

Idiopathic Left Ventricle Aneurysm

Gustavo Restrepo Molina¹, Salim Ahumada Zakzuk¹, Marcelo Luiz Campos Vieira^{2,3,4}

Clínica Medellín, Medellín, Colombia - COL¹, Instituto do Coração (InCor), São Paulo, SP - BR², Faculdade de Medicina, Universidade de São Paulo (FMUSP), São Paulo-BR³, Hospital Israelita Albert Einstein, São Paulo-BR⁴

Introduction

Idiopathic left ventricle aneurysm is an uncommon clinical entity and seldom described in literature which usually originates in the annular subaortic or sub-mitral region, and its non ischemic etiology remains unknown. This pathology may have different clinical presentations ranging from mitral regurgitation and heart failure to ventricular arrhythmias potentially fatal as well as sudden death. Even though the vast majority of cases were described among black population, it has also been described among Caucasian population with lower incidence though. At present, the use of multimodality images allows for better screening and characterization of the lesions. Hereinafter is presented a case report of a patient without evidence of known coronary heart disease, and presence in echocardiography and cardiac magnetic resonance imaging of aneurysmal idiopathic mitral sub-valvular injury compromising the functioning of mitral valve, discarding other etiologic causes, the surgical correction being concluded with an adequate evolution after the procedure. The diagnosis was confirmed by histopathological examination, serology and immune profile study.

Case Report

Female patient, 46 years, half-caste, married and a housewife. Medical appointment due to her clinical condition evolving for 4 months consisting of dyspnea with moderate efforts (NYHA II functional class). She refers episodes of oppressive chest pain when walking without typical irradiance and disappearing when she rests. She does not complain about congestive symptoms; there is no paroxysmal nocturnal dyspnea or edema. During questioning, there were no syncope episodes and she has not complained about tachycardia or palpitations. Pathological history of hypothyroidism was diagnosed in 2011 and optic neuritis for the last 6 months which was managed with acyclovir by the Ophthalmology department due to suspicion of viral etiology. There is no history of rheumatic diseases or relevant systemic diseases. She denies toxicological history or previous chest trauma. On physical examination she is in general good condition but her blood pressure values

Keywords

Hypertrophy, Left Ventricular/surgery; Heart Aneurysm; Mitral Valve/injuries; South Africa; Young Adult.

are high (150/65 mmHg) with mitral systolic murmur II/VI. An ECG was performed indicating previous lower necrosis and inversion of T wave on II, III and aVF derivations. The patient was sent to the non-invasive Cardiology department to perform a Transthoracic Echocardiogram revealing eccentric left ventricular hypertrophy with moderate systolic dysfunction (ejection fraction of around 40-45%), left ventricular dilatation with telesystolic diameter of 35 mm/m² and diastolic dysfunction due to relaxation disorder. The analysis of segmentary contractility shows segmentary contractility disorders caused by akinesia and thinning of all segmentst of the lower wall, with a large aneurysmal sac showing evidence of flow inside with Color Doppler (neck/sac ratio 0.4) (Figure 1). The mitral valve is morphologically normal with moderate eccentric failure to the back wall toward the wall of the atrium.

Aiming at discarding ischemic etiology as first diagnostic possibility, she was subjected to a coronariography and left heart catheterization, showing epicardic coronary without angiographically significant lesions and an increment of end diastolic left ventricular pressure (20 mmHg). The cardiac magnetic resonance imaging showed aneurysm of the lower wall with neck of 12.2 mm and 51.1 x 27.7 mm diameter, with homogeneous late enhancement with gadolinium and no evidence of aneurysm wall rupture into the pericardium, eliminating ventricular pseudoaneurysm (Figure 2). Serological and immunological profile was negative with the values within the normal parameters (Table 1). Therefore, the cardiovascular surgery experts proceeded to an evaluation to determine the type of intervention. The patient was taken into surgery. Dor procedure was carried out to exclude the aneurysm through the placement of a Dacron patch and closing the cavity without aneurysmectomy (Figure 3 / Table A and B). During intraoperative procedure, the mitral valve replacement by nr. 29 mechanical prosthesis was decided with the adequate postoperative gradients. Histological samples have been taken which have shown abundant fibrin and connective tissue with the hematoxylin-eosin and trichrome colorations. By staining the striated muscle with desmin for identification, traces of myocardium can be seen on the outside of the ventricular wall (Figure 3 / Table C and D). The biopsy showed no signs of vasculitis, myocarditis or granulomas. Therefore, the idiopathic etiology of aneurysm was considered as a consequence of the absence of identifiable causes through multiple extension studies.

