Concurrent Congenital Pseudoaneurysm of Mitral-Aortic Intervalvular Fibrosa and Left Sinus of Valsalva Aneurysm

Valdano Manuel¹, Humberto Morais², Mauer Goncalves³, Ana Feijão⁴, Mário Fernandes⁵

¹Cardio-Thoracic Center, Clínica Girassol, Luanda, Angola
²Departamento de Cardiologia. Hospital Militar Principal. Luanda. República de Angola
³Serviço de Cardiologia. Hospital Américo Boavida. Luanda. República de Angola

Abstract

Either the pseudoaneurysm of mitral-aortic intervalvular fibrosa or the aneurysms of Valsalva are rare cardiac conditions. Concomitant diagnosis in the same patient is extremely rare with only one case report. Herein, we described an extremely rare case of concomitant pseudoaneurysm of mitral-aortic intervalvular fibrosa and aneurysm of left sinus of Valsalva in 21-year-old black man admitted with new-onset of heart failure (NYHA class III) diagnosed by transesophageal echocardiography. Despite medical therapy optimization for heart failure, the patient died waiting for cardiac surgery. Regarding the case, we discussed possible etiology, clinical presentation and the role of echocardiography in the diagnosis and surgical approach of this very rare association.

Introduction

Pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF) is a rare condition defined as a pseudoaneurysm at the interannular zone between the mitral and aortic valves and its communication with the left ventricular outflow tract between the left coronary and non-coronary aortic cusp and the anterior leaflet of the mitral valve [1]. Infective endocarditis, aortic valve surgery are the two most common etiologies [1]. Rarely, it is congenital in origin [2-5] or due to blunt chest trauma [6]. Clinically, it is manifested by symptoms and signs of infective endocarditis, followed by heart failure or dyspnea and chest pain due to compression of coronary vessels [1]. Transesophageal echocardiography is superior to transthoracic imaging in detection of pseudoaneurysm of MAIVF [7]. Sinus of Valsalva aneurysm (SVA) is also a relative uncommon but well-known cardiac condition [8]. A coexistence of both-P-MAIVF and SVA - is extremely rare with only one case report [9].

We presented a 21-old black male patient presented with new-onset, symptoms and signs of heart failure in which a pseudoaneurysm of MAIVF associated with left SVA were detected. Regarding the case, we discuss the possible etiology, clinical presentation and the role of echocardiography in the diagnosis of this very rare association.

Case Description

A 21-old black male patient presented with palpitation, fatigue and dyspnea, associated with cough, nocturnal paroxysmal dyspnea and peripheral edema over one month. He was treated in primary care with enalapril and furosemide, and antibiotic, and was referred to the Cardiology Department of Hospital Américo Boavida, Luanda, Angola. At admission, the patient reported a slight improvement of cough and peripheral edema, maintaining, however, the remaining symptoms He reported no fever, thoracic trauma, previous cardiac surgery or syncope.

Physical examination revealed a eupneic patient with a supine blood pressure of 100/60 mmHg, a pulse rate 106 beats/min, regular rhythm. Cardiac auscultation revealed grade 3/6 pansystolic murmur, which was audible in the apex, radiating to the axila. Pulmonary auscultation revealed bilateral basal rales. The remaining physical examination was unremarkable.

Complementary diagnostic exams included routine laboratory tests, which revealed no significant abnormalities; plan chest X ray showed cardiomegaly and pulmonary interstitial edema; and 12-lead electrocardiography revealed a sinus rhythm with a heart rate of 90 beats/min, and signs of left atrial and ventricular hypertrophy, with no other alterations.

Two-dimensional transthoracic echocardiography (TTE) revealed an echolucent area adjacent to the aortic valve that was freely com-
municating with left ventricular outflow tract (LVOT). The cavity expanded during systole and bulging into left atrium (Figure 1A) and collapsing during diastole (Figure 1B), consisting with diagnosis of pseudoaneurysm of the mitral-aortic intervalvular fibrosa (P-MAIVF). Left atrium and ventricle were dilated, with depressed systolic function. The right chambers were slightly dilated, and pericardium had no alterations. Furthermore, a left sinus of Valsalva aneurysm was also detected (Figure 1C). Color Doppler examination revealed severe mitral regurgitation and mild aortic regurgitation, and confirmed blood flow into cavity during systole (Figure 1D) and flow into the LVOT during diastole.

Figure 1: Transthoracic echocardiography (TTE) showed concurrent pseudoaneurysm of mitral-aortic intervalvular fibrosa (P-MAIVF) and sinus of Valsalva aneurysm (SVA). A - TTE longitudinal axis view showing P-MAIVF expanded during systole and bulging into left atrium (white asterisk) and B - collapsing during diastole. D - TTE longitudinal axis view color Doppler interrogation. The P-MAIVF fills during systole (black asterisk) C - TTE short axis view showed left SVA (white arrow). Ao – aorta; LA – left atrium LV – left ventricle; RV - right ventricle.

