

## Submitral Left Ventricular Aneurysm. Case Report and Review of Published Brazilian Cases

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*Submitral left ventricular aneurysm is a cardiac pathology widely recognized, but relatively unknown, occurred almost exclusively in African black patients. Although still this idea of racial prevalence exists, cases have been described in patients of all the races. Ten Brazilian cases were reported. One of them was presented inside an Italian paper that refers the surgical treatment of a Brazilian patient of black race. We reported one more submitral left ventricular aneurysm case in a brown female patient, with antecedents of peripheral thromboembolism initially not identified as consequence of the cardiac pathology.*

Submitral left ventricular aneurysm is a cardiac pathology widely recognized, but relatively unknown. For a long time, it was believed that this type of aneurysm was a condition that occurred almost exclusively in African patients of the black race. Although this idea of racial prevalence still exists, cases have been described in patients of all the races including, black, white, and yellow races and also in Brazilian Indians. It was described in 1812 by Corvisart and, since this time, around 100-120 cases of these aneurysms have been reported<sup>1</sup>. The period covered by the MEDLINE database (1966 - 1999) indicates the existence of 41 published works, including five Brazilian papers<sup>2-6</sup> that report nine cases and one Italian paper that reports the surgical treatment of a Brazilian patient of the black race<sup>7</sup>. In this article one more case is reported of submitral left ventricular aneurysm in a brown female patient, with antecedents of peripheral thromboembolism initially not identified because of the cardiac pathology.

### Case Report

A 59-year-old female, brown in color who presented at

the first medical consultation with approximately a 30-year history of dyspnea on great effort. She was born in the endemic zone for Chagas heart disease, without any antecedents of tobacco use, hypertension, or diabetes. Her family history included some cases of cardiopathy. In the past, she had undergone a femoral artery embolectomy plus sympathectomy. She did not have any abnormal alterations during the physical examination that deserve mention. On the occasion of the first consultation, clinical laboratory tests were requested that disclosed the following: negative RIF for Chagas disease, glucose - 84mg/dL, cholesterol - 215mg/dL, triglycerides - 205mg/dL.

She underwent a new medical evaluation because she was experiencing daily precordial pain when walking. On this occasion, it was decided that the diagnostic inquiry should be confirmed. The electrocardiogram showed sinus rhythm with rectification of the ST segment in inferior and lateral facies, with a chest X-ray showing the pulmonary area without any abnormalities and the cardiac area with a calcified area next to the region corresponding to the left cardiac appendage (cyst, calcified myxoma?). Doppler echocardiography revealed cardiac chambers with normal dimensions, systolic performance of the left ventricle preserved, and the discrete presence of mitral valve reflux. Of significance was the presence of an aneurysmatic dilatation in the posterior area of the left ventricle with an ostial communication between the aneurysm and this cardiac chamber. The flow register through this communication was not obtained because the position of the aneurysm in the mitral subannular plan was clear (Fig. 1A).

In the following hemodynamic study, the manometry revealed a pressure gradient of 6mmHg between the right branch and the left branch of the pulmonary artery. The coronary arteriography showed: right coronary artery dominance without lesions; a long left anterior descending artery without stenosis; a short-sized left circumflex coronary artery in the atrioventricular sulcus occluded at the beginning, which received collateral circulation from the right and left coronary arteries; the angiocardiology showed an increase of the end-

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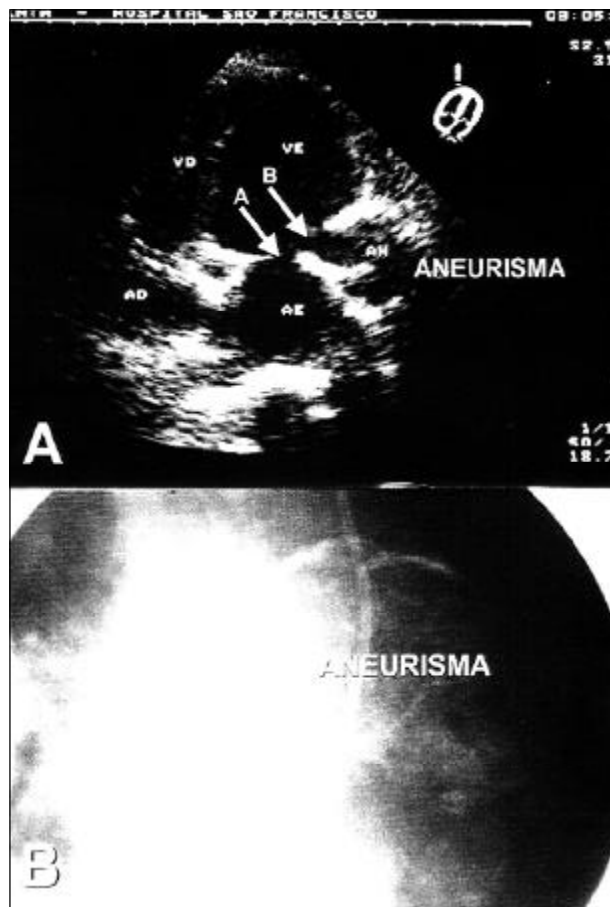


