SURGICAL CORRECTION OF PULMONARY ARTERY ANEURYSM. CASE REPORT

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ABSTRACT

Pulmonary artery aneurysm (PAA) is a rare condition that can occur in both sexes, 89% in the main pulmonary artery and 11% in the branches of the pulmonary artery. It presents a risk of compression of adjacent structures, such as the left coronary artery (LMCA), among others. Rupture of the PAA can result in high mortality rates, ranging between 50 and 100%. We report a case on the clinical evolution after the surgical approach of a patient with symptomatic PAA, with potential compression of the LMCA.

KEYWORDS: ANEURYSM; THORACIC SURGERY; PULMONARY ARTERY.

INTRODUCTION

Pulmonary artery aneurysm (PAA) is a rare condition, first described in 1860. Based on autopsies, a prevalence of approximately 1 in 13,696 individuals was estimated [1]. It generally occurs in both sexes, with 89% in the main pulmonary artery and 11% in the branches of the pulmonary artery. Among the branches, the most common incidence occurs in the left branch [2]. They can arise secondary to various etiologies such as infection, malignancies, congenital heart disease, or vasculitis [3].

PAA is characterized by focal dilatation of the pulmonary artery involving all three layers of the vessel wall [4]. The condition is diagnosed when the diameter of the vessel dilatation is greater than 1.5 times the upper limits of normal. In most studies, a gender-based cutoff was not followed, considering an aneurysm all cases with a pulmonary artery diameter greater than 40 mm [3]. PAA presents a risk of compression of adjacent structures, such as the left main coronary artery (LMCA), the main bronchus, and/or the recurrent laryngeal nerve. Among potentially fatal complications, rupture of the PAA can result in high mortality rates, ranging from 50 to 100% [4].

There is no specific guideline for the best approach when PAA is diagnosed. Among the main surgical indications are an aneurysm diameter greater than 55mm; an increase in diameter greater than 5mm in six months; compression of adjacent structures; thrombus formation in the aneurysm sac; onset of clinical symptoms; evidence of valve pathologies; pulmonary hypertension; and signs of aneurysm rupture or dissection [2].

The aim of this report is to demonstrate the clinical evolution following the surgical approach of a patient with symptomatic PAA, with potential adjacent compression of the left main coronary artery.

CASE REPORT

The patient is a 71-year-old male presenting for a cardiology evaluation for a dental implant procedure. During the consultation, he complains of typical chest pain during exertion, followed by syncope, even during routine bike rides, which started in the last few months. He reports experiencing near-syncope episodes while evacuating, but of different characteristics, with prodromes before the episodes. His medical history includes hypertension, dyslipidemia, prediabetes, and an ischemic stroke without sequelae in 1990. He brings an ultrasound (US) of the carotid arteries from 2018 showing plaques compromising 25% of the vessel lumen. He was previously diagnosed with pulmonary valve stenosis and underwent pulmonary valve commissurotomy in 1986. He is currently taking Diovan HCT®, Plenance Eze®, and Gliβ. On cardiovascular auscultation, there is a +++/6+ systolic and diastolic murmur in the pulmonary focus. The electrocardiogram shows sinus rhythm and right bundle branch block. An echocardiogram with stress, performed in 2020, revealed a left ventricular ejection fraction (LVEF) of 74% (Teichholz) with no segmental contractility abnormalities at rest or during stress.

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Due to cardiovascular risk, a new echocardiogram and carotid ultrasound were requested. The echocardiogram showed: LVEF of 55%. Posterior wall and septal wall thickness of 9 mm. Right Ventricle (RV) - Right Atrium (RA) gradient of 25 mmHg. Systolic pressure in the pulmonary artery of 30 mmHg. Significant ectasia in the pulmonary artery trunk and its left branch associated with moderate pulmonary valve regurgitation (average gradient of 9 mmHg and velocity of 2.0 m/s), as well as right chamber overload, with an internal diameter of the RV of 47 mm, S' wave of 13.2 cm/s. Due to the pulmonary artery ectasia, a CT angiography (CTA) of the pulmonary and coronary arteries was requested for further investigation, with the following findings: Pulmonary Artery Aneurysm (PAA) (internal diameter of 47 mm in the trunk region and diameter of the left branch of 24 mm), with its trunk showing intimate relationship with the TCE (1.1 mm distance) (Figure 1), without causing compression at rest. Moderate luminal reduction in the right coronary artery (RCA) and slight reduction in the second marginal artery (Mg2).

