Incidental Finding of Giant Aneurysm of Trunk of the Pulmonary Artery

CASE REPORT

Thy-ciara Fontenele Marques¹, Fernanda Moura Victor¹, José Célio Couto Vasconcelos¹, Ricardo Parente Garcia Vieira¹, Susyane Ribeiro Beserra¹, Hermes Melo Teixeira Batista^{1,2}, Samuel Soares Eduardo¹, Antonia Rayanne Xavier Barbosa¹, Italla Maria Pinheiro Bezerra², Luiz Carlos de Abreu²

¹ Hospital regional do Cariri
² Laboratório de Delineamento de Estudos e Escrita Científica. Departamento de Saúde da Coletividade. Disciplina de Metodologia Científica.

Contact information:

Hermes Melo Teixeira Batista

✉ hermesmelo@oi.com.br

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Abstract

Introduction: Pulmonary artery aneurysm (PAA) is a rare condition and associated with multiple pathologies. In most cases there are reports of dyspnea and palpitation, however, some patients are completely asymptomatic. Its natural history is uncertain and there are no clear guidelines about its treatment and monitoring.

Method: This is one case report, obtained through data from medical records of a reference hospital, located in the city of Juazeiro do Norte, Ceará, Brazil.

Case Report: Patient, female, 64 years old, healthy, sought medical assistance after a clinical picture of drug allergy. The physical examination performed at admission revealed hyperdynamic precordium associated with systolic thrills and murmur. Her chest radiography (X-ray) revealed a mediastinal widening, which after CT evaluation, it was found that it was a large aneurysm of trunk of the pulmonary artery. As it remained asymptomatic throughout the investigation and she refused to undergo surgical treatment, there was the adoption of a conservative conduct only with an outpatient treatment.

Introduction

Pulmonary artery aneurysm (PAA), a pathological dilatation of the pulmonary artery and/or one of its main branches, is a rare condition whose prevalence is still unknown. Studies have reported one case for every 14,000 autopsies [1]. Although there is no precise definition for the PAA as occurs for the

aortic aneurysm, some authors have indicated four centimeters in diameter as a cut-off point, although more recent studies involving computed tomography have reported a normal upper limit of the pulmonary artery diameter of 29 mm [2-4].

Several etiologies have been described in its pathogenesis from congenital heart defects, infections, vasculitis, connective tissue disease, tuberous

sclerosis, traumas, arteriovenous fistulas, and even atherosclerosis disease. After introduction of the use of antibiotics, the PAA of noninfectious cause became the most common etiology [5-7]. However, when no pathological condition is identified and in the absence of structural or functional abnormalities of the cardiovascular system, the PAA is named as idiopathic [8].

It is clinically manifested by nonspecific symptoms such as chest pain, dyspnea, cough, hemoptysis or palpitations [9]. Some patients, however, are totally asymptomatic and the PAA is casually detected in imaging studies realized because of other reasons [10]. Depending on their size, they can lead to significant complications, including, in rare cases, airway compression. The rupture and/or dissection of the aneurysms in low-pressure chambers are of rare occurrence [11].

There are no clear guidelines about the treatment of the PAA and the patients can be treated individually. The treatment can be clinical, surgical or endovascular. Clinically, the control of the pulmonary pressure, predisposing factors and underlying disease is imperative [12]. Surgical treatment is usually indicated in the most unstable aneurysms with hemoptysis and in which a fatal outcome because of increased risk of rupture is foreseeable [13]. The endovascular therapy is reserved for when the clinical treatment alone fails to control the evolution of the disease, especially when the surgical repair entails high rates of morbidity and mortality [13, 14].

Case Report

Patient, female, 64 years old, without previous comorbidities, was admitted in a service of reference with diffuse pruritic rash and facial edema after the use of anti-emetic medication. She denied chest pain, cough, hemoptysis, palpitations, dyspnea or cyanosis and prior history of drug allergy. She was not using any other drugs, alcohol or cigarettes.