Fig. 1 – Echocardiogram showing a subvalvar mitral aneurysmatic dilatation of the basal left ventricle posterior wall (arrow A shows the mitral valve and arrow B shows the aneurysm neck); B) angiography showing radioccontrast inside the submitral calcified ventricular aneurysm.

systolic volume of the left ventricle, with diffuse hypokinesis. The mitral valve was competent, and passage of contrast from the mitral subvalvar region to a retrocardiac and calcified aneurysmatic formation could be observed (Fig. 1B). The aorta and the main pulmonary artery were normal, with a relative stenosis evident in the right branch of the pulmonary artery by extrinsic compression. After the physician-surgeon meeting, surgical treatment was opted for.

During surgery, the inspection after median sternotomy revealed a great calcified aneurysm with strong adherences in the lateral and posterior walls of the left cardiac chambers (Fig. 1A). The aneurysm was located between the left appendage and the atrioventricular sulcus, and it was compressing the right branch of the pulmonary artery against the aorta. After the cardiopulmonary bypass circuit was established, the aneurysm was dissected in all its extensions. Its calcified wall was broken, exposing its unique multilocular chamber, with a great amount of organized and recent thrombus (Fig. 2B), which had been removed (Fig. 2C). Its ostial neck was calcified and measured about 2.5 X 2.5 cm. Through it, the subvalvar apparatus of the mitral valve, which was competent, could be seen (Fig. 2D). The surgery consisted of the suture of a double patch of bovine pericardium, occluding

the aneurysm ostia, completed with additional stitches, including the left appendage. A laceration of the origin of the left descending coronary artery, which was dislocated and deformed by the aneurysm, occurred during release of the aorta. After observing that the circumflex artery was free with independent output at the injured location, a saphenous vein bypass was opted for to correct the accident. A spontaneous recovery of heart beats occurred after removal of the aortic clamp, but until the end of the surgery, important polymorphic cardiac arrhythmias occurred that were resistant to the lidocaine, amiodarone, and metoprolol treatment.

In the postoperative care unit, the great problem was control of cardiac arrhythmias, ventricular and supraventricular. Enzymatic variations, including CKMB, were not observed, nor were the electrocardiographic findings compatible with acute myocardial infarction. The attempts at associating various antiarrhythmic medications was started, but this treatment was not successful, and the patient died in the seventh day of the postoperative period, in an episode of atrioventricular dissociation, not responsive to cardiopulmonary resuscitation maneuvers. It was not possible to rule out acute myocardial infarction as the final event because an autopsy was not performed.

The anatomical pathology examination of the elements of the aneurysmectomy was constituted of multiple fragments of fibrous tissue, inelastic and gross, with areas of irregular calcifications, measuring 13.0 x 15.0 x 2.0cm. Fragments of thrombus and extensive fibrous cicatricial of the cardiac wall with foci of unspecific chronic pericarditis were found.

## Discussion

Submitral aneurysms are injuries of obscure origin, which suggests rare and varied causes, such as inflammation, infection, or traumatic illness. The infectious illnesses mentioned most often as possible causes of these types of aneurysms are syphilis and tuberculosis. Doubts do not exist about whether such things as congenital aneurysms exist. The occurrence of noninfectious and nontraumatic aneurysms supports the suggestion that mitral aneurysms subvalvar, aneurysms of the aortic Sinus of Valsalva, result from a congenital defect of the valvar ring. Submitral aneurysms only occur underneath the posterior leaflet. On the other hand, aneurysms of the Sinus of Valsalva can appear in any of the three sinuses, but subaortic aneurysms only occur below of the intermediate portion of the left aortic sinus<sup>7</sup>. Moreover, confirmation of these injuries through fetal echocardiography confirms without a doubt the congenital origin of many of these aneurysms<sup>8</sup>.