Discussion

Aneurysms of the LV can be divided according to its etiology in congenital or acquired, whether the cause is of cardiac origin or not. The most common cause in clinical practice is

Mailing Address: Dr. Gustavo Restrepo Molina •
Clínica Medellín - Calle 53 N.46-38, Medellín - Colombia
E-mail : gustavorestrepomd@une.net.co
Manuscript received November 27, 2013; revised November 28, 2013;
accepted February 09, 2014.

Case Report

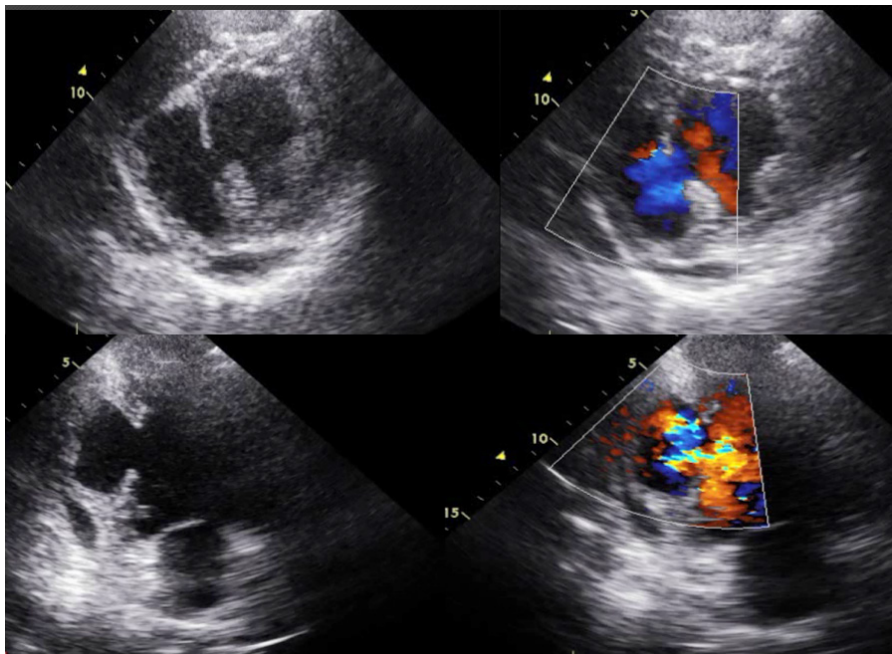


Figure 1 - Echocardiogram with evidence of aneurysm in the lower wall with flow inside with color Doppler. Akinesia in the lower wall.

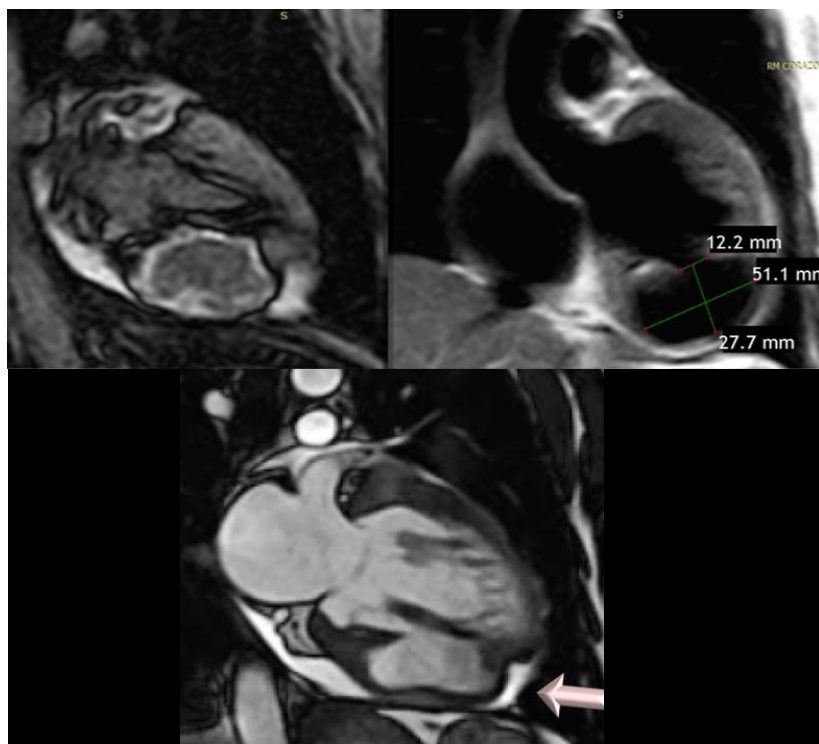


Figure 2 - Cardiac Magnetic Resonance Imaging. In the upper-left image late enhancement is evidenced with gadolinium of ventricular aneurysm. Top right image with diameters of the aneurysmal sac. Bottom cine imaging with location of aneurysm in lower wall.

