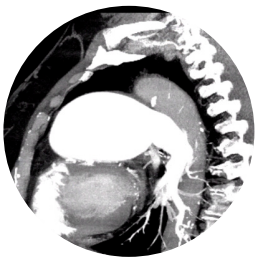

Pulmonary artery aneurysm: a review and two illustrative cases

Aneurisma de arteria pulmonar: una revisión y dos casos ilustrativos

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Summary

Pulmonary artery aneurysm (PAA) is a rare disease with an estimated prevalence of 0.06%. Although its pathophysiological mechanisms are still under study, the main cause is believed to be an increased pressure and/or flow through the pulmonary artery (PA), as occurs in pulmonary hypertension and Eisenmenger syndrome. PAA often has an asymptomatic course or triggers nonspecific symptoms, but its complications can occur regardless of the symptoms and be fatal. In this article, a literature review is carried out and two cases of PAA are reported.

Resumen

El aneurisma de la arteria pulmonar (AAP) es una patología poco frecuente, con una prevalencia estimada en 0,06 %; si bien sus mecanismos fisiopatológicos aún se encuentran en estudio, se cree que la principal causa es el incremento de la presión y/o del flujo a través de la arteria pulmonar (AP), como sucede en la hipertensión pulmonar y el síndrome de Eisenmenger. Con frecuencia el AAP tiene un curso asintomático o desencadena síntomas inespecíficos, pero sus complicaciones pueden presentarse con independencia de los síntomas y llegar a tener un desenlace fatal. En el presente artículo se realiza una revisión de la literatura y se presentan dos casos de AAP.

Introduction

Un A true aneurysm is the focal dilatation of a blood vessel that involves the three layers that make up its wall (1), unlike pseudoaneurysms that do not comply with this rule. Aneurysmal processes of the pulmonary artery (PAA) occur in the main trunk, which normally has a diameter of up to 29 mm in men and 27 mm in women, and/or in some of its branches (2). Despite being a pathology known for more than a hundred years, there are still discrepancies in the literature on the parameters to define the presence of a PAA; some authors consider PAA when it is greater than 40 mm (3), and others take into account a truncal diameter greater than 45 mm and/or greater than 30 mm in one of the branches of the pulmonary artery (PA) (4). However, there is a partial consensus that a dilatation greater than 1.5 times the upper limit of normal (43.4 mm in men and 40.4 mm in women) is diagnostic of this pathology (5).

PAA are rare, their global epidemiology is unknown. A study with more than 100,000 autopsies identified eight cases, which shows a ratio of 1 in 13,696, and a prevalence of 0.06% (6), which is low compared to abdominal aortic aneurysm (2-7% in men and 1% in women over 65 years) (7) and thoracic aortic aneurysm (0.16%) (8). It has been identified that PAA can be idiopathic or secondary to various diseases, including congenital heart disease, bacterial and mycobacterial infections, Marfan syndrome and pulmonary hypertension, among others (9); however, a significantly higher incidence (1.25%) is observed in patients with severe pulmonary hypertension of any cause and in patients with Eisenmenger syndrome (6%) (4).

Although its prevalence is low, the mortality rate associated with PAA rupture has been estimated between 50% and 100%, resulting in death by aspiration and asphyxia after intrapulmonary hemorrhage; in addition, PAA can lead to PA dissection and sudden death even in asymptomatic patients (10, 11). In this article, a review of the literature and two cases of PAA are presented.

Pathophysiology

Although there is little information on the pathophysiology of PAA, it has been proposed that structural changes in collagen and elastin of the arterial walls may play an important role, especially under the influence of elevated pulmonary sphenoidal pressure (1). It has

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been observed that both pulmonary arterial hypertension and thoracic aortic aneurysm share molecular cascades that favor changes in the extracellular matrix and vascular remodeling, so there could be some common mechanisms between the formation of thoracic aortic aneurysms and PA aneurysms, taking into account the association between pulmonary arterial hypertension and PAA (12).

Classification

PAAs are classified according to the pressure in the pulmonary artery, into high and low pressure aneurysms, and are subdivided according to their respective etiologies (2).

- High pressure:
  - Pulmonary hypertension associated with congenital heart disease
  - Other types of pulmonary hypertension.
- Low pressure:
  - PA dilatation with tissue abnormalities, such as genetic, infectious and inflammatory factors
  - Idiopathic and post-stenotic PA dilatation
  - PA dilatation caused by high flow through the PA, such as severe pulmonary valve insufficiency
  - Restrepo and Carswell (4) propose another classification according to the type of vessel affected, in which they divide PAA into congenital and acquired causes.

