

Idiopathic Mitral and Aortic Subvalvular Aneurysm in a Young Patient

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Abstract

Left ventricular aneurysms are primarily caused by coronary artery disease. Among them, there are also less common types, such as those located in the subvalvular regions of idiopathic origin. This article describes the case of a patient with a history of aortic subvalvular and Sinus of Valsalva aneurysm repairs who developed a new mitral subvalvular aneurysm.

Introduction

Left ventricular aneurysms primarily result from coronary artery disease, often as a complication following acute myocardial infarction. Other non-ischemic cardiac causes include arrhythmogenic heart disease, hypertrophic cardiomyopathy, and myocarditis. Notable systemic causes include Chagas disease, tuberculosis, syphilis, and HIV, among others.¹

A rarer entity is the idiopathic mitral subvalvular aneurysm, first described in Africa in the 1960s.²

Case Report

Male Afro-descendant patient, 17 years old, with a history of treatment for aneurysm in the left ventricular outflow tract and ruptured aneurysm of the Sinus of Valsalva, both repaired in 2018 with two percutaneous devices. Since the procedure, the patient had moderate aortic reflux, secondary to the anatomical distortion caused by the Sinus of Valsalva aneurysm and was maintained under clinical treatment.

Four years after the procedure, he began experiencing progressive dyspnea and declined overall condition, and sought emergency care to investigate the symptoms. Anamnesis gathered no fever, chest pain, syncope or palpitations.

The patient performed a transthoracic and transesophageal echocardiogram, which demonstrated enlarged left cavities, mild systolic dysfunction of the left ventricle and severe aortic reflux, progressively worsened compared to previous exams.

Additionally, a mitral subvalvular aneurysm was identified, located on the lateral wall of the left ventricle, distorting the mitral annulus and posterior leaflet anatomy. This resulted in a noticeable regurgitant jet emanating from the aneurysm cavity and traversing the posterior leaflet, which appeared perforated, likely due to trauma from the jet (Figure 1). This particular mitral subvalvular aneurysm had not been reported in any previous examinations, making it a new finding in the current investigation.

Considering ischemic disease as the primary cause of aneurysms related to the left ventricle, the patient underwent cardiac tomography angiography with analysis of the coronary arteries and aorta, revealing no lesions and preserved anatomy and patency. Additionally, an aneurysm in the supra-ventricular aortic region, partially occluded with a percutaneous prosthesis and containing contrast and parietal thrombus, was identified (Figure 2). Moreover, another device was observed in the left ventricular outflow tract, below the aortic valve plane, where a previous aneurysm was repaired, hindering visualization of the cavity or flow, likely due to thrombosis.

Blood samples were collected to rule out systemic causes. The exams evidenced slightly increased inflammatory markers, negative blood cultures, unaltered autoimmune markers and negative serologies, as described below (Table 1).

After initial clinical compensation, the patient underwent surgery on August 01, 2022, with resection of the mitral subvalvular aneurysm, patch repair and placement of a mitral mechanical prosthesis #29. Furthermore, the Sinus of Valsalva aneurysm was excluded with a bovine pericardium patch, and an aortic mechanical prosthesis #23 was placed. The mitral valve fragment was sent for culture and deemed negative for endocarditis. Post-operative control exams showed normal functioning prostheses, a well-positioned patch and moderate systolic dysfunction of the left ventricle.

The extensive etiological investigation with negative results resulted in a diagnosis of idiopathic mitral subvalvular aneurysm.

Discussion

Idiopathic subvalvular aneurysm of the left ventricle is a rare condition with uncertain etiologies. Its estimated incidence is around 34 cases for every 10,000 cardiovascular diseases.³ It appears to have an epidemiological correlation with people of African descent, as it was described for the first time in Africa, in the Bantu population.

The first descriptions come from two series of cases, published in 1962 by Abrahams et al.² and in 1965 by Chesler et al.,⁴ with the mitral subvalvular as the main affected region. In the case series published by Chesler et al.,⁴ all patients

