Giant Left Atrial Thrombus with Double Coronary Vascularization

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Introduction

Rheumatic valvular disease, especially mitral stenosis (MS) and atrial fibrillation (AF), are the main factors related to the formation of left atrial (LA) thrombi. Its incidence can range from 16 to 64%, and the most affected site is the left atrial appendage. Systemic embolism is responsible for 10 to 45% of the complications, being the most frequent clinical presentation. Despite being rare, the mechanical mitral valve obstruction caused by thrombus may be considered as potentially severe, especially among those patients with previous MS. The clinical manifestations of this condition are variable, presenting from worsened functional class (NYHA) to cardiogenic shock.

The left atrial thrombus related to MS, even if rarely, may present as a large and organized mass with undistinguishable characteristics from vascularized tumors, especially the atrial myxoma. Clinical and echocardiographic aspects may not be sufficiently specific to distinguish one from the other safely, and additional examinations are often required.

This is the case of a patient with moderate rheumatic MS and no anticoagulation for AF, with a large thrombus organized in LA mimicking an atrial tumor, with difficult clinical differentiation by complementary examinations.

Case Report

It is the case of a 57-year-old female patient, with dyspnea that progressed to orthopnea, paroxysmal nocturnal dyspnea and lower extremity edema for two months. She had been diagnosed with rheumatic valvulopathy for seven years, permanent AF with no anticoagulation; she was a smoker. In the physical examination, she presented with good general status, 44 bpm heart rate, 150 x 80 mmHg blood pressure, cardiac auscultation with hyperphonetic sound, second to second sound, rumbling 2+/6+ diastolic murmur in the mitral area and pulmonary auscultation with fine crepitation in both bases. The electrocardiogram presented AF rhythm. The thoracic x-ray showed bilateral pulmonary congestion and increased cardiothoracic index. The transthoracic echocardiogram showed mitral valve with commissural fusion, thickened cusps and reduced valve opening, mean LA-LV diastolic gradient of 4 mmHg and maximum of 16 mmHg, with valve area of 1.2 cm², which is compatible with moderate rheumatic compromise.

A 51 mm LA and the presence of hyperechoic image from the left atrial roof to its lateral wall were described, measuring 65 x 54 mm (Figure 1A and B). There was also severe pulmonary arterial hypertension (SPAP 66 mmHg) and important increase in the left chambers, eccentric hypertrophy and left ventricular systolic dysfunction (35% ejection fraction), with diffuse hypokinesis.

The patient was submitted to cardiac catheterization, which excluded obstructive coronary lesions and showed the presence of extensive vascularization of the atrial mass, with irrigation originated from the left (Figure 1C) and right (Figure 1D) coronaries. Cardiac nuclear magnetic resonance showed important LA increase, with thickened walls and image compatible with large thrombus adhered to its walls (roof, floor and lateral wall), besides positive parietal diffuse enhancement, compatible with hypertrophy and atrial fibrosis, probably associated with MS. However, in the interface between the thrombus and the lateral LA wall, there was a positive perfusion, so it was not possible to prevent other expansive processes in the atrial wall. The late left ventricular enhancement was negative.

By considering the presence of an image in the LA in a patient with moderate MS and no anticoagulation for AF, there was the hypothesis of a giant thrombus. Due to the progression of the symptoms, size of thrombus and risk of embolism or mechanical obstruction of the mitral valve, the surgical treatment was chosen.

The patient was submitted to surgery, therefore, the mass that presented red coloration, with soft and friable consistency was removed, which confirmed the diagnosis of left atrial giant thrombus and impairment of the entire LA posterior wall, weighing 80 g (Figure 2A and B). Anterior and posterior commissurotomy was conducted in the mitral valve. The postoperative transthoracic echocardiogram showed mitral valve with mean LA-LV diastolic gradient of 4 mmHg and maximum diastolic gradient of 12 mmHg, and also a 1.6 cm² valvular area. There was still systolic dysfunction (EF 42%) resulting from diffuse hypokinesis. The patient evolved well in the postoperative period and received asymptomatic hospital discharge, on warfarin anticoagulant.
Discussion

MS and its marked atrial increase predisposes to AF in 40 to 75% of the symptomatic patients. Its occurrence increases with age and with the level of valvular obstruction. In this condition, blood stasis on the left atrial appendage and LA favors the formation of multiple thrombi in those areas, therefore, it is very important to identify them due to the risk of systemic embolism and, less frequently, mechanical valve obstruction.

In this case, atrial mass was related to MS with AF with no anticoagulation and to increased LA, being diagnosed as thrombus by the echocardiogram. However, the vascularized mass in the preoperative catheterization created doubts as to the nature of the mass. The discovery of a large mass in the LA in the echocardiogram forces the doctors to distinguish the cardiac myxoma and the thrombus, which are the most common aspects in cases of round masses in this chamber.
The myxoma is located in the LA in 90% of the cases, and there are also numberless reports of abnormal coronary vascularization in these tumors, thus being the main differential diagnosis. The precise diagnosis is important due to the different therapeutic proposals. The myxoma requires surgical resection, while the thrombus can be solved with anticoagulation. Nuclear magnetic resonance plays an important role in this differentiation; however, the organized thrombus may acquire the same characteristics of the image of a myxoma. Therefore, the presence of vascularization is not pathognomonic for atrial myxoma and cannot be used alone to tell a thrombus from a myxoma.

Giant thrombi that develop inside the atrial chamber are usually immovable, well organized and fibrotic, with a close relation to the wall. They have an unfavorable response to thrombolytic therapy, which is not safe due to the high risks of systemic embolism. The rare cases described in literature suggest that surgical removal should be the treatment of choice.

The mechanical obstruction of the mitral valve is rare, however, potentially severe, especially for people with previous MS. In this case, the patient had presented symptomatic moderate MS for two months (NYHA III) and pulmonary arterial hypertension (SPAP 66 mmHg), which were sufficient to indicate surgical treatment. Besides this indication, there was the presence of a large mass in the LA, which could not only have contributed with the symptoms presented by the patient, but also added the risk of systemic embolism. With regard to left ventricular dysfunction, the other etiologies were ruled out, therefore, such a dysfunction may be justified by preload reduction and by changes in ventricular geometry caused by the calcification and immobility of the mitral valve ring.

In literature, angiographic findings of neovascularization or coronary artery fistula formation for the LA have occasionally been described in association with atrial thrombus among patients with MS. Most of the time, the irrigation of these organized thrombi originates from the circumflex artery, so, the double irrigation by the left and right coronaries is extremely rare, as observed in this case.

MS and AF are the main factors related to the formation of thrombi in the LA, and sometimes they cannot be distinguished from vascularized tumors. Even though the patient had sufficient thrombogenic substrate, subsidiary tests brought up this diagnostic doubt, especially due to the preoperative angiographic finding. Surgery was the best strategy both in the diagnostic and in the therapeutic approach. This case leads to the diagnosis of giant left atrial thrombus with double coronary vascularization, thus configuring the second case in the world and the first one in Brazil.

Author contributions
Conception and design of the research, Acquisition of data, Analysis and interpretation of the data, Writing of the manuscript and Critical revision of the manuscript for intellectual content: Ciambelli GS, Baptista ML, Rosa VEE, Lopes ASSA, Accorsi RAD, Tarasoutchi F.

Potential Conflict of Interest
No potential conflict of interest relevant to this article was reported.

Sources of Funding
There were no external funding sources for this study.

Study Association
This study is not associated with any thesis or dissertation work.

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