

Submitral Aneurysm: An Echocardiography Study in a Tertiary Center in Angola

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Introduction

Submitral Aneurysms (SMA) are a relatively rare and poorly understood cardiac condition, although there are multiple reports of its incidence in specific populations [1-5]. Clinically it is manifested by symptoms and signs of heart failure due to mitral regurgitation, and/or ventricular arrhythmias. Transthoracic echocardiography (TTE) plays a key role in the definitive diagnosis of this pathology. Transesophageal echocardiography is of paramount importance in assessing the rupture of the aneurysm into the left atrium [1].

In recent years, a number of clinical cases using others imaging modalities have been published including real-time three-dimensional echocardiography, nuclear magnetic resonance, Cardiac Tomography (CT), as well as the use of different imaging modalities in the same patient [2,6-8]. The purpose of this paper is to present a brief review of the literature on the subject of SMA, regarding six patients studied in echocardiography laboratory of our Hospital in an eleven-year period.

Methodology

Based on database of the Echocardiography Laboratory of the Hospital Militar Principal/Instituto Superior, demographics data, echocardiography features and data from follow-up of patients with the diagnosis of SMA, in the period from May 2001 to April 2012, were analyzed.

All patients underwent Transthoracic Echocardiography (TTE). Four patients underwent real time three-dimensional TTE (RT3DTTE), three patients underwent Transesophageal Echocardiography (TEE), two patient performed real time three-dimensional TEE (RT3DTEE). One patient underwent cardiac computed tomography angiography.

TTE and Doppler studies were performed in accordance with the recommendations of the American Society of Echocardiography [9,10]. TEE studies were performed in accordance with the recommendations of the European Society of Cardiology [11]. Real time three-dimensional echocardiography was performed as previously reported [12].

Results

Demographic data, clinical presentation, NYHA functional class and follow-up are shown in Table 1. Data from transthoracic echocardiography, cardiac complications and associated pathologies are presented in Table 2. During the review period six patients had a diagnostic of SMA. Four are male and two are female. The mean age was 27.3 ± 7.0. Two patients had rupture of the aneurysm into the left

No	Gender	Age	Symptoms	Class NYHA	Follow-up
1	Male	21	Dyspnoea	III	Lost
2	Male	27	Dyspnoea	III	Lost
3	Male	22	Chest pain	I	Surgery
4	Female	30	Dyspnoea	I	Lost
5	Female	40	Dyspnoea	II	Lost
6	Male	24	Dyspnoea	III	Refused Surgery

Table 1: Demographic data, clinical presentation, NYHA functional class, and follow up.

atrium. One patient presented together with SMA an aneurysm of the right sinus of Valsalva dissecting into the interventricular septum and left ventricular noncompaction. Five of the six patients had severe mitral insufficiency. All patients were proposed for surgery: one was successfully operated in Angola, in Hospital Josina Machel, one refused surgery and died one year after the diagnosis was made, the remaining four patients were lost in follow-up.

Discussion

Once SMA is a relative rare disease there are no data on the prevalence and incidence of this condition in general population. However, there are few epidemiological data reported in the literature that it is worth highlighting. In a study by Gaultier et al. at Hospital de Miamex in Nigeria, SMA accounted for 0.04% of hospitalizations and 0.34% of cardiovascular disease [4]. Sliwa and Mocumbi found 10 cases of SMA in 5200 patients with heart failure diagnosed in the period 2006-2008 in Soweto, corresponding to 0.19% of cases [13]. Morais et al. in Angola found SMA in 4.1% of congenital heart disease diagnosed in patients aged greater than or equal to 15 years within 10 years [14].

Its etiology is still the subject of debate, although there is agreement that it can be congenital or acquired. The most common causes of acquired SMA are inflammatory or infectious diseases or trauma. Among infectious causes are syphilis, tuberculosis and infectious endocarditis [3,15]. It can also result from cardiomyopathy [4,16]. Congenital SMA arises from a defect of a valve ring and is sometimes associated with sinus of Valsalva aneurysm, which suggests a congenital weakness of aortic or mitral annulus insertion in the underlying

NO	Year	Systolic Function	Mitral regurgitation	Complications	Other findings
1	2001	Good	Severe	Rupture into LA	None
2	2004	Good	Severe	None	None
3	2008	Good	Mild	None	None
4	2010	Good	Severe	None	None
5	2010	Good	Severe	Rupture into LA	None
6	2011	Severe Depressed	Severe	None	SVA; LVNC

Table 2: Date of diagnosis, and findings on echocardiography, LA: left atrium, SVA: sinus of Valsalva aneurysm, LVNC: left ventricular non-compaction.

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myocardium, probably due to a congenital defect in the posterior portion of the mitral annulus [3-5,15,17,18].

The most common forms of clinical presentation of SMA are congestive heart failure, mitral insufficiency, thromboembolism, and ventricular arrhythmias [1,3,5,17-21]. In some cases, it presents as myocardial ischemia due to compression of the coronary arteries by the aneurysm or as cardiogenic shock [17,22,23]. In present study, the main clinical presentation was heart failure: 50% of the patients were in class III of NYHA, requiring hospitalization. Two patients with dyspnoea were referred to the echocardiography laboratory in class I and II of NYHA respectively; one patient had complaints of atypical chest pain. In contrast, none of the patients debuted with thromboembolic events, ventricular arrhythmias or myocardial ischemia. In all patients, holosystolic murmur in the mitral area suggestive of mitral regurgitation was observed.