The diagnosis of pseudoaneurysm of the MAIVF along with left SVA was confirmed using transesophageal echocardiography (TEE) It demonstrated a large pseudoaneurysm of MAIVF communicating with LVOT (Figure 2A); Color Doppler interrogation showed that the P-MAIVF fills during systole (black asterisk) Figure 2B). A left SVA was also detected (Figure 2C,2D, Figure 3 and Movie 1). Furthermore, presence of vegetations and fistulous communication of the P-MAIVF to the left atrium or aorta were excluded by TEE. Despite the ideal medical treatment for heart failure, the patient died of congestive heart failure refractory to treatment waiting for cardiac surgery.

Figure 2: Transesophageal echocardiography (TEE) confirmed the findings of TTE. A - TEE longitudinal axis view of the aorta showing P-MAIVF expanded during systole and bulging into left atrium (white asterisk) B - TEE longitudinal axis view of the aorta color Doppler interrogation confirmed blood flow into pseudoaneurysm during systole (black asterisk). C,D - TEE short axis view showed left sinus of Valsalva aneurysm (white arrow). Ao – aorta; LA – left atrium LV – left ventricle; RA – Right atrium; RV - right ventricle.

Figure 3: A - Transthoracic echocardiography short axis view showed left sinus of Valsalva aneurysm (SVA) (asterisk) B - Transesophageal echocardiography (TEE) short axis view and C Doppler study confirmed the presence of left SVA asterisk and flow (arrowhead) between left coronary sinus and the aneurysm. D – TEE transgastric view showed left SVA (asterisk). Ao – aorta; LA – left atrium RA – Right atrium; RV - right ventricle.

Discussion

Pseudoaneurysm between the aortic and mitral valves in the region of the MAIVF was firstly described in 1966 by Waldhausen, et al. [10]
It is characterized by aneurismal dilatation between the left coronary and noncoronary aortic cusp and the anterior leaflet of the mitral valve.

The two most common etiologies of P-MAIVF are infective endocarditis and aortic valve surgery [1]. Congenital pseudoaneurysm of MAIVF in adults is very rare with a few case reported in literature [2-5]. In our case, no probable etiologic factor could be identified. The patient had no history of infective endocarditis, previous cardiac surgery or chest trauma. Moreover, the association of P-MAIVF with SVA supports the theory that in our case, the aneurysms are congenital in origin.

The clinical presentation can vary depending on the etiology, and complications. Known complications of MAIVF pseudoaneurysm are rupture into the pericardium, left atrium or aorta or coronary artery compression. Most common modes of presentation are features of active endocarditis, dyspnea and heart failure [1]. Our case presented with symptoms and signs of heart failure due to mitral regurgitation. Mitral regurgitation may be due to rupture of the pseudoaneurysm into left atrium, [4,5,12] or due to disconnection of the anterior mitral leaflet [12]. In present case, fistulous communication of the P-MAIVF to the left atrium was excluded by TEE; the mitral regurgitation was secondary to deformation of mitral valve.

Two-dimensional echocardiography with Doppler study plays an important role in the diagnosis of P-MAIVF [1,7] and in the diagnosis of SVA as well [8]. However, the sensitivity of TTE for the diagnosis of P-MAIVF is 43%, while the sensitivity of TEE is 90% [7]. The echocardiographic appearance of P-MAIVF is that a false lumen below the aortic valve annulus at the level of MAIVF. The pseudoaneurysmal cavity exhibits marked pulsatility with systolic expansion and diastolic collapse and communication with LVOT. Color Doppler interrogation demonstrates a toward and fro-flow into the cavity-flow from LVOT into the cavity in systole and reverse flow in diastole [7]. In our case, the diagnosis was suspected by TTE. Transesophageal echocardiography confirmed the diagnosis, and excluded presence of vegetation or rupture of P-MAIVF to left atrium or aorta. Furthermore, TEE gave important information about the mechanism of mitral regurgitation and spatial relationship between the P-MAIVF and left SVA.

Differential diagnosis of P-MAIVF include paravalvular/ring abscesses, contained/nor contained aortic root ruptures and sinus of Valsalva aneurysm and subaortic aneurysm. In our case, given the presence of both: pseudoaneurysm of MAIVF and left SVA, the correct diagnosis required a very careful study of the mitral and aortic rings as well as the MAIVF.

Surgery is recommended for the treatment of P-MAIVF [7] and SVA [8]. Despite the ideal medical treatment for heart failure, the patient died of congestive heart failure refractory to treatment waiting cardiac surgery.

Conclusion

We presented a patient in whom a pseudoaneurysm of mitral-aortic intervalval fibrosa associated with left sinus of Valsalva aneurysm was detected. This association is extremely rare with only one case report. In absence of obvious etiologic factor, we consider that the coexistence of P-MAIVF with SVA supports the theory that in our case the aneurysms are congenital in origin.

References