Clinical manifestations

PAAs are usually asymptomatic, therefore, their finding is usually incidental through diagnostic images in the study of other pathologies (1), as in the cases that will be presented later in this article.

The symptoms of a PAA, if present, are indeterminate, especially chest pain, cough, dyspnea, palpitations, hemoptysis, central cyanosis, and other less common symptoms such as systolic and/or diastolic murmur in the left third intercostal space, ventricular hypertrophy, tricuspid regurgitation, syncopal episodes and multiple pulmonary embolisms (1).

Hemoptysis is considered a sign of poor prognosis, since it may indicate imminent rupture of the aneurysm, a situation in which lethal bleeding, asphyxia and sudden death occur, requiring immediate intervention (1, 13, 14).

Diagnosis

The diagnosis of PAA is usually made at necropsy or on diagnostic imaging, usually requested for another reason. The radiological features on chest radiography are non-specific, and the most suggestive finding is the prominence of the pulmonary hilum; although indirect signs, such as the enlargement of right heart cavities, can also be found (14).

The echocardiogram may reveal an increase in the diameter of the PA trunk greater than the upper limit of normality observed in the right ventricular outflow tract/PA window, as well as indirect signs of the presence of a PAA (2).

Contrast angiotomography and cardioresonance imaging allow a detailed appreciation of the PA anatomy and are therefore the ideal methods to determine its diameter (1, 14).

Treatment

Treatment should be oriented to the etiology of the PAA, prioritizing the least invasive with the most durable results. Conservative management can be chosen, which includes radiological follow-up of the PAA; or surgical management, which has various techniques that, however, carries a higher risk of morbidity and mortality, especially in patients with pulmonary hypertension or multiple comorbidities (5, 10). Conservative management may be preferable in asymptomatic patients; however, if the disease progresses or symptoms appear, changes in treatment should be reevaluated (1).

Prognosis

PAAs are an unusual vascular anomaly, difficult to diagnose and of uncertain prognosis, but with potential risk of fatal complications. A critical diameter that suggests imminent rupture of the PAA or other complications has not been determined, so that the conduct to be followed must take into account the etiology, size, presence of pulmonary hypertension and the patient's clinical condition (15).

It is necessary to emphasize that this disease can be a direct consequence of pulmonary hypertension and Eisenmenger's syndrome, and one of its possible complications is pulmonary embolism (4).

As we have seen, it is essential to have the appropriate tools for its diagnosis and timely management, based on the best available scientific evidence and, in the absence of clear recommendations in this regard, individualization of each patient.

Case 1

A 77-year-old female patient attended the emergency department for a clinical picture of two weeks of evolution consisting of progressive dyspnea that developed into orthopnea two days before the consultation. The patient has a history of severe group 1 pulmonary hypertension, atrial septal defect, permanent atrial fibrillation, severe left ventricular hypertrophy, hypothyroidism in substitution, and overweight, pharmacologically managed with bosentan, sildenafil, dabigatran and levothyroxine.

On admission, the patient has peripheral oxygen saturation of 76%. As positive data, on physical examination the patient is found with polypnea, arrhythmic heart sounds, symmetrical edema of lower limbs grade I, and acropaquia.

A portable chest X-ray in anteroposterior projection shows widening of the vascular pedicle, prominence of pulmonary hilarity and global cardiomegaly. In addition, he observed a homogeneous rounded opacity in the middle third of the right pulmonary parenchyma with a diameter of 77 × 56 mm (Figure 1).

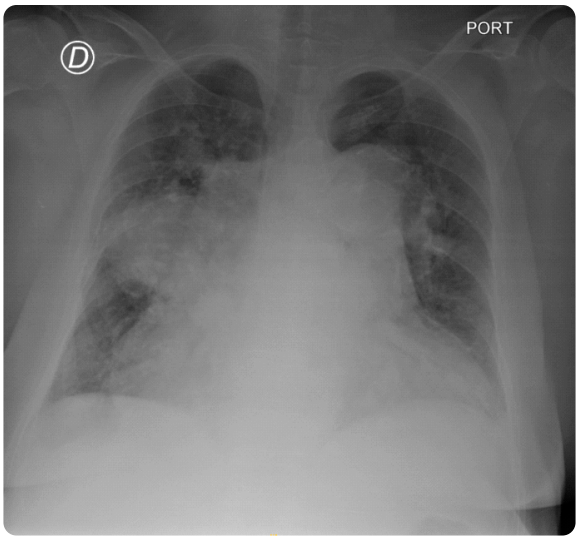

Figure 1. Portable chest X-ray in anteroposterior projection: widening of the vascular pedicle, prominence of the pulmonary hilae and global cardiomegaly. Additionally, a homogeneous rounded opacity is observed in the middle third of the right lung parenchyma with a diameter of 77 × 56 mm.

