Idiopathic Aneurysm of Pulmonary Artery

Júlio B. Cota Pacheco, Patrícia N. Pimentel, Beatriz S. Knust
Clinica de Cardiologia Cota Pacheco, Mogi das Cruzes, SP - Brazil

Introduction

Because it is a very rare isolated lesion, we decided to present this case of idiopathic pulmonary artery aneurysm (IPAA) and review the cases published in the literature in order to correlate our clinical and imaging findings, as well as management based on patient data. IPAA was first described in a case of autopsy by Bristowe in 1860 and later in 1947 by Deterling and Claggett, whose prevalence was lower than eight to one hundred thousand[1,2].

Although the use of diagnostic imaging methods has been applied in a very large population in recent decades, this lesion has been most often described in postmortem examinations[3]. Therefore, it is important to be aware of possible clinical symptoms, at times non-specific, as well as the signs on imaging studies. In this study, therefore, the report of a case of an asymptomatic patient whose diagnosis was done through color Doppler echocardiography in a routine test in 2012, later confirmed by simple chest computed tomography (chest CT) and cardiac catheterization as IPAA and its branches. We discussed the literature available and the possibilities for treatment and the use of color Doppler echocardiography as an initial diagnostic tool for such a rare and intriguing disease.

Case Report

Asymptomatic female patient, white, 82, who, two years before, came to this service for treating arterial hypertension (AH) and diabetes mellitus type II (DM II). Since then, the patient had been taking olmesartan 20 mg, amlodipine besylate 5 mg, metformin 850 mg and simvastatin 20 mg. Laboratory tests showed adequate control of DM II, HA and dyslipidemia, with preserved renal function, liver enzymes and blood count without abnormalities, serological reaction to negative syphilis. On physical test, the patient presented, on cardiac auscultation, normophonetic rhythmic sounds (NFRS) with systolic murmur in mesocardium ++. Electrocardiogram (ECG) — sinus rhythm, no abnormalities. Color Doppler echocardiography revealed mild left ventricular hypertrophy (LVH), good left ventricular (LV) and right ventricular (RV) contractile function, abnormal LV relaxation, discrete pulmonary valve reflux without pathological systolic gradient, good valve opening and aneurysmal dilatation of the pulmonary artery and its branches, pulmonary trunk (PT) = 5.0 cm, right branch = 2.0 cm and left bundle branch = 2.5 cm (Figure 1). Spectral color Doppler revealed a discrete PT flow turbulence TP (Figures 2 and 3). No signs of pulmonary hypertension were observed on Doppler (PH).

A chest CT scan showed aneurysmal dilatation of the pulmonary artery and branches with a PT diameter = 4.9 cm (Figure 4). Cardiac catheterization showed coronary arteries free of stenosis and aneurysmal dilatation of the pulmonary artery and its branches (Figure 5) with no signs of PH. Pulmonary artery systolic pressure (PASP) = 30 mmHg and medium pressure (Mp) = 20 mmHg.

Due to the stable condition of the patient age, only clinical follow-up and treatment of comorbidities were proposed.

After two years of clinical follow-up, laboratory and imaging tests, the patient is stable with no abnormalities in the tests described above.

Discussion

When diagnosed by imaging methods, patients with IPAA may be asymptomatic, but the most common symptoms are chest pain, dyspnea, cough, hemoptysis and palpitations[4,5]. Diagnosis is established with invasive diagnostic procedures and can be started with color Doppler echocardiography because it is easy to apply and to obtain information about anatomy and function. It is necessary to complement the study with other tests such as chest CT, MRI and cardiac catheterization for a more accurate analysis of this disease.

