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 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 ± 7.6. 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

| 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  | 2011 | Severe Depressed | Severe               | Rupture into LA | None          |

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 dyspnea 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 Gautier 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 echocardiogram noted a paravalvular jet of mitral regurgitation (Figure 3) [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.
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

1. Morais H, Branco LM, Cunha R, Martins T (2007) Rupture of a submural ventricular aneurysm into the left atrium diagnosed by transesophageal echocardiography. Rev Port Cardiol 26: 367-372.
2. Chen CC, Hsiung MC, Wei J, Chang WT, Yin WH, et al. (2005) Mitral annular subvalvular left ventricular aneurysm. Echocardiography 22: 434-437.
3. Ribeiro PJF, Mendes RGG, Vicente WVA, Menardi AC, Évora PRB (2001) Aneurisma ventricular subvalvar mitral. Case presentations and surveys of published Brazilian cases. Arq Bras Cardiol 76: 395-398.
4. Gauthier Y, Cénac A, Aoua HO, Touteé I (1989) [Idiopathic annular subvalvar aneurysm. Contribution of echography apropos of 5 cases]. Arch Mal Coeur Vaiss 82: 897-902.
5. Damasceno A, Haussie AO, Ferreira B, Teixeira R (1993) [Mitral subvalvular aneurysm]. Rev Port Cardiol 12: 963-971, 902.
6. Hotte VT, Cruz CB, Rassi Ddo C, Vieira ML, Mathias W Jr, et al. (2010) Subvalvular mitral pseudoaneurysm evaluated by three-dimensional echo. Echocardiography 27: 473-475.
7. Warren O, Athanasius T, Massey R, Hamady M, Stanbridge R (2006) Large annular subvalvar left ventricular aneurysm: diagnostic evaluation using computed tomographic angiography. Tex Heart Inst J 33: 529-531.
8. Kharwar RB, Sethi R, Sanguri R, Singh V, Narain VS (2014) Multimodality imaging of submural left ventricular aneurysm. Echocardiography 31: E24-47.
9. Henry WL, DeMaria A, Gramia R, King DL, Kisslo JA, et al. (1980) Report of the American Society of Echocardiography Committee on Nomenclature and Standards in Two-dimensional Echocardiography. Circulation 62: 212-217.
10. Quiliones MA, Otto CM, Stoddard M, Waggoner A, Zoghbi WA; Doppler Quantification Task Force of the Nomenclature and Standards Committee of the American Society of Echocardiography (2002) Recommendations for quantification of Doppler echocardiography: a report from the Doppler Quantification Task Force of the Nomenclature and Standards Committee of the American Society of Echocardiography. J Am Soc Echocardiogr 15: 167-184.
11. Flachsrandk FJ, Decoodt P, Fraser AP, Daniel WG, Roelandt JRTC (2001) Guidelines from working group. Recommendation for performing transesophageal echocardiography. For the subgroup on transesophageal echocardiography and valvular heart disease, on behalf of working group on echocardiography of the European Society of Cardiology. Eur J Echocardiography 2: 8-21.
12. Morais H, Martins T (2010) Ecocardiografia transesofágica tridimensional em tempo real: Experiência inicial em Angola An HMP/IS 1: 13-19.

13. Sliwa K, Mocumbi AO (2010) Forgotten cardiovascular diseases in Africa. Clin Res Cardiol 99: 65-74.

14. Morais H, Martins T Cunha R (2014) Spectrum of Congenital Heart Disease in Adolescents and Adults in Angola: An Echocardiography Study “2001-2009”.

15. Deshpande J, Vaideeswar P, Sivaraman A (2000) Subvalvular left ventricular aneurysms. Cardiovasc Pathol 9: 267-271.

16. Cenac A, Chaingeau C, Sueur JM, Orfila J (2002) Submitral annular aneurysm, postpartum cardiomyopathy, and anti-Chlamydia pneumoniae antibodies in Niamey (Niger). Med Trop (Mars) 62: 81-84.

17. Chockalingam A, Gnansou I, Alagesan R, Subramaniam T (2004) Congenital submitral aneurysm and sinus of valsalva aneurysm. Echocardiography 21: 325-328.

18. Mohanty A, Saxena A (2003) Submitral aneurysm: unusual echocardiographic features. Heart 89: 552.

19. Esposito F, Renzulli A, Festa M, Cerasuolo F, Caruso A, et al. (1996) Submitral left ventricular aneurysm. Report of 2 surgical cases. Tex Heart Inst J 23: 51-53.

20. Moisés VA, Vieira Filho JP, Andrade JL, Leão LE, Martinez Filho EE (1993) [Submitral left ventricular aneurysm in a Brazilian Indian]. Arq Bras Cardiol 60: 343-345.

21. Chi NH, Yu HY, Chang CI, Lin FY, Wang SS (2004) Clinical surgical experience of congenital submitral left ventricular aneurysm. Thorac Cardiovasc Surg 52: 115-116.

22. Skoularigis J, Sareli P (1997) Submitral left ventricular aneurysm compressing the left main coronary artery. Cathet Cardiovasc Diagn 40: 173-175.

23. Terzi CB, Pomerantzeff PM, Monachini MC, Kopel L, Medeiros CC, et al. (1996) [Mitrval subvalval aneurysm of the left ventricle]. Arq Bras Cardiol 67: 351-353.

24. Antunes MU (1987) Submitral left ventricular aneurysms. Correction by a new transatrial approach. J Thorac Cardiovasc Surg 94: 241-245.

25. Peters F, Essop R (2011) Congenital submitral aneurysm with rupture into the left atrium: assessment by 2D and 3D transesophageal echocardiography. Echocardiography 28: E121-124.

26. Simpson L, Duncan JM, Stainback RF (2006) Perforated submitral left ventricular aneurysm resulting in severe mitral annular regurgitation. Tex Heart Inst J 33: 492-494.

27. Sai Krishna C, Naresh Kumar PV, Panigrahi NK, Suman K (2007) Submitral aneurysm with left atrial communication. Eur J Cardiothorac Surg 32: 547-549.

28. Duggal B, Inania R, Panchani N (2012) Rupture of a submitral aneurysm into the left atrium. Pediatr Cardiol 33: 854-856.

29. Pektok E, Cikirikcioglu M, Didier D, Kalangos A (2008) Submitral left ventricular aneurysm: a rare but challenging pathology to treat. J Card Surg 23: 533-535.

30. Sharma A, Kulkarni V, Barde S, Bobade S, Bhargava V, et al. (2009) Submitral aneurysmorrhaphy with mitral valve replacement: A case report. LTCVS 26: 118-120.

31. Singh S, Agarwal S, Dutta N, Mishra S, Upreti L, et al. (2012) Surgical repair of a submitral aneurysm in a three-year-old child. J Card Surg 27: 238-240.

32. Ran JN, Gaijar T, Desai N (2011) Submitral left ventricular aneurysm – our experience Indian J Thorac Cardiovasc Surg 27: 91-95.