Transthoracic echocardiography was diagnostic in all cases, revealing an aneurismal dilatation clearly in submitral location behind to the posterior leaflet of the mitral valve, communicating with the left ventricular cavity through one or more necks (Figures 1 and 2).

In the present study, transthoracic echocardiogram showed Mitral Regurgitation (MR) in all patients. MR was considered severe in five of them and mild in one patient (Figures 3 and 4). It is in agreement to those reported by other authors [4,24]. In the series reported by Antunes eight of the nine operated patients had significant mitral regurgitation. In the series reported by Gaultier all patients had a murmur of mitral regurgitation [2,24].

Rupture of SMA into the left atrium is a serious complication. In the present series, this complication was found in two of six patients, constituting 33.3% of cases. Antunes found rupture of the aneurysm into the left atrium in one patient of nine operated patients, representing 11% of cases [24]. These data, together with cases of rupture of the



Figure 3: (A) Color-Doppler TTE study in apical four-chamber view showing severe mitral regurgitation. (B) Color-Doppler TEE study confirming the severity of mitral regurgitation.

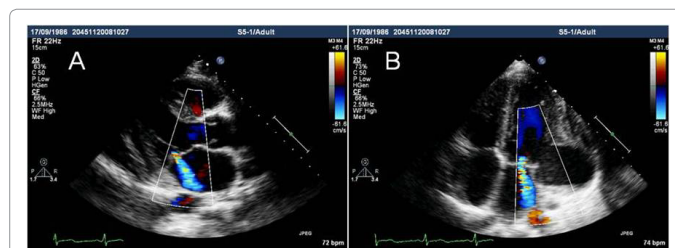
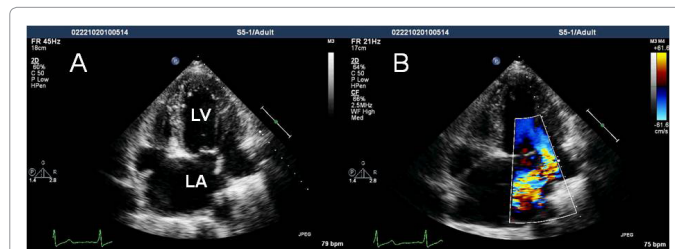
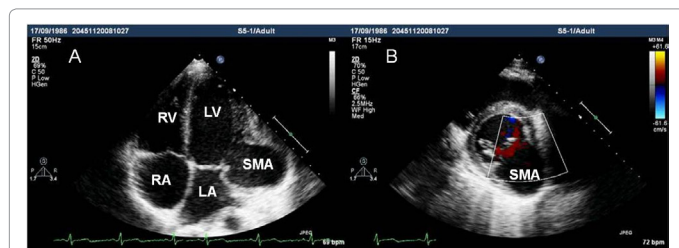


Figure 4: (A) Color-Doppler TTE in parasternal long axis view revealing mild mitral regurgitation. (B) Color-Doppler TTE apical four-chamber view showing mild mitral regurgitation.



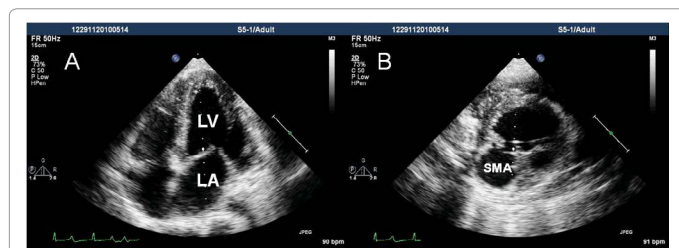
LV: Left ventricle; LA: Left atrium

Figure 5: TTE (A) apical four chambers, revealing submitral aneurysm with possible rupture into the left atrium. (B) Color-Doppler in apical four chambers that reveals paravalvular regurgitation jet into the left atrium, suggesting rupture of the aneurysm into the left atrium.



RV: Right ventricle; LV: Left ventricle; LA: Left atrium; AD: Right atrium; SMA: Submitral aneurysm

Figure 1: TTE (A) Apical four chambers view reveals submitral aneurysm. (B) Short axis view shows aneurismal dilatation behind of the posterior mitral valve leaflet near the anterolateral commissure.



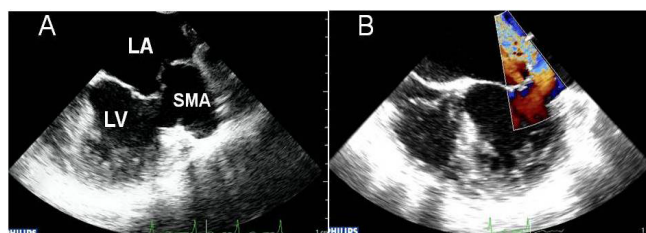
LV: Left ventricle; LA: Left atrium; SMA: Submitral aneurysm

Figure 2: TTE (A) Apical four chambers view apparently unchanged. (B) Short axis view shows aneurismal dilatation behind of the posterior mitral valve leaflet near the posteromediana commissure.

aneurysm into the left atrium reported in the literature leads us to suppose that this complication may not be as rare as commonly thought. This complication should be suspected whenever the transthoracic echocardiogram noted a paravalvular jet of mitral regurgitation (Figure 5) [25-28].