Table 1 - Serology and immune profile. Autoimmune, viral and parasitic causes are discarded.

Lab tests/Serology and immune profile	
Trypanosomes Cruzi (Chagas), Acps	Negative
Antinuclear Acp	Negative
Serology HIV, Acps	Negative
Extractable Nuclear Acps	
* Anti-Sm	2.1 (NEG)
* Anti-RNP	2.5 (NEG)
* Anti-SSA	3.1 (NEG)
* Anti-SSB	2.7 (NEG)
THS/Free T4	10.5 / 1.03

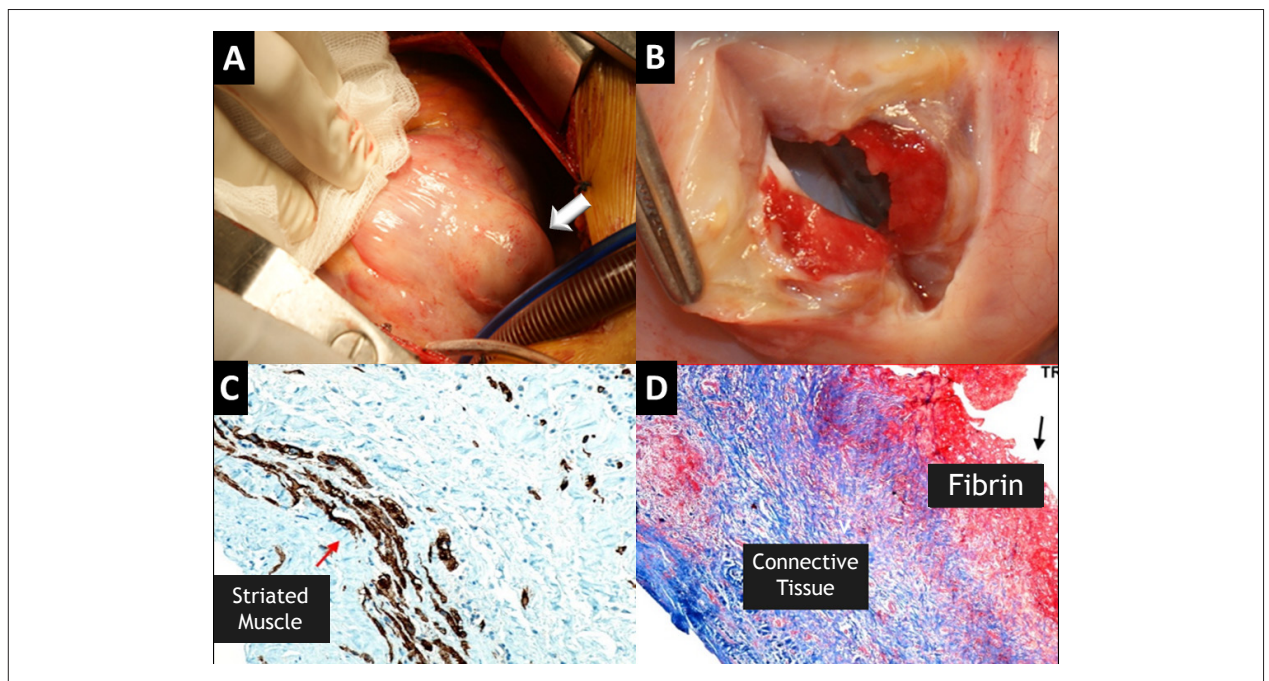


Figure 3 - (A). Photo of aneurysm during surgical procedure, in which ventricular wall integrity without breaking for pericardium is evidenced.(White arrow). (B). Aneurysm after incision of the sacin in which a small neck communicating with the ventricular cavity is seen. (C). Histologic myocardial biopsy sample with desmin coloring for identification of striated muscle, which is evidenced in coffee like color (Red arrow). (D). Trichrome coloring with fibrin in deep red and blue connective tissue. Due to the following findings it is considered that there is a ventricular wall with striated muscle remaining on its outside.

usually attributed to acute myocardial infarction, however, they can develop in the context of other pathologies as RV arrhythmogenesis dysplasia, hypertrophic cardiomyopathy or myocarditis. Other non-cardiac causes include sarcoidosis, Chagas disease, systemic lupus erythematosus, tuberculosis and HIV. On some occasions it may not be possible to determine the etiology of this pathology which is catalogued as idiopathic¹ Abrahamse et al² in 1962 described a pathology in the report of 12 cases in patients coming from Nigeria, introducing the expression “Annular subvalvular left ventricular

aneurysm” due to its location immediately beneath the mitral and aortic valves, and extending around and in the substance of the annular fibrous ring where they are formed. Subsequently, Chesler et al.³ in 1965 described other 6 cases of similar characteristics, suggesting weakness of the left ventricular wall in the auriculoventricular groove region with herniation of endocardium by ventricular pressures influence, and resulting in the formation of a fibrous wall aneurysm with calcium and thrombus deposits in their walls, sometimes with adherence to adjacent pericardium.

