

Giant Pulmonary Artery Aneurysm Associated to severe mitral stenosis: Review and case report

Abstract:

Pulmonary aneurysms are rare abnormalities of the pulmonary vessels that are often associated with pulmonary hypertension. However, the occurrence and treatment of this pathology are still unclear. A 33-year-old patient with a medical history of transcatheter mitral commissurotomy was admitted with NYHA class III dyspnea and lower extremity edema. The clinical examination found the semiology of mitral stenosis and signs of both right and left heart failure. The electrocardiogram showed complete atrial fibrillation and right ventricle hypertrophy. Chest X-ray revealed cardiac hypertrophy and a prominent aspect of the left median arch reminiscent of a left pulmonary aneurysm. Doppler echocardiography found pure severe mitral stenosis (mitral valve area = 0.8 cm²), dilation of the pulmonary trunk (diameter = 74 mm), and its branches. The right heart chamber was also dilated, with significant tricuspid regurgitation and severe pulmonary hypertension (systolic pulmonary artery = 95 mmHg). The thoracic angioscan angiography revealed aneurysmal dilation of the trunk of the pulmonary artery and its branches without dilation of their distal parts (diameter of the pulmonary artery = 76 mm; Right pulmonary artery diameter = 51 mm, left pulmonary artery diameter = 40 mm). showed aneurysmal expansion of the pulmonary trunk and its branches without expansion of the distal portion (pulmonary artery diameter = 76 mm, right pulmonary artery diameter = 51 mm, left pulmonary artery diameter = 40 mm). Clinical symptoms improved with diuretic, anticoagulant, and analgesic treatment, and the patient was subsequently referred to cardiovascular surgery for mechanical mitral valve replacement and plastic surgery of the pulmonary artery. Pulmonary aneurysms are uncommon pathology that can be potentially life-threatening. Its clinical manifestations are usually nonspecific or asymptomatic and the appropriate treatment is a challenge, as there are no clear guidelines on the recommended treatment.

Keywords: Pulmonary artery aneurysm, mitral stenosis, pulmonary hypertension, case report

Introduction:

Pulmonary aneurysms represent uncommon abnormalities of the pulmonary arteries that can be potentially life-threatening. Patients often present with nonspecific symptoms, and the diagnosis may be incidental. Consensus on diagnostic criteria and imaging follow-up for patients diagnosed with this entity is limited. Additionally, treatment strategies vary according to underlying disease, etiology, center-specific expertise, and available resources.

This article aims to report our rare case and review the pertinent literature.

Case Presentation:

A 33-year-old woman with a medical history of transcatheter mitral commissurotomy for severe mitral stenosis 8 years ago, presented with worsening dyspnea associated with lower-extremity edema that appeared 2 weeks before her admission.

The general examination found a conscious patient. Her heart rate was 94 b/m, and her blood pressure was 110/68 mm Hg. She was polygenic with a respiratory rate of 20 breaths/min, and an Oxygen saturation of 97% on ambient air. Physical examination revealed infundibula pulmonary shock, apex diastolic rolling of 4/6th intensity with fine bibasilar crackles, positive spontaneous turgidity of the jugular veins, and pitting edema involving her lower extremities bilaterally. Subsequent physical examination was normal.

Her blood test did not reveal any abnormalities.

The electrocardiogram showed an arrhythmia complete by atrial fibrillation with an average ventricular rate of 94 cycles per minute, a QRS axis at $+110^\circ$, and a right ventricular preponderance.

Chest X-ray (Fig. 1) demonstrated cardiomegaly (with a cardiothoracic ratio of 67%) due to the expense of the right heart chambers and the left atrium, a double contour aspect of the lower right arch, a prominent aspect of the left middle arch, large vascular hilum and rarefaction of the vascular network at the periphery.



Fig. 1: Chest X-ray showing cardiomegaly with the prominent middle left arc and double outline of the lower right arc.

