneurysms in multiple organs including the pulmonary artery.

On the other hand, Takayasu arteritis is a chronic inflammatory disease that affects large vessels including the pulmonary vessels ultimately leading to aneurysm formation.

- **Infections:** Syphilis and tuberculosis are identified as the leading causes of PAA formation. Syphilis: It is caused by Treponema pallidum and can lead to tertiary syphilis with vascular involvement, including pulmonary artery aneurysms. Tuberculosis: Severe or disseminated tuberculous infection can cause tuberculous arteritis affecting the pulmonary artery. Pseudoaneurysms that are secondary to tuberculosis are commonly referred to as Rasmussen aneurysms.(5)

Although rare other infections include bacterial infections like infective endocarditis, bacterial pneumonia, lung abscess, and staphylococcus bacteremia. Some fungal infections like candida, mucormycosis, aspergillus, and parasitic infections like schistosomiasis are also responsible for PAA.

- **Trauma:** Blunt and penetrating trauma involving the chest can result in injury to the pulmonary artery which is quite life-threatening and lead to aneurysm formation.

1) **Miscellaneous causes:** Pulmonary embolism: recurrent or severe pulmonary emboli can cause chronic changes in pulmonary vascular resistance leading to aneurysm formation. Radiation Exposure: this may be the result of high-dose radiation therapy to the chest for conditions like lung cancer leading to vascular damage and aneurysm formation in the pulmonary artery.

- **Idiopathic:** In some cases, the cause of pulmonary artery aneurysm can not be ascertained however, it has been postulated that the cause is secondary to congenital weakness of the vessel wall and/or cystic medial degeneration of the vessel wall.(6)

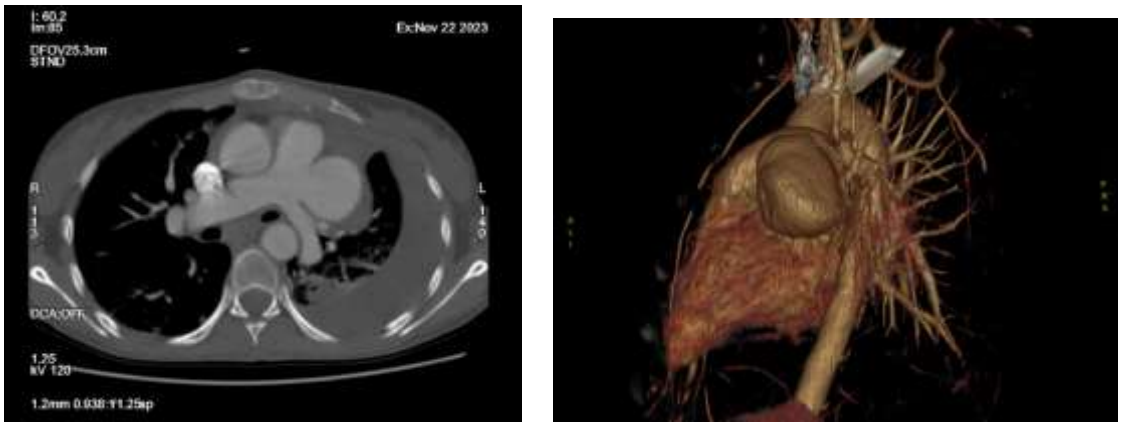

Fig 1: Historical form of a PPA in one of our patients

A- Fissured PPA with hemothorax and hemopericarditis

B- Huge PPA in CT reconstruction view

3. CLINICAL MANIFESTATIONS:

As we have previously discussed pulmonary artery aneurysm is a rare but fatal condition, it presents a typical set of clinical manifestations that require immediate attention. However clinical manifestations of both PAA and PAPA are nonspecific and can be noticed in many other clinical conditions, It is characterized by abnormal dilation of the pulmonary artery, and it can lead to various symptoms depending on its size and location. Some of the commonly observed clinical manifestations are shortness of breath, chest pain, and coughing which is often accompanied by haemoptysis.(7)

Additional symptoms include palpitations, dizziness, or syncope (fainting episodes) due to the proximity of the pulmonary artery to the heart.

In more severe and critical cases, the aneurysm may compress neighbouring structures resulting in symptoms such as hoarseness of voice and difficulty in swallowing.(8)

4. DIAGNOSIS:

The diagnosis of PAA and PAPA is very challenging and it usually involves imaging modalities like CT scans or MRI scans which are then followed by treatment directed at treating and managing the symptoms and focusing on the underlying cause which may involve surgical intervention either to repair or replace the arterial segment that is affected. However, the gold standard for both detection and follow-up of PAA and PAPA is Computed tomography angiography (CTA)(9).

Due to non-specific symptoms of PAA and PAPA, CTA focus can be placed on the pulmonary arterial system or the aorta and bronchial artery system.

Catheter-directed angiography has been considered the gold standard for the diagnosis which not only helps in the determination of vascular involvement but also the assessment of right-sided cardiac pressures(10). In addition, prompt endovascular treatment can be performed. However, the role of noninvasive imaging modalities such as CT and MRI is expanding due to the advancement in technologies.