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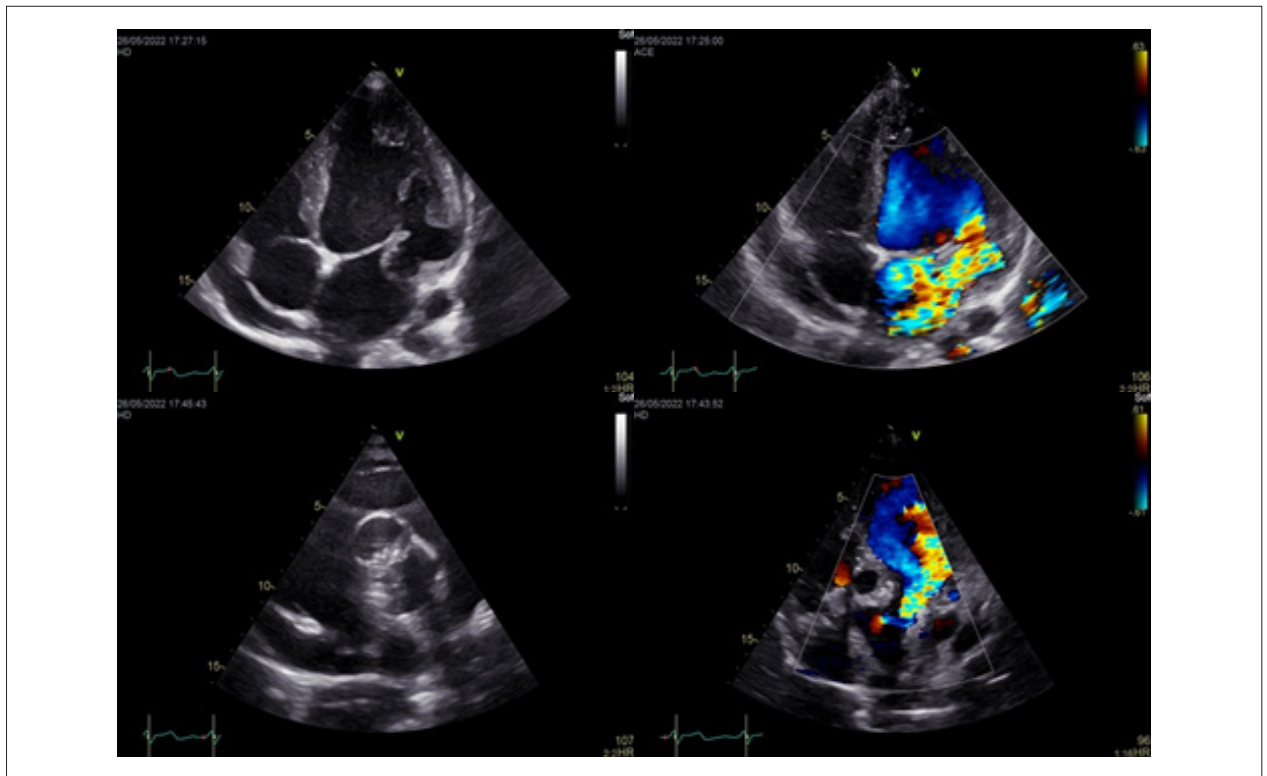


Figure 1 – Within the top-row images, the mitral subvalvular aneurysm is evident, located near the lateral wall of the left ventricle, with and without color Doppler. The lower-row images exhibit a device adjacent to the Sinuses of Valsalva and severe aortic reflux.