The echocardiography (Fig. 2) found mitral valves thickened, very altered, and of limited opening (mitral area = 0.8 cm² by mean gradient planimetry OG-LV = 17 mmHg) without associated mitral regurgitation. The aortic sigmoids were remodeled but of correct opening without aortic regurgitation. Significant tricuspid insufficiency with Major PAH (95 mmHg). A significant expansion of both the left and right atrium. Dilation size of the right ventricle. The pulmonary artery is dilated (diameter = 74 mm in parasternal section left centered on the pulmonary artery,

the diameter ratio of pulmonary artery/aorta diameter = 2.5) as well as its two branches without contrast in the pulmonary artery or its branches.

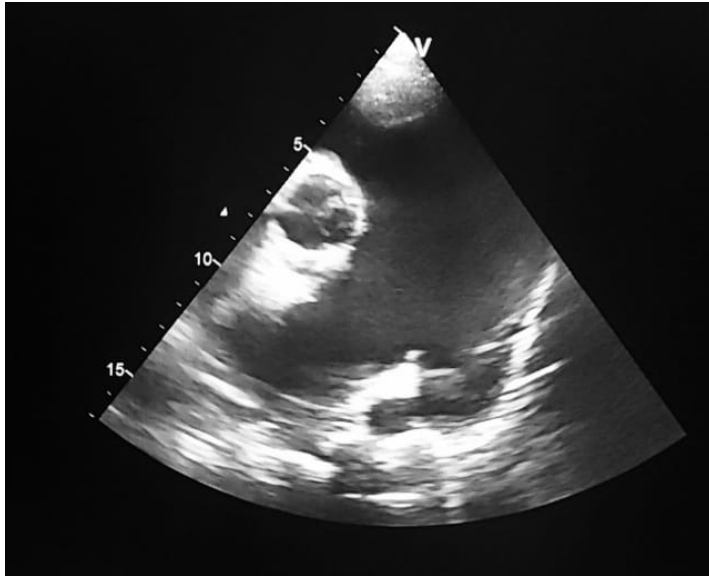


Fig. 2: ETT 2D short-axis parasternal view centered on the pulmonary artery showing significant dilation of the trunk of the pulmonary artery and its branches.

Chest CT angiography (Fig. 3 and 4) revealed aneurysmal dilation of the pulmonary artery and its Branches without dilation of their distal parts (diameter of the pulmonary artery = 76 mm; Right pulmonary artery diameter = 51 mm, left pulmonary artery diameter = 40 mm).



Fig. 3: Chest CT showing aneurysmal dilatation of the trunk of the artery lung and its branches.