IPAA is rare and its pathogenesis and incidence are little known, but it is estimated that it is lower than eight for each hundred thousand in autopsy findings[3]. To diagnose an IPAA, the initial criterion that should be used is a PT dilation greater than 3.0 cm with no cardiac or pulmonary cause. It is known that when such lesion occurs, the three layers of the artery are compromised and this may extend to its branches[6]. So when a PT dilation above 3.0 cm is found, an etiological investigation of the possible causes should be conducted: congenital diseases with increased pulmonary blood flow, pulmonary hypertension, pulmonary valve stenosis with post-stenosis dilation, systemic arteritis, including the Behcet's disease and collagen diseases (Marfan's syndrome, Ehler's - Danlos syndrome), trauma and infections (syphilis, bacterial endocarditis and tuberculosis)[3]. The latter also includes an interesting case report of a patient with schistosomiasis, which evolved with pulmonary artery aneurysm and pulmonary hypertension, and had a fatal complication: artery rupture[7]. When the patient does not fit in any of these cases, we call it IPAA. It is known that the risk of

Keywords

Pulmonary Artery/injuries; Echocardiography, Dopper; Tomography, X-Ray Computed; Cardiac Catheterization.

Mailing Address: Júlio Batista Cota Pacheco •
Clínica de Cardiologia Cota Pacheco
Av. Braz de Pina, 455, Postal Code 08730-020, Vila Vitória,
Mogi das Cruzes, SP - Brazil
E-mail: jcota@uol.com.br
Manuscript received on February 12, 2015; revised on March 9, 2015; accepted on April 22, 2015

DOI: 10.5935/2318-8219.20150023

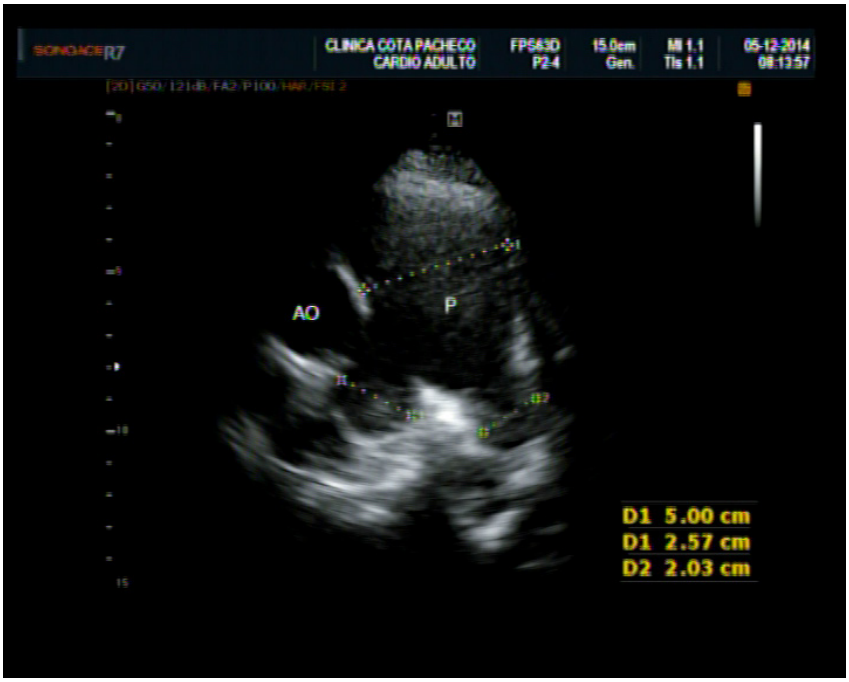

Figure 1 - *Color Doppler echocardiography (base cross section) showing severe dilatation of the pulmonary trunk and proximal segments of its branches. Ao (Aorta); P (pulmonary); Pulmonary artery trunk diameter (D1 5.0 cm); Diameter of the left pulmonary artery (D1 2.57 cm); Pulmonary artery right branch diameter pulmonary (D2 2.0 cm).*