Clinically, submitral aneurysms are characterized by cardiac insufficiency, mitral insufficiency, and the absence of coronary disease associated with thromboembolic cardiomyopathy, other phenomena, and cardiac arrhythmias. Although it is not described as a coronary disease, the compression of these vases by the aneurysm can have ischemic manifestations<sup>10</sup>.

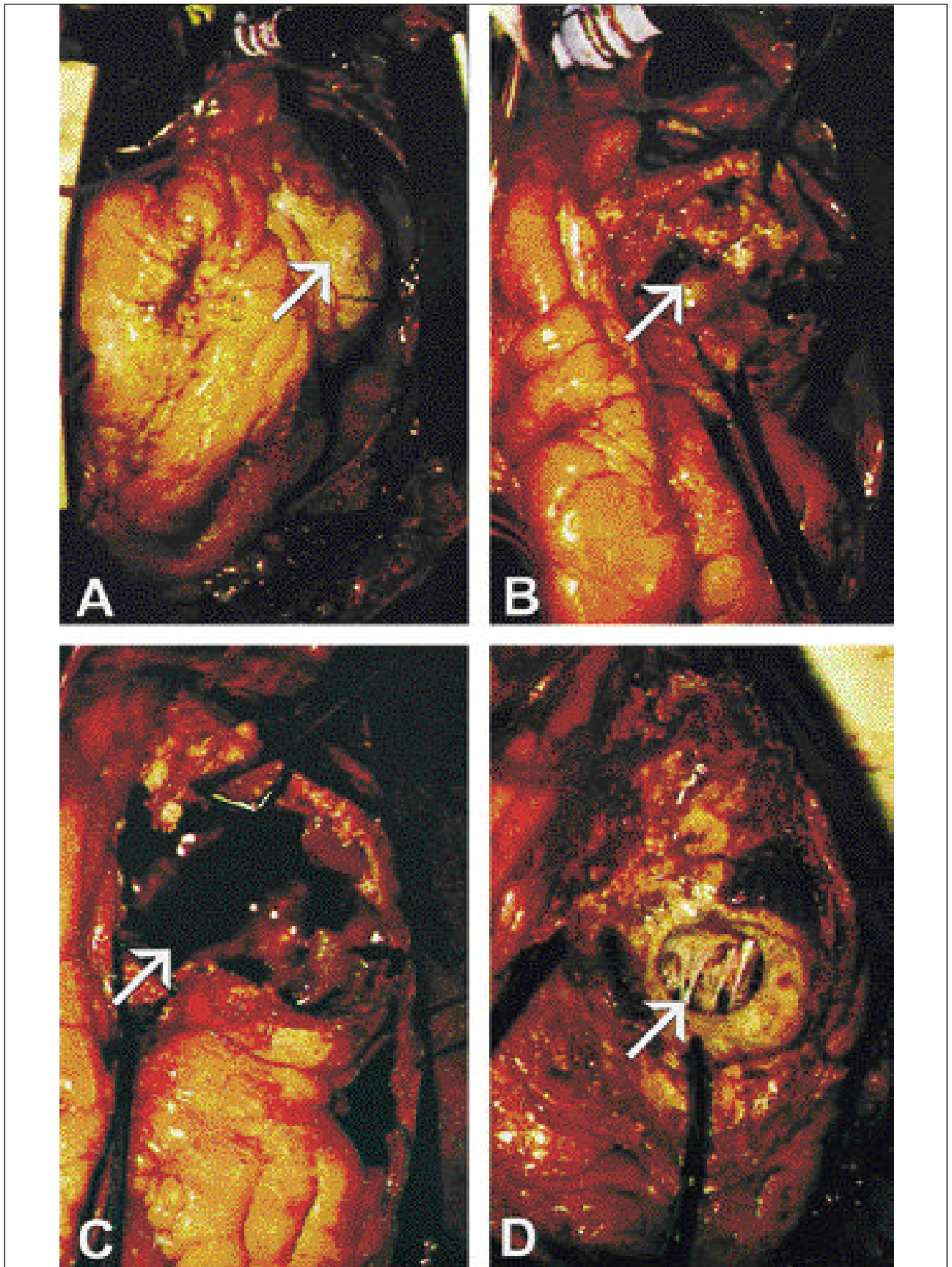


Fig 2 – Surgical aspects: A) Calcified submitral aneurysm firmly adherent to the lateral and posterior walls of the left cardiac chambers; B) aneurysmatic cavity is showing a great amount of organized and recent thrombi; C) aneurysmatic multiloculate cavity after thrombi removal; D) neck of the aneurysm measuring 2.5 x 2.5cm, also heavily calcified, observing the submitral structure.

The treatment is, eminently, surgical with the aid of cardiopulmonary bypass. The access can be extracardiac, confining the aneurysm, or using a transatrial access, exposing the orifice of the aneurysm through an incision in the floor of the left atrium (roof of the aneurysm). In the case of mitral insufficiency, this must be treated by implantation of a valvar prosthesis, but the possibility of mitral valvoplasty is considerable<sup>6,11</sup>. Surgical mortality rate is extremely important information; however, the literature does not mention a specific number. Reports have been published by some who consider the surgical treatment of submitral aneurysms to be "of high mortality and only suitable in the cases of important deterioration of the heart function"<sup>6</sup>.

In the present case it is open to question whether the diagnosis and treatment had been delayed. The patient was in functional class III of the N.Y.H.A. The aneurysm was extremely calcified, needing, literally, to be broken. About the problem of the calcification, doubt persists as to whether the aneurysms calcify over time, or whether they can present with calcification at birth. The literature makes reference to great calcified aneurysms in children below the age of 4-6 years<sup>12</sup>. The evolution of fetal medicine will verify whether calcifications can occur before the birth.

Cardiac arrhythmias were the definitive cause of the death of the patient, because cardiac insufficiency persisted completely unchanged in the postoperative period. Reports exist that cardiac arrhythmias are not frequent in the submitral ventricular cases of aneurysm, and that resection of this type of aneurysm can eliminate these arrhythmias<sup>13</sup>. These arrhythmias may have been explained by the compression of the circumflex