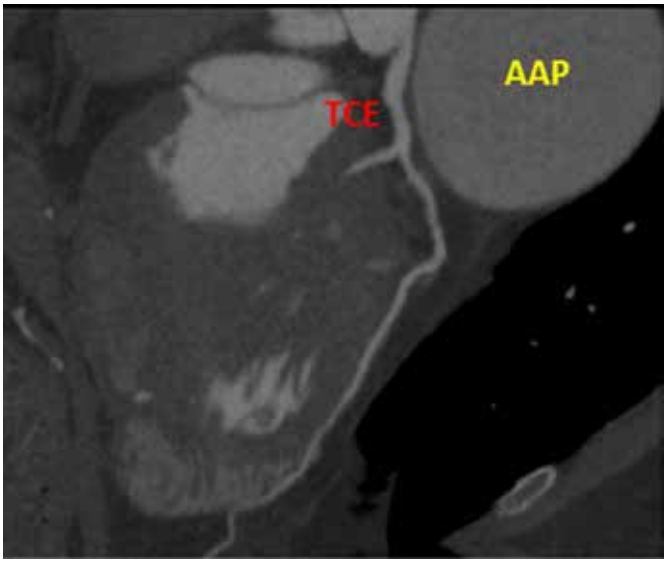

Figure 01. Angiotomography showing pulmonary artery aneurysm (PAA) in close proximity to the left coronary artery trunk (LCA).

Cardiac magnetic resonance imaging was performed for better anatomical evaluation, showing: Left Ventricular Ejection Fraction (LVEF) of 65% and Right Ventricular Ejection Fraction (RVEF) of 69%. Cardiac chambers with preserved dimensions. Preserved biventricular systolic function. Absence of myocardial fibrosis. Mild pulmonary valve regurgitation (regurgitant fraction of 18%). Presence of significant aneurysmal dilation of the pulmonary artery, measuring 55 mm in its largest diameter (Figure 2).

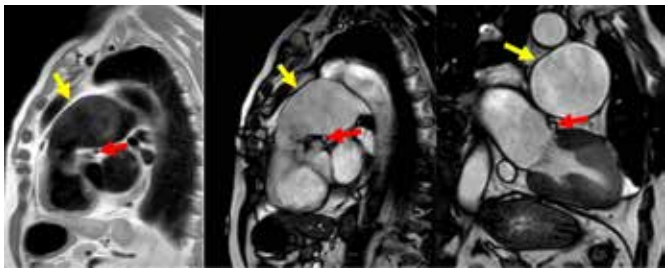

Figure 02. MRI showing pulmonary artery aneurysm (yellow arrow) revealing proximity to the left coronary artery (red arrow).

After discussion with the Heart Team, surgical correction of the aneurysm was indicated. The procedure was performed with a median sternotomy, the heart in sinus rhythm, and an 80mm diameter aneurysm identified. Heparinization followed by aortic and atrial cannulation with initiation of cardiopulmonary bypass (CPB) under hypothermia below 25º. Aortic clamping was performed, and cardioplegia was administered into the aortic root with Custodiol®. The tricuspid valve was calcified. The procedure continued with pulmonary arteriotomy and pulmonary valve commissurotomy, end-to-end anastomosis of a 30 Dacron tube with the left pulmonary artery, side-to-side anastomosis with a tube, and end-to-end anastomosis of the pulmonary origin with the tube. Reinforcement with biological glue and closure of the aneurysm over the tube. Warm-up and air removal maneuvers were performed. Aortic declamping occurred at 44 minutes with return to sinus rhythm after 10J defibrillation. CPB weaning was done at 55 minutes, mediastinal drainage with two Blake drains, two temporary pacemaker wires in the right ventricle, hemostasis revision with biological glue, and pericardium reconstruction with Goretex®. Layer closure and dressing. He had a good postoperative evolution without surgical complications during hospitalization. Twice, after 9 and 24 days post-procedure, he presented acute atrial fibrillation (AF) in the emergency room, returning to sinus rhythm after chemical reversion with amiodarone.

In the post-surgical follow-up appointment, the patient brought an echocardiogram (ECO) and an angioCT of the pulmonary arteries. The ECO showed a diagnosis of mild pulmonary valve stenosis, with a peak gradient of 35 mmHg and mild pericardial effusion. The angioCT revealed a prosthesis in the main pulmonary artery that starts from the valve and extends to the bifurcation of the right and left branches, which has a preserved caliber and is patent. The maximum caliber of the prosthesis measures 30 mm. Correction of the pulmonary artery aneurysm with a prosthesis was successful, without signs of complications.