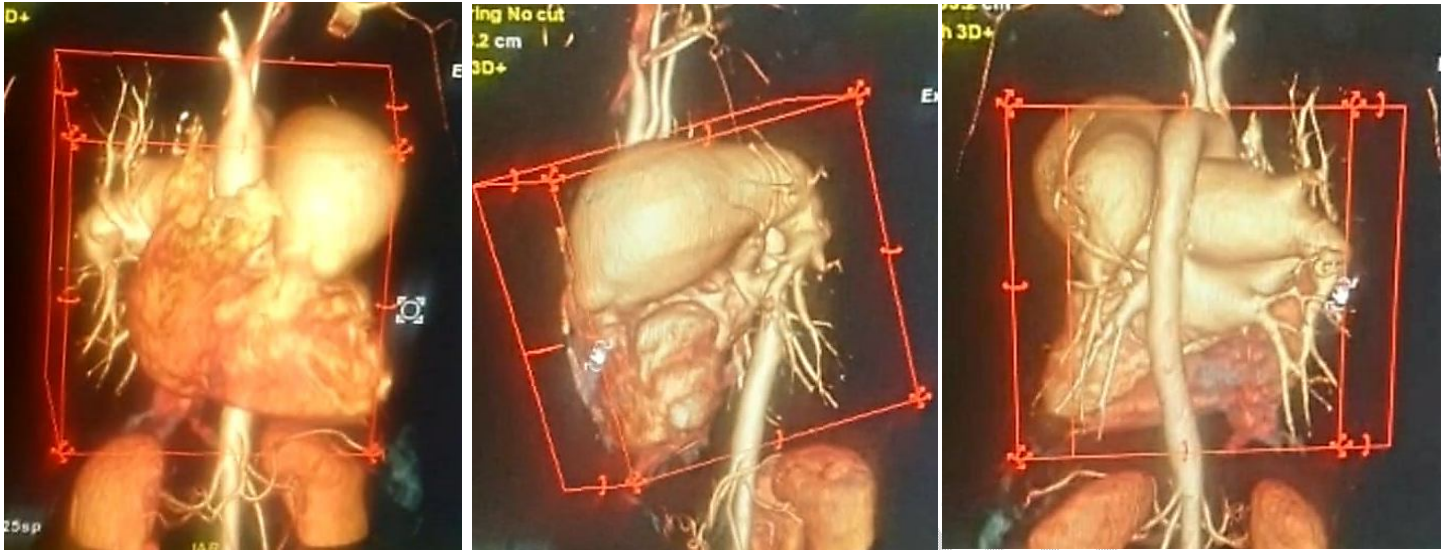


Fig. 4: Three-dimensional reconstruction of the heart and the massively dilated trunk of the pulmonary artery and its branches main pulmonary artery

Good evolution was noted under diuretic, anticoagulant, and analgesic treatment, and the patient was referred to the cardiovascular surgery department for mechanical mitral valve replacement and plastic surgery of the pulmonary artery.

Definition and etiologies:

Pulmonary aneurysm incidence has been estimated to be 1 in 14,000 autopsies [1]. In contrast to aortic aneurysms, PA aneurysms do not have a clear definition. There was no standard diameter size parameter to define PAA. Most commonly, PAA is defined as dilation greater than 40 mm. A main pulmonary aneurysm can be defined, based on the recent literature, as a dilatation greater than 1.5 times the upper limit of normal. This corresponds to 43.4 mm for males and 40.4 mm for females. The maximum MPA diameter was 106–170 mm [2]. The frequency of pulmonary aneurysms (PAA) is independent of age and gender [3].

“An aneurysm can be a true aneurysm or a pseudoaneurysm. By definition, a true aneurysm is a local dilation of a vessel that affects all three layers of the vessel wall: Intima, media, and adventitia. Otherwise, pseudoaneurysms do not affect all arterial walls and are at increased risk of rupture” [4].

“These lesions occur in 70% of central aneurysms involving the pulmonary trunk, right or left main pulmonary artery, and in 30% of peripheral aneurysms originating from segmental or intrapulmonary branches” [1].

The pathophysiology of PA aneurysms is associated with stress on the vessel wall, which can lead to progressive vasodilatation and rupture, resulting in very high mortality (50-100%) [4]. Aneurysm dissection, airway compression, and pulmonary artery thrombosis are other serious complications of large PA aneurysms.

“Clinical manifestations of PAA are usually nonspecific or asymptomatic” [5]. “Most patients present with unremarkable symptoms such as dyspnea, cough, chest pain, and hemoptysis and are referred to imaging studies for vascular dilatation” [4]. Symptoms of heart failure may also be

observed secondary to pulmonary insufficiency, tracheal or bronchial compression, or pulmonary embolism due to her enlarged MPA [3,6].

Table 1: PAA etiology can be congenital or acquired [5,7]

CONGENITAL	ACQUIRED	
<p>*Increased pulmonary blood flow (Eisenmenger's syndrome)</p> <p>*Heart defects (Pulmonary valvular abnormalities, Ductus arteriosus, Atrial septal defects, Ventricular septal defects, Hypoplastic aortic valve)</p> <p>*Connective tissue abnormalities (Ehlers–Danlos syndrome, Marfan syndrome, Cystic medial necrosis)</p>	True aneurysms	Pseudoaneurysms
	<p>*Pulmonary arterial hypertension related</p> <p>Lung conditions other than pulmonary hypertension: Bronchiectasis and pulmonary; fibrosis; Interstitial lung diseases COPD</p> <p>*Pulmonary artery hypertension</p> <p>*Vasculitis: Behcet's disease</p> <p>*Idiopathic</p>	<p>*Post-infectious (Endocarditis Tuberculosis, Syphilis, Pyogenic bacteria, Pneumonia)</p> <p>*Malignancy related: Metastasis, Primary lung cancer</p> <p>*Iatrogenic: Cardiac surgery; Chest tube placement; Lung biopsy; PA catheter placement; PA arteriography; Lung resection; Radiation in the past</p> <p>*Traumatic</p>

Diagnosis:

The physical examination can detect a systolic murmur combined with a diastolic murmur due to pulmonary regurgitation [6]. The ECG is usually normal, but right axis deviation or bundle branch block may be present [8].