Figure 1: Chest radiography (X-ray) showing mediastinal widening with prominence of the middle arc corresponding to trunk of the pulmonary artery and the outer edge of the right atrium.

On examination, she was normotensive, with symmetric peripheral pulses and normal capillary refill. A hyperdynamic precordium with thrill to palpation was evidenced, severe systolic murmur best heard in pulmonary and tricuspid focus, and heart rate of 64 beats per minute. Her laboratory evaluation revealed no abnormalities with normal inflammatory tests, negative serology for schistosomiasis, nonreactive to ANF, HIV and VDRL. The chest radiography (**Figure 1**) revealed a mediastinal widening; the electrocardiogram suggested overload of the right chambers and the chest tomography (**figures 2A and 2B**) showed an ectasia of the trunk of the pulmonary artery, measuring 9.0 cm in its largest diameter and dilatation of the main pulmonary arteries and cardiomegaly. The three-dimensional reconstruction (**Figure 3**) allowed a better visualization of aneurysmal dimensions and its relationship to adjacent structures. In addition, the transthoracic echocardiography showed moderate tricuspid insufficiency associated with pulmonary artery hyperten-

Figure 2A: Axial chest CT with window to mediastinum showing marked dilatation of the pulmonary trunk (9.0 cm) and the main branches.

Figure 2B: Chest CT with coronal plane reconstruction of the aneurysmal dilatation.

Figure 3: 3D reconstruction showing massive expansion and its relationship to the aortic arch.

sion (70 mmHg) and maintained systolic function. The patient evolved well and because of the lack of symptoms attributed to the aneurysm, the shortage in the literature of recommendations about surgical intervention and the desire of non-intervention by the patient, there was the decision for performance of an expectant approach with outpatient monitoring of the use of nifedipine retard 40 mg/day and periodic computed tomography control.

Discussion

Pulmonary artery aneurysm is an uncommon clinical entity, diagnosed mainly in autopsy study. Although the majority of patients relate some symptoms such as dyspnea, cough, hemoptysis or palpitation, a part of them evolves asymptomatic. It is noteworthy, therefore, the importance of a thorough physical examination. Currently, because of easier access to imaging methods, some patients with PAA are incidentally diagnosed in radiologic

studies, even in the cases of giant aneurysms like the patient described.

According to Nguyen et al., the normal value for the upper limit of the diameter of the main pulmonary artery is of 29 mm in CT image [4]. The CT scan of our patient showed an aneurysmal dilatation 3 times greater than this limit, with few cases report-

ed in the literature with similar diameters.

There are still many divergences regarding the treatment of the PAA and there is no specific recommendation. The surgical treatment is usually indicated in the most unstable aneurysms with hemoptysis and in cases in which a fatal outcome because of the increased risk of rupture is foreseeable, such as mycotic aneurysm, the ones associated with Behcet's disease, Hughes-Stovin syndrome, Marfan syndrome and Osler-Weber-Rendu syndrome [1, 12]. Several surgical techniques have been reported as replacement for the Dacron graft, the substitution with the combination of Dacron prosthesis and bioprosthesis, aneurysmorrhaphy and repair of lung allograft [15].

The greatest concern is over the long term evolution of the cases not treated surgically, because there is little data in the literature. However, there are reports of clinical monitoring that reached 40 years, without complications.

Because of the lack of symptoms attributed to the aneurysm, the shortage of recommendations about surgical intervention in the literature, evidence that the periodic monitoring is safe, absence of an underlying disease and the desire of non-intervention by our patient, there was the decision of performance of an expectant approach with outpatient monitoring and periodic CT control.

Conclusion

The rarity of the PAA confirms the scarcity of data in the literature and reinforces the importance of the description of case reports, even isolated. As some patients are asymptomatic, a simple physical examination associated with thoracic imaging can identify cases that would be missed in a normal routine. Despite the availability of several corrective

surgical techniques there is growing evidence that outpatient treatment is safe. More data are needed in the literature to reinforce the indications of treatment and monitoring of these patients.

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