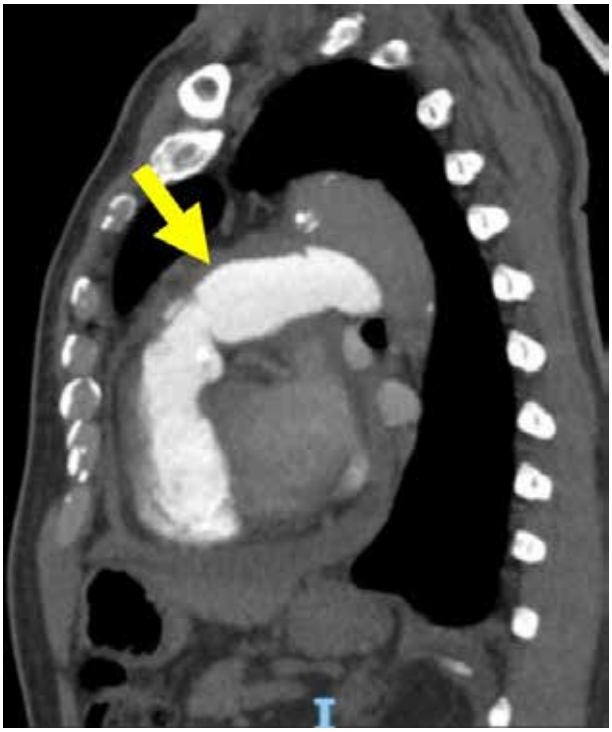

Figure 03. Post-procedure angiotomography showing Dacron tube (yellow arrow) demonstrating correction of the PAA.

The patient is currently using Eliquis, Diovan HCT, Amiodarone, and Dexilant. He is clinically stable, with no new episodes of atrial fibrillation, pre-syncope, or syncope, reporting an improved quality of life without complaints of angina-like chest pain.

DISCUSSION

The PAA is a rare entity among pulmonary vascular diseases, but with unfavorable and potentially fatal outcomes. Patients may go for long periods without a diagnosis, as a considerable portion of cases are asymptomatic, with diagnosis often occurring incidentally through imaging exams done for other reasons. Fatality can result from aneurysmal rupture with frank hemoptysis, pulmonary artery dissection, and compression of the coronary artery, leading to acute coronary syndrome.

It presents a varied spectrum of clinical manifestations depending on the size, pressure, growth rate, and contact with adjacent structures. In situations of low pressure and slow growth, patients can remain asymptomatic for years to decades [4].

Studies unanimously report the difficulty in finding the ideal management after diagnosis, primarily because there are no specific guidelines for PAA to date. Treatment should be tailored to each case, considering the underlying cause, symptoms, risk-benefit of surgical intervention, and comorbidities. In cases where conservative management is chosen, treatment should include managing underlying conditions, pulmonary hypertension if present, and regular imaging follow-up.

When there is dilation of the pulmonary artery (PA), extrinsic compression on the left main coronary artery (LMCA) can occur, as in the case reported. This becomes likely when the diameter exceeds 55 mm or when the ratio between this diameter and the diameter of the aortic root is at least 1.98. Its development depends on a chronic increase in the diameter of the PA, usually associated with pulmonary hypertension (PH). The most common clinical presentation is angina-like chest pain, associated or not with dyspnea. Syncope, arrhythmias, or myocardial infarction may also be present. It has been observed that abnormal flow through the pulmonary valve can cause tension in the vessel wall due to the eccentric jet outflow from the right ventricle, leading to weakness in the vessel wall. A review identified 19 cases due to pulmonary valve abnormalities, with no association with PH. The median age was 50 years, with cases reported up to 79 years, and a similar gender distribution [3].

After an extensive review inferring the rarity of the diagnosis, we also identified numerous cases discovered in individuals younger than the case in question. The low pressure in the PA explains the long asymptomatic period, with symptoms starting from a significant increase in arterial diameter of 55 mm, which, according to the review, increases the risk of compression of adjacent structures

Some factors increase the risk of PAA rupture, such as chronic PAH, PA pressure greater than 50 mmHg, PA diameter greater than 75 mm, and annual growth exceeding 2 mm. There are several options to reduce the risk of these complications, including addressing the underlying cause, continuous monitoring through imaging studies as already mentioned, and even the need for surgical intervention.

Aneurysmorrhaphy and aneurysmectomy surgeries are well described for the treatment of aneurysms confined to the pulmonary trunk. Early surgery should be considered in patients with reasonable surgical risk to prevent worsening of cardiac function due to diameter progression and difficulty in ventilation due to chronic bronchial obstruction and atelectasis. Procedures in younger patients reduce postoperative morbidity and mortality [2].

The patient in question had more than one formal indication for surgical intervention, including a diameter of 55 mm, presence of symptoms (chest pain/syncope), and associated pulmonary valve pathology. Due to the clinical benefits of reducing symptoms, preventing progression, and avoiding complications related to the PAA, early surgical treatment, with its proper indications, showed good results and improved the patient's quality of life.

This study, along with the scarce number of published cases, may contribute to advising medical teams on possible interventions for PAA, while awaiting guidelines to guide the ideal management.

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