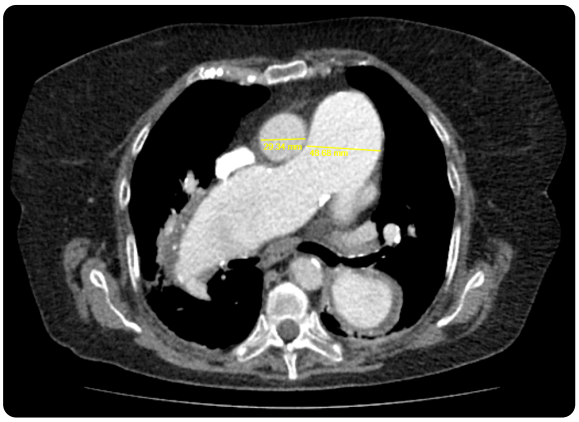

Figure 2. Axial CT, with contrast medium in mediastinal window: dilatation of the PA trunk (48 mm).

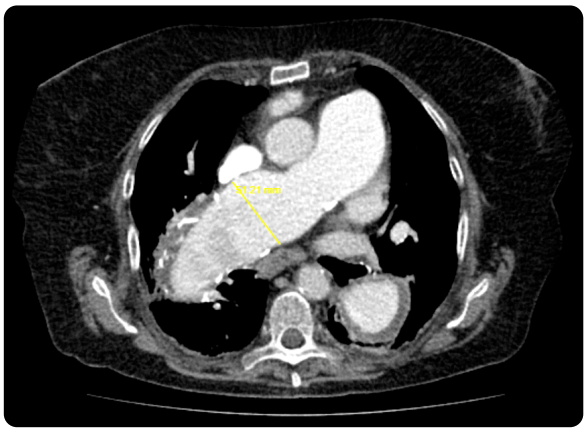

Figure 3. Chest CT, axial, with contrast medium in mediastinal window: dilatation of the right branch of the pulmonary artery (52 mm), with eccentric opacification defect related to changes due to chronic pulmonary embolism.

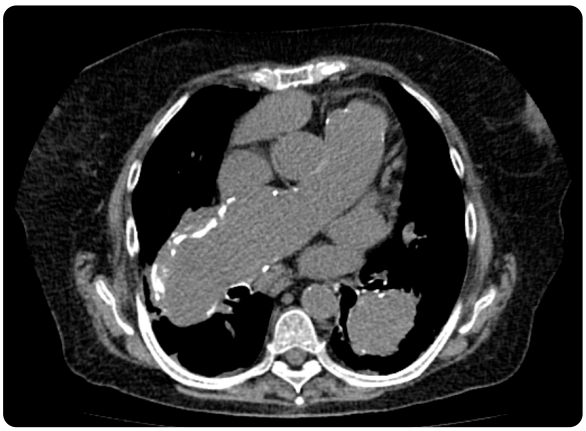

Figure 4. Axial CT in simple phase in mediastinal window: dilatation of the right branch of the pulmonary artery and linear calcifications of the arterial walls as signs of chronic pulmonary embolism.

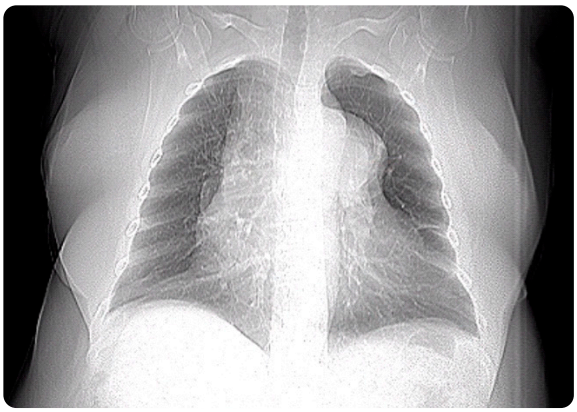

Figure 5. Chest angiotomography scout view image in anteroposterior projection: cardiomegaly, mediastinal widening, dilatation of the PA trunk and prominence of the left pulmonary hilum are identified as indirect signs of pulmonary hypertension.