Case Report

The idiopathic sub-mitral LV aneurysm (ISLVA) is a rare entity, of non-ischemic origin, described most commonly in black young individuals in Africa with an estimated incidence of 34 per each 10,000 cardiovascular diseases⁴. Although initially considered unique to this population, probably because the studies were directed to this continent, there are reports of cases in the Caucasian population. The higher prevalence in certain ethnic groups and the absence of a clear etiology suggests a congenital etiology. Typically, they have been described at the basal level of the heart, in the posterolateral wall and/or bottom, occurring just below the back flap and associated with the mitral regurgitation in structurally normal valve, invariably with different degrees of severity. So far it has not been linked to a higher incidence of other congenital anomalies⁵.

The ISLVA seems to be caused by a change in the junction of the heart muscle with the fibrous structure of the heart. It may vary in size from millimeters to several centimeters, extending behind the left atrium or LV, or even distorting the mitral valve apparatus causing loss of posterior flap support and mitral insufficiency. ISLVA patients may show asymptomatic for years, and sometimes the first symptom can be the appearance of ventricular arrhythmias, sudden death or systemic embolism. Clinically, it is characterized by symptoms of heart failure, mitral insufficiency and on occasions due to the expansion of the aneurysm the compression of coronary vessels may occur⁶.

The echocardiogram is the initial method of diagnosis because it is a non-invasive and cost-effective technique, and the absence of lesions in coronary anatomy during coronariography, associated with the typical location allows a high suspicion of this entity. Other types of imaging used are the ventriculography, CT, and magnetic resonance imaging of heart, the latter being perhaps the one that provides a better anatomical characterization of the aneurysm allowing setting the surgical planning for its correction. On several occasions, cardiac mesothelial excrescences associated with idiopathic aneurysms have been described, which are a benign non neoplastic injury and should be subjected to histopathological study for a proper diagnosis and differentiation of other neoplasms or metastatic carcinomas⁷.

The definitive treatment of ISLVA is surgical however, if the surgical risk doesn't allow it, medical treatment and cardio defibrillator implant in case of malignant ventricular arrhythmia, should be considered. The main surgical indications are: the size of the aneurysm, growth during follow-up, formation of thrombi, presence of symptoms with hemodynamic deterioration and the presence of arrhythmias (VT/VF)⁸. In the case of our patient, symptoms of heart failure, the size of the aneurysm and the presence of significant mitral valvulopathy determined the mitral valve replacement and Dor surgery for the correction of the defect with its the exclusion.

References

1. Paul M., Schafers M., Grude M., Reinke F, Juergen KU., Fischbach R, et al. Idiopathic left ventricular aneurysm and sudden cardiac death in young adults. *Europace*. 2006; 8(8):607-12..
2. Abrahams DG, Barton CJ, Cockshott WP, Edington GM, Weaver EJM. Annular subvalvular left ventricular aneurysm. *Q J Med*. 1962; 31: 345-60.
3. Chesler E, Joffe N, Schamroth, L, Meyers, A. Annular subvalvular left ventricular aneurysms in the South African Bantu. *Circulation*. 1965;32:43-51.
4. Kazemi, B. Arya, A. Haghjoo, M. Sadr-Ameli, MA. Idiopathic Submitral Left Ventricular Aneurysm: an unusual substrate for ventricular tachycardia in Caucasians. *Indian Pacing Electrophysiol J*. 2005; 5(3):244-9.
5. Esposito, F. Renzulli. A. Festa, M. Cerasuolo, F. Caruso, A. Sarnicola, P, et al. Submitral left ventricular aneurysm. Report of 2 surgical cases. *Tex Heart Inst J*. 1996; 23(1):51-3.
6. Ribeiro, PJ. Mendes, RG. Vicente, WV. Menardi, AC. Evora, PR. Submitral left ventricular aneurysm. Case report and review of published Brazilian cases. *Arq Bras Cardiol*. 2001; 76(5):395-402.
7. Exadaktilos, N. Marsonis, A. Moschovitis, I. Giannopoulos, D. Gavaliatsis, I. Papadimitriou, C. Kakkavos, D. Left ventricular aneurysm with normal coronary vessels and cardiac mesothelial excrescences. *Hellenic J Cardiol (HJC)*. 2002;43:71-5.
8. Chen, CC. Hsiung, MC. Wei, J. Chang, WT. Yin, WH. Young, MS. Mitral annular subvalvular left ventricular aneurysm. *Echocardiography*. 2005; 22(5):434-7.