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 [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 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 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

<table>
<thead>
<tr>
<th>NO</th>
<th>Year</th>
<th>Systolic Function</th>
<th>Mitral regurgitation</th>
<th>Complications</th>
<th>Other findings</th>
</tr>
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<tr>
<td>1</td>
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<td>Severe</td>
<td>Rupture into LA</td>
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</tr>
<tr>
<td>2</td>
<td>2004</td>
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<td>Severe</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>3</td>
<td>2008</td>
<td>Good</td>
<td>Mild</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>2010</td>
<td>Good</td>
<td>Severe</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>5</td>
<td>2010</td>
<td>Good</td>
<td>Severe</td>
<td>Rupture into LA</td>
<td>None</td>
</tr>
<tr>
<td>6</td>
<td>2011</td>
<td>Severe Depressed</td>
<td>Severe</td>
<td>None</td>
<td>SVA: LVNC</td>
</tr>
</tbody>
</table>

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 aneurysmal 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 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 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.

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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.
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 submtral 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.

References