A chest x-ray shows enlarged PA, hilar enlargement, pulmonary nodule, or lung mass. An accurate echocardiogram focusing on the RV outflow tract/PA view allows early diagnosis.

"The value of echocardiography in the diagnosis of peripheral aneurysms is limited, with an echocardiographic reference value of 26 mm for MPA dilatation" [6]. "Cardiac CT helps to assess pulmonary artery size and rule out causes of dilated pulmonary arteries. Because of its high spatial resolution, contrast-enhanced CT offers a unique opportunity to assess the presence, size, shape, precise location, and associated cardiovascular abnormalities of aneurysms and is, therefore, the leading technique for diagnosing pulmonary artery dilatation. is considered. The upper limit of the normal diameter of the main pulmonary artery on computed tomography is 29 mm and that of the right interlobar artery is 17 mm" [9]. "CT angiography is a superior modality because it can easily identify mural thrombi, dissections, or other abnormalities. Multiplanar reconstruction is useful for surgical planning" [8]. "In addition to cardiac computed tomography angiography, magnetic resonance imaging also confirms the diagnosis and provides additional information regarding the size, number, location, and extent of the PAA, especially when detecting the possibility of an intimal flap. Considered the gold standard for delivery. It shows arterial wall thickening in connective tissue disease and also provides hemodynamic information in dilation after stenosis due to pulmonary valve disease" [10]. "CMR is a useful noninvasive modality for quantifying pressure gradients in pulmonary artery stenosis and detecting other causes of dilated pulmonary arteries without the need for contrast agents or radiation exposure" [8].

Cardiac catheterization is considered essential for diagnosis, helping to rule out causes of dilated pulmonary arteries and to determine intracardiac pressure, which is diagnostically important [8].

Treatment:

After a pulmonary aneurysm is diagnosed, it is difficult to determine the appropriate treatment, as there are no clear guidelines for recommended treatment. As previously mentioned, PAA has multiple etiologies and therapeutic strategies vary and are based on underlying etiology, hemodynamics, and associated complications.

Most PAA appears to be relatively benign, especially if they remain asymptomatic. On the other hand, PAA anatomy is associated with poor prognosis. There are still no clear risk factors for PAA dissociation and rupture. Analysis of case reports found that predictors of high risk were:

Rapid progression of PAA diameter (>2 mm/year), tissue wasting due to infection and/or pregnancy, PA diameter >75 mm, or PA systolic pressure >50 mm Hg [2].

Conservative treatment options include targeted therapy of underlying disease, treatment of pulmonary hypertension, and regular radiological follow-up for PAA. Surgical techniques such as aneurysm, lobectomy, bilobectomy, aneurysmectomy, and pneumonectomy have been described in the literature, but are associated with increased risk of morbidity and mortality, especially in patients with pulmonary hypertension. Endovascular interventions such as coil embolization and vascular plugging are excellent treatments, but there are no specific guidelines for their use [7].

Conservative treatment should be selected in asymptomatic patients without pulmonary hypertension or unstable PAA size. In case of vasculitis or infectious etiology, causative treatment should be assigned. Interventional treatments such as spiral embolization and vascular occlusion are treatment options for iatrogenic causes and small branches [7,11].

Symptomatic patients or those with accelerated PAA increase with pulmonary hypertension (defined as RVSP >35 mmHg) should undergo surgery [12,13]. Asymptomatic patients with a PAA ≥80 mm in diameter should also be referred for cardiac surgery [6,12,13].