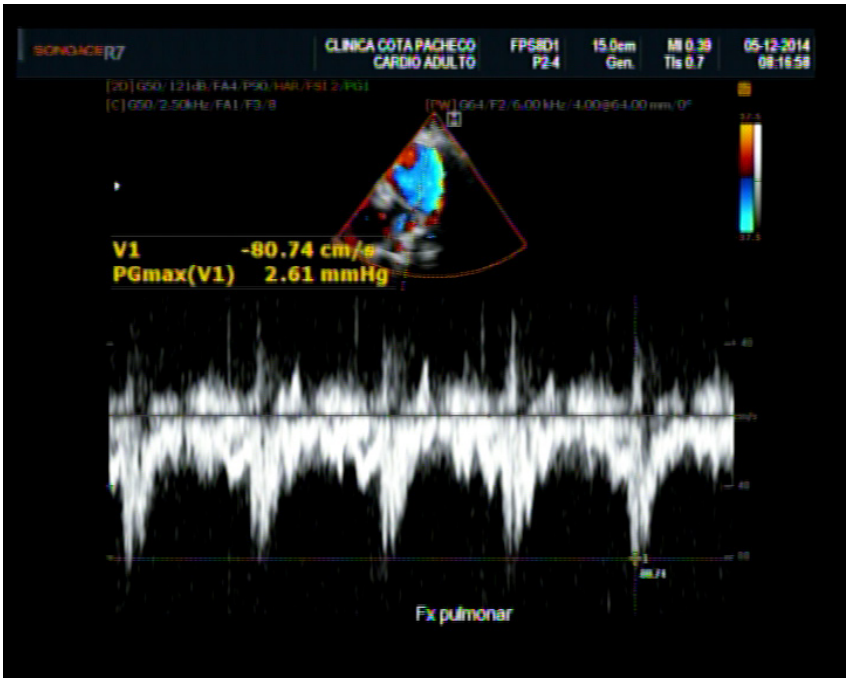

Figure 2 - *Spectral Doppler in pulmonary valve showing the absence of pathological systolic gradient and discrete turbulence. Pulmonary flow (pulmonary Fl).*

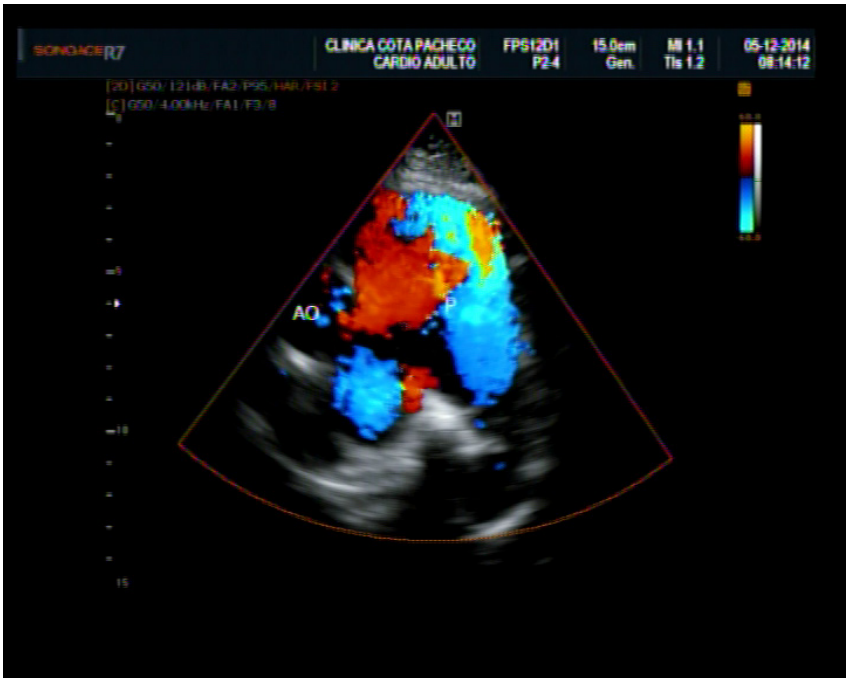

Figure 3 - *Echocardiography with color flow in the pulmonary artery. AO (Aorta); P (Pulmonary).*