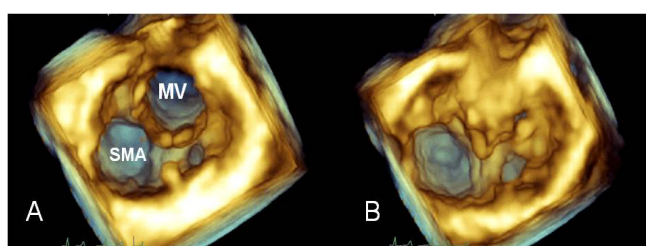
The transesophageal echocardiography is essential in diagnosis of this complication, clearly showing the rupture of the aneurysm into the left atrium in the two cases described herein, one of which is showed in Figure 6. Thus, given the suspicion of rupture of the aneurysm into the left atrium we strongly recommend performing TEE whenever possible. Multidetector cardiac CT may also be useful in this context [28].

Recently, real-time three-dimensional echocardiography has been shown to be superior to conventional echocardiography in the evaluation of SMA [6,25]. In our experience, the real-time three-dimensional echocardiography was useful in evaluating the relationship of the aneurysm with other cardiac structures including the mitral valve and its leaflets, the anatomical characterization of the aneurysm. Moreover in one patient, real-time three-dimensional echocardiography has allowed the identification of a bilobular aneurysm with two necks communicating with the left ventricle, providing additional data



LV: Left ventricle; LA: Left atrium; SMA: Submitral aneurysm

Figure 6: (A) TEE revealing submitral aneurysm with rupture into the left atrium. (B) Color-Doppler study showing regurgitation jet from the aneurysm into the left atrium, confirms the rupture of the aneurysm into the left atrium.



MV: Mitral valve; SMA: Submitral aneurysm

Figure 7: RT3DTTE (A) view from the left ventricle in diastole showing submitral bilobular aneurysm. (B) The same view in systole.

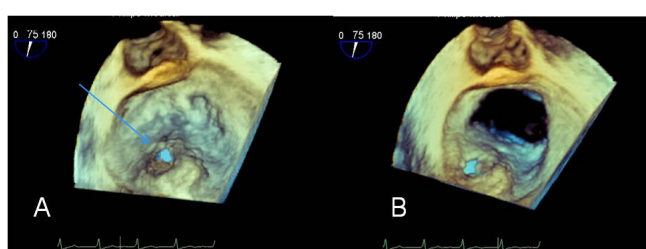


Figure 8: RT3DTTE (A) "en face" view from the left atrium in systole, showing the rupture of the aneurysm into the left atrium (blue arrow). (B) The same view in diastole.

to two-dimensional echocardiography. Real-time 3DTEE provided unambiguous images in case of rupture of the aneurysm into the left atrium (Figures 7 and 8).

Average age found in present series (27.3 ± 7.0) is similar to those reported by Antunes (28 ± 3), but slightly higher than reported by Gaultier (20 years). In the present study a 3:1 male: female ratio was found it is higher than reported by Antunes who found a 1.25:1 ratio [4,24].

Definitive treatment of SMA requires surgery [23,24,29-32]. The technique that is now more frequently utilized was described by Antunes in 1987 where the aneurysm is approached through the atrium [24]. This technique allows excluding the aneurysm and often preserving the mitral valve [22]. When this is not possible, the valve should be replaced.

Conclusions

Submitral aneurysm, although uncommon, should always be entered in the list of differential diagnosis in young patients presenting with a murmur suggestive of mitral insufficiency or signs and symptoms

of heart failure or thromboembolic events. The definitive diagnosis is made by transthoracic echocardiography in the presence of an aneurismal dilatation in submitral location behind the posterior leaflet that communicated with the left ventricular cavity through one or more necks. The Doppler study frequently reveals mitral regurgitation that is often severe.

In the present series the most frequent complication was rupture of the aneurysm into the left atrium. Transesophageal echocardiography was fundamental in the diagnosis of this complication in both cases, and we strongly recommend performing TEE whenever possible. Contrast-enhanced Cardiac CT may also be useful in this context, clearly revealing the passage of the contrast from the aneurysm into the left atrium through the point of rupture. Cardiac CT angiography also allows evaluating the coronary arteries.

Real-time three-dimensional echocardiography is not necessary for the diagnosis of SMA. However it is very useful in the evaluation of the relationship of the aneurysm with the other cardiac structures. In the evaluation of the anatomical characteristics of the aneurysm allowing the identification of one or more apertures through which aneurysm communicates with the left ventricle, providing additional data to two-dimensional echocardiography, and improving the plan for surgery. Real-time 3DTEE provides clear and unique images in cases of the rupture of the aneurysm into the left atrium.

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