This raises the question of when to choose risky procedures and when to choose conservative treatments. There is no consensus on how to treat PAA and currently, there are no treatment guidelines outlining the choice between risky procedures and conservative management. However, some authors have proposed management algorithms. The management scheme proposed by Reisenauer [12] is described below (Fig 5).

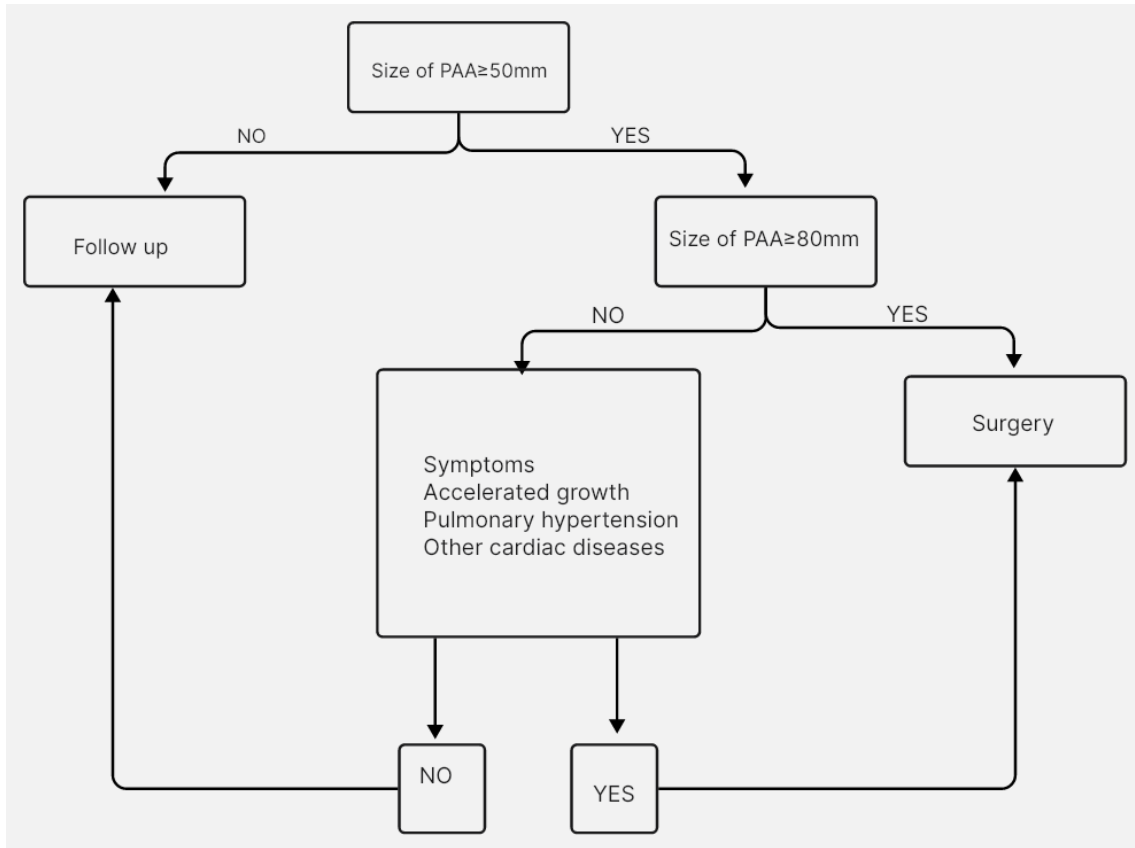


Figure 5: Algorithm for management of pulmonary artery aneurysm proposed by Reisenauer.