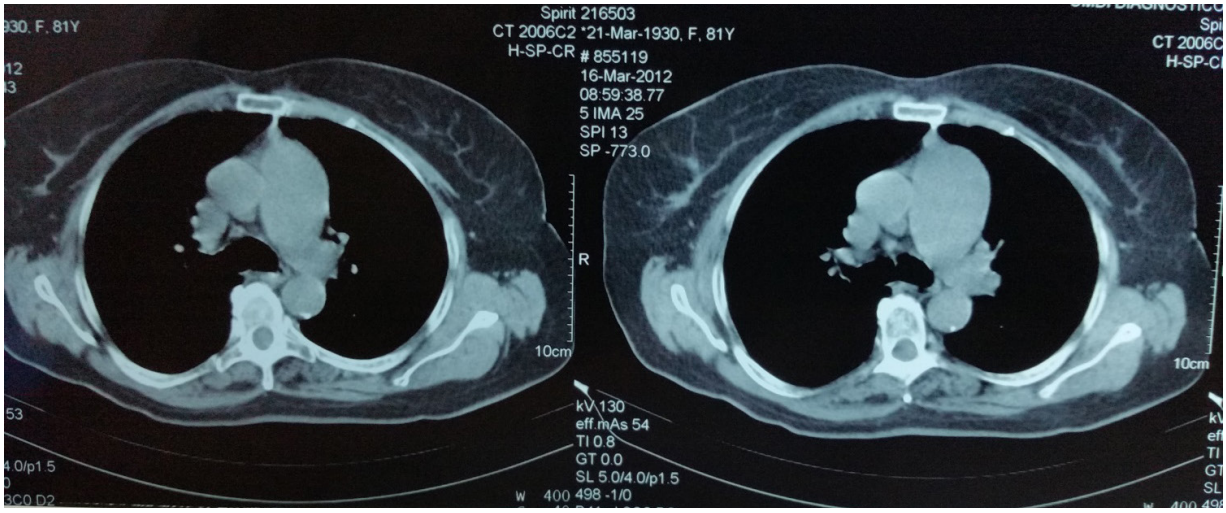

Figure 4 - *Simple chest Computed Tomography showing significant dilation of the pulmonary trunk.*

IPAA complications is smaller than aortic aneurysm, particularly if pulmonary artery pressures are normal[3].

Treatment for IPAA remains controversial, and for asymptomatic and stable patients, clinical follow-up is recommended, with description in the literature with clinical improvement for more than three decades of follow-up[4]. For symptomatic and unstable patients, surgery or endovascular intervention have been performed. So far, the maximum diameter of the pulmonary artery that would determine surgical indication has not been established, just like with aortic aneurysms; however, surgical intervention is recommended when its diameter is equal to or greater than 60 mm[5]. It is recommended that surgery be prescribed for low-risk patients with symptoms and/or signs of constant progressive increase of the aneurysm[8].

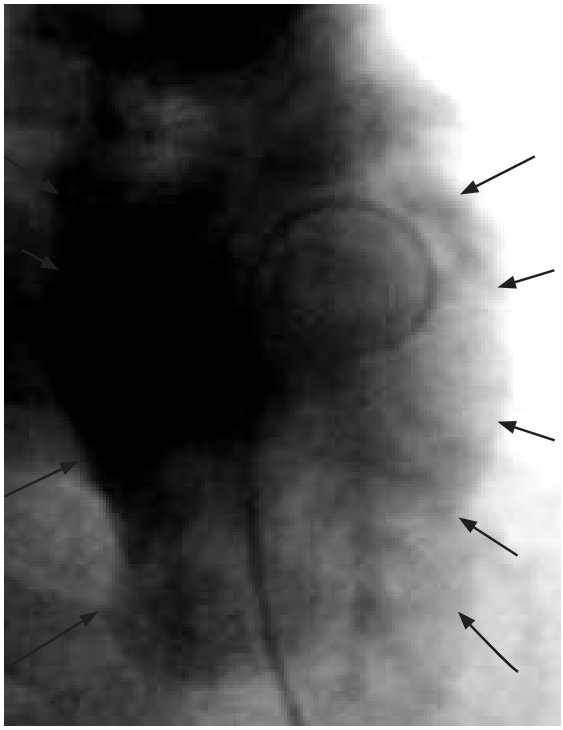

Figure 5 - *Cardiac catheterization showing aneurysmal dilation of the pulmonary artery.*

The surgical alternatives proposed are: 1 correction with Dacron prosthesis or reconstruction with pericardial patch; 2 aneurysmorrhaphy; or 3 arterioplasty[8,9].