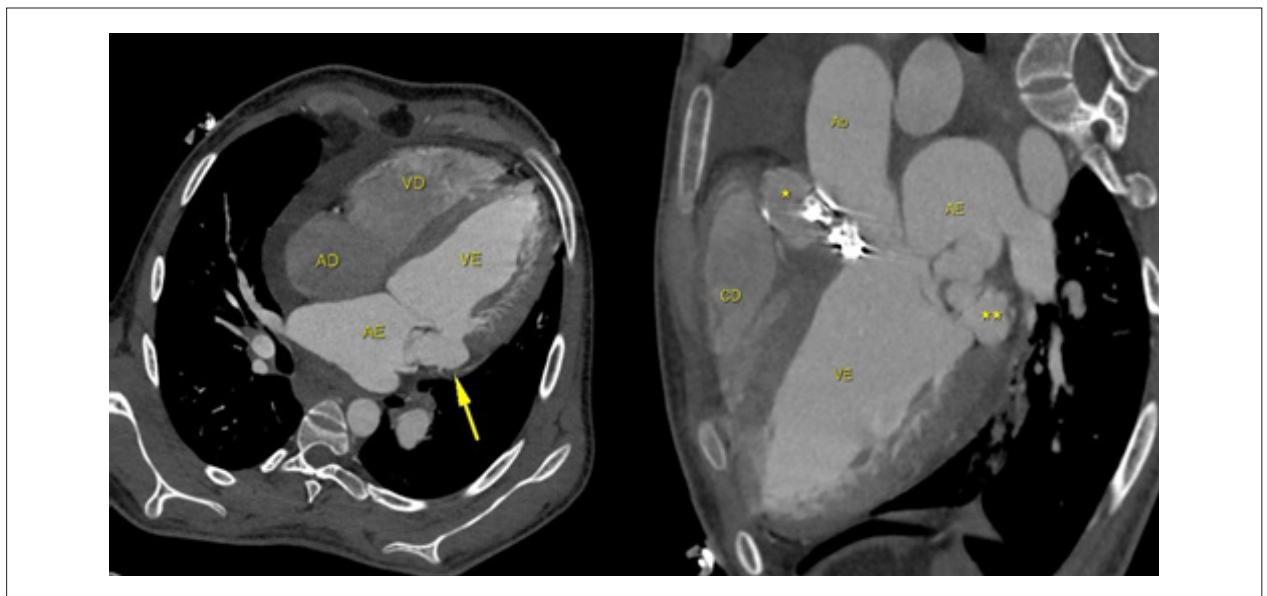


Figure 2 – In the left image, the yellow arrow indicates the mitral subvalvular aneurysm. In the right image, two hyperdense devices from previous percutaneous corrections can be seen: the first (*) is located next to the Sinuses of Valsalva; the second, just below, is in the left ventricular outflow tract.

were young (not older than 42), which may suggest the congenital nature of the disease. Of the six cases studied, only one was described in the subaortic region and five in the subaortic region, either below the anterior or posterior

leaflet. From their histopathological analyses, the authors observed no evidence of syphilis, tuberculosis, or rheumatic activity and noted thinned aneurysm walls. This led to the postulation that a weakness in the left ventricular wall

Case Report

Table 1 – Laboratory Tests

Leukocytes	13,000/mm ³	ASLO	109 U/mL (up to 250)
VHS	34 mm	HIV	Non-reactive
C-reactive protein 3.2	mg/dL	Treponema pallidum antibodies	Not detected
FAN	Non-reactive	Rheumatoid factor	Non-reactive
Bartonella and Coxiella Burnetii PCR	Negative	Epstein Barr and Cytomegalovirus IgM	Negative

Source: authors. HIV: human immunodeficiency virus; FAN: Anti-nuclear factor; VHS: erythrocyte sedimentation rate; ASLO: antistreptolysin O antibody.

in the atrioventricular groove region, when subjected to chronically increased intracavitary pressure, could lead to herniation of the wall and the formation of the aneurysm.

As previously described, left ventricular aneurysms are primarily ischemic, mainly after acute myocardial infarction. Exclusion of coronary artery disease is always the first step, whether aided by coronary angiography or CT angiography. In the latter cases, in addition to excluding coronary artery disease, it is also possible to evaluate the origin and course of the coronary arteries, excluding ischemia caused by anomalous origin or fistulas.

Several cardiac causes can also lead to ventricular aneurysms. In hypertrophic cardiomyopathy, the mid-ventricular region's involvement can lead to the development of an apical aneurysm due to increased pressure gradients in that area. Similarly, Chagas disease can cause apical aneurysms by affecting the ventricular wall in that region. Patients with myocarditis may exhibit a spectrum of ventricular function impairment, ranging from asymptomatic to cardiogenic shock. In more severe cases, extensive myocardial involvement with inflammation and cavity remodeling can occur, potentially resulting in aneurysms in various locations. Furthermore, infectious endocarditis with abscess formation and subsequent fistulization into the cavity can mimic subvalvular aneurysms, especially in the mitral-aortic curtain region.

A mitral subvalvular aneurysm can compromise valve function, leading to varying degrees of reflux, either due to distortion of the leaflets' anatomy or the valve ring itself. In this case, there was anatomical distortion and perforation of the posterior leaflet. We hypothesize that the flow from the aneurysm caused a jet lesion to the posterior leaflet, weakening it and resulting in perforation, leading to severe regurgitation into the left atrium.

The patient had a history of percutaneous repair of a Sinus of Valsalva Aneurysm and another aneurysm in the left ventricular outflow tract, supporting Chesler et al.'s hypothesis that the disease's pathophysiology involves ventricular wall weakening and subsequent aneurysm formation. Chen et al.⁵ also confirmed the histopathological finding in their case, showing fibrous tissue with a thin myocardium layer. Another unique aspect of our case is

that the patient has two subvalvular aneurysms in different locations (previous aortic; current mitral), which has not been previously described. The Sinus of Valsalva aneurysm, addressed in this surgery with a patch, further supports the notion that the patient has some reduction in ventricular and/or aortic wall resistance, predisposing them to these cavities' formation.

Conclusion

Left ventricular subvalvular aneurysm is a rare condition with unclear pathophysiological mechanisms, which can manifest with a range of clinical presentations, from incidental findings to heart failure with marked valve dysfunction. Ruling out treatable causes is crucial to halt its progression and determine the anatomical and functional implications to provide the most effective therapeutic approach.

Author Contributions

Conception and design of the research, writing of the manuscript and critical revision of the manuscript for intellectual content: Laydner JP, Siciliano AP, Silva RC, Felix AS, Dutra MF; obtaining financing: Laydner JP.

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Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.

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