Case 04/12 — A 44-Year-Old Male with Rheumatic Valvular Heart Disease with Multiple Previous Surgeries of Aortic Valve Replacement Admitted for Treatment of Congestive Heart Failure

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Male, 44 years old, with a history of rheumatic attacks in childhood and adolescence and multiple aortic valve replacement surgeries, was admitted with decompensated heart failure.

The present illness began with rheumatic attacks characterized by fever, arthritis of the right knee, when he was 7-11 old. At age 15, paroxysmal palpitations appeared. At age 16, the patient initiated follow-up at the outpatient services at InCor. Double aortic lesion and rheumatic disease activity were diagnosed.

At the time, the electrocardiogram (ECG) revealed severe left ventricular hypertrophy; a chest radiograph revealed cardiomegaly +++/ 4+.

At age 17, the palpitations became more frequent, dyspnea on moderate exertion appeared.

Physical examination in primary care (May 1977) revealed pulse rate of 80 bpm, blood pressure 100/0 mmHg; auscultation was normal, cardiac auscultation revealed systolic murmur +++/ 4+ in the aortic area and diastolic murmur in the left sternal border. There was also the presence of a third heart sound; the abdomen showed no changes, no edema, and the pulse was of rapid ascent and descent. The patient was under use of digoxin (0.25 mg daily) and prophylaxis of rheumatic fever with benzathine penicillin 1.2 million units intramuscularly (IM) every two weeks. Surgical treatment of the aortic valve was prescribed.

After the indication for surgical treatment, there were several attempts of hospitalization. However, these were frustrated by frequent episodes of fever and arthralgia of the knees treated as rheumatic disease activity and, due to the persistence of this condition, an investigation was conducted for infective endocarditis. Finally, the patient underwent aortic valve replacement with dura mater bioprostheses in 1979 (at age 19) and remained asymptomatic for five years, until 1984, when complaints similar to those prior to the surgery reappeared. Rough systolic murmur +++/4+ appeared and irradiated to the wishbone and tip.

ECG revealed left chamber overload. Radiography revealed cardiomegaly ++/4+.

An echocardiogram showed normal left ventricular dimensions (diastole 53 mm and systole 36 mm). The left atrium was 36 mm and there was dilatation of the aortic root (42 mm). There was concentric hypertrophy of the left ventricle and the prosthesis was considered to be normofunctioning.

Medication in this period was 0.25 mg digoxin, 40 mg furosemide daily and benzathine penicillin every 15 days.

In two years, the patient complained of chest pain and dyspnea initially on major exertion, then on moderate exertion. There were complaints of generalized arthralgias on several occasions.

A new evaluation revealed the following echocardiographic changes (July 1986): concentric hypertrophy of the left ventricle with septal and posterior wall thickness of 14 mm, left ventricular dilatation (66 mm) with preserved systolic function (left ventricular ejection fraction of — LVEF = 64%), aortic dilatation (44 mm), normal left atrium (36 mm) and non-coronary leaflet rupture of the aortic prosthesis with moderate heart failure.

Cardiac catheterization (August 1986) revealed right atrium 2 mmHg; right ventricle (S/Di/Df) 51/0/12 mmHg; pulmonary artery pressure (S/D/Med) 62/30/45 mmHg; pulmonary capillary wedge pressure (average) 31 mmHg; left ventricle (S/Di/Df) 138/0/37 mmHg; aorta (S/D/Med) 90/44/61 mmHg. In summary, there was right ventricular hypertension, pulmonary artery hypertension and left ventricular end diastolic hypertension. The left ventricle showed moderate diffuse hypokinesia and pronounced reflux of the aortic prosthesis.

The patient underwent a new valve replacement for bovine pericardial prosthesis (August 20, 1986). There were complications such as pericarditis and atelectasis on the left base. However, the patient was discharged without alterations, except for the presence of pericardial friction on the 13th day after the surgery.

A new echocardiographic assessment (August 28, 1989) revealed normal left ventricular dimensions (53 mm in diastole) and ejection fraction of 74%; aorta of 42 mm, left atrium of

Keywords

Rheumatic fever / complications; heart valve diseases / surgery; aortic valve / abnormalities; aortic valve / surgery.

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30 mm, right ventricle of 20 mm and septal and upper wall thickness of 18 mm. There was a mean gradient of 10 mm Hg in the aortic prosthesis, which was competent.

Five years later (October 1991), the patient was admitted with fever, general malaise and conjunctival petechiae. Heart rate was 90 bpm, blood pressure 120x 70 mmHg; the lungs were clear, there was systolic murmur ++ in the aortic area; there was no change in the examination of the abdomen and lower limbs. Infective endocarditis was diagnosed and the patient was hospitalized to receive antibiotic treatment with crystalline penicillin, oxacillin and gentamicin. There was leukocytosis (18.200/mm^3 – 25% baton-like cells; 61% segmented; 9% lymphocytes and 5% monocytes).

The electrocardiogram revealed sinus tachycardia and left ventricular overload (Figure 1).