Gupta and his team proposed another management algorithm, shown in the figure below, based on the experience of the medical school of Monmouth Medical Center in the United States [7]:

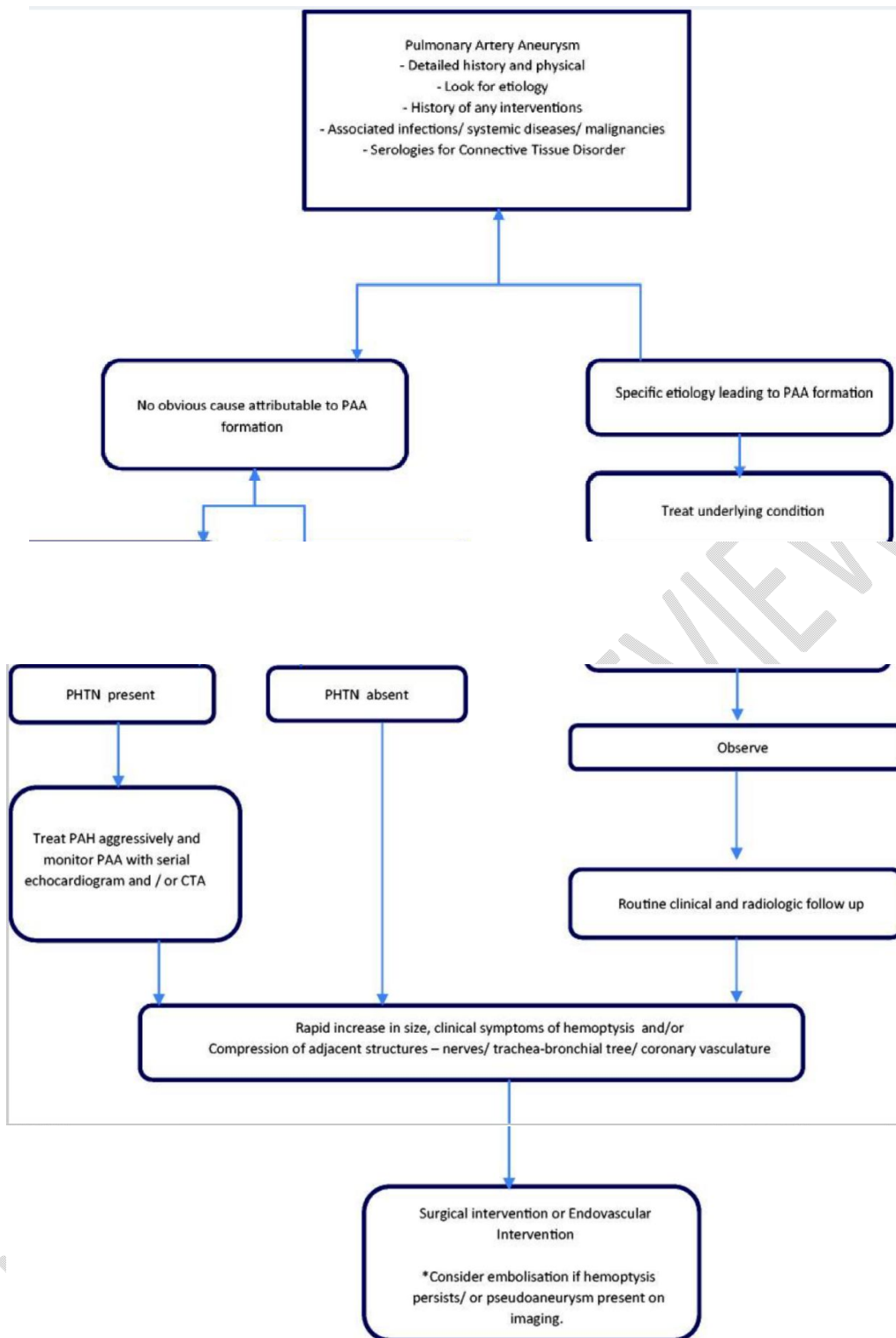


Figure 6: Algorithm for management of pulmonary artery aneurysm proposed by Gupta. PAH: pulmonary arterial hypertension; PAA: pulmonary artery aneurysm; PHTN: pulmonary hypertension; CTA: computed tomography angiography. *indicates a special point to be considered.

Outcomes:

Higher risk of sudden death, left main coronary artery compression, right pulmonary artery thrombosis, pulmonary compression, and atelectasis are associated with pulmonary artery

diameters greater than 55 mm and high PPA pressures [2]. PAA rupture and PAA dissection with pulmonary hemorrhage are major complications with a fatal prognosis [14].