Endovascular treatment may be an option when there is progression of the aneurysm, onset of symptoms or impairment of the right chambers due to pulmonary reflux and when there is high risk for open surgery[10].

The most dramatic and terrible complication is rupture. However, even when there is indication for surgery because of symptoms, medical treatment is recommended if there is high surgical risk. Clinical and radiological control should be performed every six months or at any time if any acute changes are found in the clinical status of the patient[11,12].

In our case, we opted for the clinical and radiological follow-up due to the high age of the patient and because the patient was clinically stable and with no abnormalities in subsequent radiological controls (color Doppler echocardiography and chest CT), since diagnosis two years before.

Conclusion

Although IPAA is rare, a first suspicion may be raised by color Doppler echocardiography, a key test for diagnostic suspicion and control, which should be taken into account due to its accuracy and ease of execution, as well as an objective assessment of ventricular function. The support of other imaging methods and laboratory tests allows a complete and reliable assessment for choosing the best treatment for each patient. Due to the low incidence of the disease, surgical treatment protocols have not yet been established, strengthening the clinical and radiological data for making therapeutic decisions.

Authors' contributions

Research creation and design: Pacheco JBC; Data collection: Pacheco JBC; Data analysis and interpretation: Pacheco JBC; Manuscript drafting: Pacheco JBC, Pimentel PN, Knust BS; Critical revision of the manuscript for important intellectual content: Pimentel PN, Knust BS.

Potential Conflicts of Interest

No relevant potential conflicts of interest.

Sources of Funding

This study had no external funding sources.

Academic Association

This study is not associated with any graduate program.

Referências

1. Bartter T, Irwin RS, Nash G. Aneurysms of the pulmonary arteries. Chest. 1988;94(5):1065–75.
2. Deterling RA Jr, Clagett OT. Aneurysm of the pulmonary artery; review of the literature and report of a case. Am Heart J. 1947;34(4):471–99.
3. Nair KK, Cobanoglu AM. Idiopathic main pulmonary artery aneurysm. Ann Thorac Surg. 2001;71(5):1688–90.
4. Van Rens MT, Westermann CJ, Postmus PE, Schranmel FM. Untreated idiopathic aneurysm of the pulmonary artery: long-term follow-up. Respir Med. 2000;94(4):404-5.
5. Deb SJ, Zehr KJ, Shields RC. Idiopathic pulmonary artery aneusrysm. Ann Thorac Surg. 2005;80(4):1500-2.
6. Singh A, Singh K, Aditi P, Aneja P. Pulmonary Artery Aneusrysm. Indian J Chest Allied Sci. 2014;56(1):45-57.
7. Piveta RB, Arruda AL, Rodrigues AC, Pinheiro JÁ, Andrade JL. Rupture of a Giant aneurysm of the pulmonary artery caused by schistosomiasis. Eur Heart J. 2012; 33(9):1159.
8. Henn LW, Esmailian F. Repair of a large main pulmonary artery aneurysm in a 71-year-old Jehovah´s witness patient. Tex Heart Inst J. 2013;40(3):350-2.
9. Arom KV, Richardson JD, Grover FL, Feris G, Trinkle JK. Pulmonary artery aneurysm. Ann Surg. 1978;44(10):688-92.
10. Gomes O, Morais J, Borges C, Moreira A, Leite J, Fernandes C. Giant aneurysm of the main pulmonary artery: Case Report. Rev Port Cardiol. 2008;27 (11):1463-8.
11. Vural AH, Turk T, Ata Y, Goncu T, Ozyazicioglu A. Idiopathic asymptomatic main pulmonary artery aneurysm: surgery or conservative management? A case report. Heart Surg Forum. 2007;10(4):E273–5.
12. Shafiq A, Bokhari A, Nahin I, Rabbani B. Extreme dilatation of pulmonary artery: a literature review. BMJ Case Reports.2014; doi: 10.1136/bcr-2013-202223.