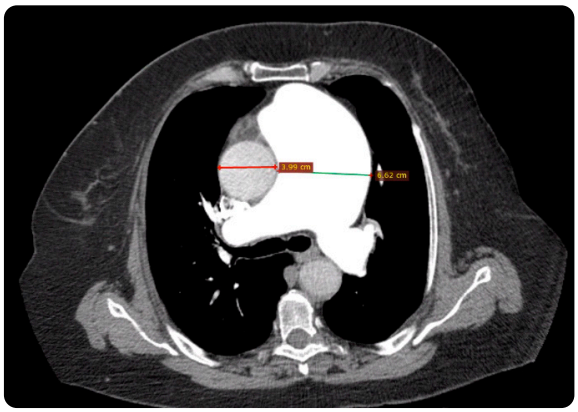

Figure 6. Chest angiotomography, axial, in contrasted phase in mediastinal window: there is evidence of fusiform dilatation of the PA trunk of 66 mm, the diameter of the PA trunk is more than 1.5 times the upper limit of normality.

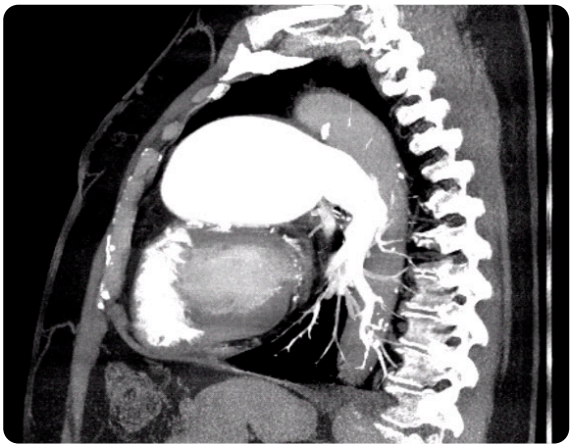

Figure 7. Sagittal maximum intensity projection (MIP) reconstruction of chest angiotomography with contrast medium: there is evidence of fusiform dilatation of the PA trunk without involvement of the main branches.

Likewise, a chest CT scan with contrast medium was performed showing dilatation of the pulmonary artery trunk (48 mm) (Figure 2), and coexistence of dilatation of the right branch of the PA (corresponding to the opacity visualized in the chest X-ray) reaching a maximum diameter of 52 mm, associated with eccentric opacification defects due to thrombosis of chronic characteristics and linear calcifications of the arterial walls (Figures 3 and 4).

Additionally, arterial blood gases reveal that the patient has a mild respiratory acidosis and a severe oxygenation disorder, with a PaO2/FiO2 ratio of 55.8. The management of the patient's underlying pathologies was optimized and she was subsequently discharged. Six days later, the family reported the patient's death at home.

Case 2

69-year-old female patient hospitalized by the general surgery service for clinical symptoms of intestinal obstruction by flanges, resolved with medical management, who has a history of hypertension, asthma and chronic lung disease under study due to exposure to wood smoke, under pharmacological management with salbutamol, beclomethasone, losartan and omeprazole.

During the hospital stay he presented sudden oppressive chest pain of intensity 7/10, associated with dyspnea and diaphoresis with peripheral oxygen saturation of 88%. Physical examination revealed stable vital signs, with no relevant findings. An electrocardiogram was performed showing signs of left bundle branch block; cardiac biomarkers were taken, which were negative, so acute coronary syndrome was ruled out. Chest X-ray showed cardiomegaly with no other abnormalities (Figure 5).

A transthoracic echocardiogram showed mild right ventricular dilatation with preserved systolic function and biauricular dilatation predominantly on the right, with probable mild pulmonary hypertension.

In addition, chest angiotomography was performed, showing fusiform dilatation of the PA trunk (66 mm) (Figures 6 and 7), with no evidence of endoluminal emboli in the trunk or its main branches; no other abnormalities were found. Normal paraclinical examinations ruled out other alterations.

Conclusions

PAA is a rare pathology, usually asymptomatic, which is usually diagnosed in necropsy studies and can be found incidentally in chest imaging studies in different modalities. Although the diagnostic criteria are still debated, pulmonary artery trunk diameter greater than 1.5 times the upper normal diameter (43.4 mm in men and 40.4 mm in women) is the most accepted parameter for CT and MR imaging.

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