- **IMAGING FINDINGS:**

Although pulmonary artery aneurysms are quite rare various imaging techniques aid in the diagnosis of PAA. Some of the common imaging findings associated with PAA are discussed below:

1) Chest X-ray (CXR): A CXR usually shows an enlargement of the pulmonary arteries, although it is non–specific for aneurysm it can provide some clues for the need for further imaging and workup.
2) CT Angiography (CTA): It is one of the most efficacious imaging tools for detecting Pulmonary artery aneurysms as it provides detailed information regarding the size, location, and shape of the aneurysm along with its relationship to surrounding structures.
3) Magnetic Resonance Imaging (MRI): It usually provides high-resolution images and provides useful information on arterial wall thickening, and blood flow and specifies aortic and pulmonary hemodynamics without any radiation exposure.
4) Echocardiography: Transthoracic or transesophageal echocardiography may show the presence of PAA. Although it is not as sensitive as CTA or MRI, it may help in detecting pulmonary artery aneurysms especially if they are large or located near the heart.
5) Pulmonary Angiography: This is an invasive procedure in which dye is injected into the pulmonary arteries and then X-ray images are obtained. It can give proper details and information about the anatomy of the aneurysm especially where the information provided by other imaging modalities is inconclusive and further detail is required for treatment planning.

5. DIFFERENTIAL DIAGNOSIS:

The list for the differential diagnosis of pulmonary artery aneurysm is quite broad and includes various conditions that usually share similar symptoms or imaging findings. Here are some of the key differential diagnoses which should be kept in mind:

- Pulmonary Embolism (PE): It usually results when a blood clot which originated somewhere else, travels to the lungs and blocks a pulmonary artery or any of its branches. Although it is not an aneurysm it may cause dilation and distortion of the pulmonary artery which on imaging may be mistaken for an aneurysm. However, it cannot be denied that chronic pulmonary embolism is another relatively common cause of pulmonary artery aneurysm.
- Pulmonary Arteriovenous Malformation (PAVM): These are rare vascular anomalies of the lungs resulting from an abnormal connection between a pulmonary artery and vein leading to shunting of blood. PAVMs may sometimes mimic the findings of pulmonary artery aneurysms in imaging studies.
- Pulmonary Hypertension: Enlargement or remodeling of the pulmonary arteries may result from chronic pulmonary hypertension. In severe cases, they may resemble aneurysms in imaging studies due to dilation of the pulmonary arteries.
- Vasculitis: Some conditions including Behcet's disease, Hughes-Stovin syndrome, Takayasu arteritis, and systemic lupus erythematosus (SLE) may result in inflammation and weakening of the wall of blood vessels resulting in aneurysm formation.
- Trauma: Traumatic injuries resulting from blunt or penetrating trauma to the chest can result in vascular injuries leading to the formation of pseudoaneurysms of the pulmonary arteries, which on imaging may resemble true aneurysms.
- Neoplastic Lesions: Abnormal masses or tumors near or within the pulmonary arteries can sometimes mimic pulmonary artery aneurysms in imaging studies.

6. TREATMENT:

Determining the appropriate treatment remains extremely challenging once a PAA or PAPA is diagnosed. The treatment must be directed to the underlying cause while using the least invasive procedure and achieving promising results which is quite difficult. Depending on the severity of the disease and co-morbidities, there are two kinds of treatment approaches including conservative and surgical approaches.

1) **Conservative Treatment:**

It includes medical management of the underlying disease like pulmonary hypertension along with the routine radiological follow-up of PAA. Conservative treatment seems favorable for asymptomatic patients or patients with normal pulmonary artery pressure.

In the case of PAH, calcium channel blockers, diuretics, and anticoagulants must be included in the treatment, and in some cases use of vasoactive substances such as endothelin receptor antagonists, phosphodiesterase type 5 inhibitors and prostacyclin derivatives may be beneficial. However, in general, patients with PAH should be thoroughly evaluated for surgical treatment owing to the risk of impending dissection and rupture (11).

Interventional treatment is a relatively new treatment option for PAA's and coil embolization seems to be a good treatment option for iatrogenic causes and small branches.(9)

1. **Surgical treatment:**

Without any doubt surgery remains the keystone of therapy for lesions which are involving the main pulmonary trunk. According to the clinical experience and going through all the available data about aortic aneurysms, it is suggested to operate on all the patients with pulmonary trunk aneurysms with a diameter of >5.5cm according to the guidelines of aortic disease.(12)

Aneurysmorrhaphy is a simple procedure for surgical repair of PAA, although it only decreases the diameter of the vessel wall and does not treat the abnormal vessel wall which may increase overall wall stress.(13) However, nowadays aneurysmectomy and repair or replacement of the right ventricular outflow tract seems to be the method of choice (14, 15) and it is the only feasible treatment for patients with connective tissue disorders.(15)

As compared to proximally located aneurysms, the treatment of distal aneurysms may be more challenging and may require lung resection, therefore it is considered more fatal. (16)Lung or complete heart-lung transplantation is the ultimate treatment, especially in patients with PAH. (17, 18).

7. CONCLUSION:

In conclusion, pulmonary artery aneurysm are although quite rare but may result in serious complications that require prompt diagnosis and treatment. They can be secondary to various underlying causes including congenital, connective tissue disorders, infections and trauma. Management is typically aimed at treating the underlying cause, monitoring for complications like thrombosis or rupture and identifying the need for prompt surgical intervention in certain cases to avoid potentially life-threatening consequences. Hence early detection and timely management are mandatory for optimizing outcomes in patients with pulmonary artery aneurysm.

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