artery. Although a great part of the aneurysm was resected, its calcified neck was contiguous with the mitral ring, and was not surgically removed (Fig. 2D). Thus, not even this aspect was controlled by the surgical act. As an autopsy was not preformed because the family refused it, it cannot be affirmed that the final morbid event was a myocardial infarction, associated or not, with the ventricular dysfunction and arrhythmia. In the event

that a severe myocardial infarction did occur, the intraoperative laceration of the left anterior descending artery could have been decisive for the fatal event. Contrary to this hypothesis, although not definitive, is the fact the death happened on the seventh day after the surgery; death would be expected to have been a more precocious event. Through a review of the literature, as broad as possible, no reports were found of lesion of the left anterior descending coronary artery. It is more probable that the event occurred with the circumflex artery. This detail deserves emphasis, together with the fact that submitral ventricular aneurysm compresses the left coronary artery including branches and even the trunk<sup>8,10</sup>.

The patient had thromboembolism in the lower limbs and underwent thromboembolism and lumbar sympathectomy. A possible cardiac embolic source in the case of the aneurysm in question was not considered. The thromboembolism can cause sudden death. Although the presence of thrombus inside the submitral aneurysm is a constant, systemic embolism is not common, possibly due to the presence of a small ostial neck in many of the cases, with only two described cases of systemic embolism in this form<sup>6</sup>. This point must be emphasized. Because of the growing incidence of embolism among nonblack populations, the use of echocardiography in patients with a history of embolic episodes in the absence of apparent risk factors for embolism is mandatory.

The hypothesis that the prevalence of this type of ventricular aneurysm is almost exclusive to African patients of the black race, because the studies had been concentrated in that continent, is a speculative matter. This could be a similar phenomenon to that occurring in relation to Chagas disease and to endomyocardial fibrosis in Brazil. This hypothesis justifies the distinction of the Brazilian cases. So, it is a strong message to medical services, especially diagnostic imaging services, pathology laboratories, and cardiology services, to pay special attention and to report new cases of this cardiac pathology.

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