“Because PAA is rare and uncommon, data on surgical outcomes are scarce. However, perioperative morbidity is similar to that associated with the repair of ascending aortic aneurysms. Difficult ventilation is the most important postoperative problem” [11]. “Endovascular treatment of PAA has fewer inherent risks than surgical procedures, but risks remain similar to other systemic endovascular embolization procedures. These risks include contrast nephropathy, non-targeted embolization, arterial dissection, arterial thrombosis, and partial or total end-organ infarction” [3].

Discussion:

Proximal pulmonary artery aneurysms with a pulmonary-to-aortic diameter ratio greater than 2 are rare, and bilateral aneurysms have been described exceptionally [15]. Clinical manifestations are nonspecific and rare. PAA may remain asymptomatic and the diagnosis in this case is made incidentally on a chest x-ray showing dilation of the pulmonary arch. Sudden death can also be a diagnostic situation [16]. The patient's symptoms of exacerbation and lower extremity edema were most likely secondary to left heart valve disease, represented by significant mitral stenosis requiring surgical intervention. The etiology of PAA is dominated by bacterial (syphilis, tuberculosis) or fungal infections, inflammatory arteritis (giant cell arteritis, Behcet), congenital or acquired heart disease, and valvular disease. Causes of trauma are also mentioned. The primitive type is the exception and is associated with cardiac defects in half the cases.

Persistent ductus arteriosus, atrial septal defect or interventricular defect [7]. Chronic PAH can cause pulmonary aneurysms by directly attacking the wall with atherosclerosis and medial necrosis, leading to aneurysmal dilatation [17]. However, it has not yet been proven that treatment to lower pulmonary artery pressure can have a significant impact on the lack of progression of pulmonary hypertension [8]. For this patient, the most likely mechanism of PA dilatation was pulmonary hypertension secondary to severe mitral stenosis.

Many complications are observed, including pulmonary embolism, aneurysm dissection or rupture, right heart failure, left coronary trunk, right pulmonary vein, superior vena cava, tracheal compression, and recurrence [19].

If these aneurysms are found, surgical repair (dacron insertion [Dupont], homograft, prosthesis or pericardial patch reconstruction, aneurysm, or angioplasty) is recommended [20]. Most commonly, surgery should be suggested if the risk of surgery to the patient is sufficiently low, if there is a gradual increase in pulmonary artery diameter, or if there is a pulmonary artery dissection [21]. Considering the risk of cardiac surgery, repairing an aneurysm was judged to be the most appropriate treatment for the above patient to prevent arterial dilatation due to future mitral valve replacement surgery. A Japanese study found that areas of PA aneurysms have increased expression of the EP4 receptor (cyclooxygenase-2-dependent PGE2) compared to non-PAA areas [18]. Increased expression of this receptor has also been reported in areas of non-striated muscle cells, macrophages, and abdominal aortic aneurysms [22,23]. The activity of metalloproteinase 2 and the production of interleukin 6 is increased by PGE2 through the EP4 receptor, causing elastic fiber breakdown and thus progression of AAA [22, 24]. Non-steroidal anti-inflammatory drugs and selective COX-2 inhibitors suppress PGE2 synthesis. However, these drugs should not be used long-term because of side effects (such as gastrointestinal bleeding with NSAIDs and increased risk of cardiovascular events with coxibs). An interesting goal of drug therapy is to inhibit aneurysm

progression via selective EP4 receptor antagonists, which may be one of the future therapeutic options [5].

Conclusion:

PAA is rarely diagnosed because most patients are asymptomatic or exhibit nonspecific symptoms. To date, due to the low prevalence of PA aneurysms, there are no guidelines for diagnosis, treatment, or follow-up of these patients. Surgical treatment should be considered for pulmonary artery dilatation greater than 80 mm, accelerated aneurysm growth, pulmonary hypertension, and other concomitant heart disease. However, increasing survival while minimizing procedure-related morbidity and mortality requires a multidisciplinary action plan.

Consent

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

Ethical approval

The study was exempt from ethical approval in our institution.

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