Another echocardiogram revealed no changes except for aortic regurgitation. There was no growth of microorganisms in blood cultures. However, the patient received antibiotics for six weeks.

The patient had few symptoms for two years. Then, in 1993, he complained of dyspnea on minimal exertion.

A new evaluation by hemodynamic study (November 1st, 1993) revealed average right atrial pressure 2 mm, right ventricle (S/Di/Df) of 65/0/5 mmHg; pulmonary artery (S/D/med) 65/28/43 mmHg; pulmonary capillary pressure (S/D/med) 25/35/30 mmHg; left ventricle (S/Di/Df) 153/0/30 mmHg; aorta (S/D/Med) 115/52/74mmHg; severe aortic regurgitation; no obstructive coronary lesions, left ventricular dilatation and moderate diffuse hypokinesis.

The patient was again submitted to aortic valve replacement, now with St. Jude mechanical prosthesis (1993).

The patient remained asymptomatic and used losartan 50 mg, spironolactone 25 mg, digoxin 0.25 mg and warfarin.

About eight years after (2002), the patient had dyspnea on moderate exertion, and clinical examination and echocardiography revealed severe mitral regurgitation.

There was progression of dyspnea until minimal exertion and rest. The patient was admitted with signs of atrial fibrillation, which was cardioverted without hemodynamic improvement. He was readmitted two weeks after decompensated heart failure.

Physical examination (September 10, 2004) revealed a heart rate of 140 bpm, blood pressure of 110/80 mmHg; pulmonary auscultation revealed decreased vesicular murmur at the base and crackles until the middle third; cardiac auscultation revealed rhythmic sounds, systolic murmur in the aortic and mitral area; abdomen showed no changes and there was edema ++ of lower limbs.

The ECG (September 11, 2004) revealed atrial flutter, heart rate of 72 bpm, left bundle branch block (Figure 2). Laboratory tests (September 15, 2004) detected leukocytosis (Table 1).

Echocardiogram (October 8, 2004) revealed left ventricle (systole/diastole) 83/73 mm; septum 14 mm; posterior wall 13 mm, dilated and hypokinetic right ventricle; aortic transvalvular gradient 15 mm Hg and systolic pulmonary artery pressure of 65 mmHg. There was moderate mitral regurgitation, with thickening and decreased mobility of the posterior leaflet.

During admission, the patient improved dyspnea and edema. The mitral valve replacement surgery was not considered useful for the patient. One month later, the patient had syncope and ventricular tachycardia; he underwent electrical cardioversion and amiodarone was introduced.

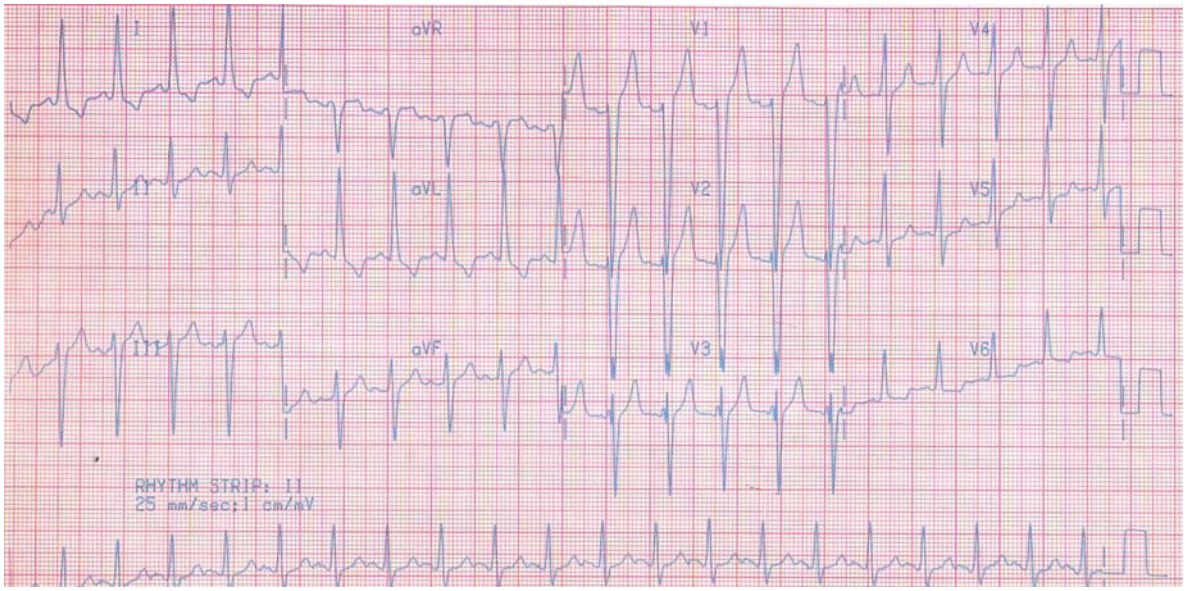

Figure 1 – *ECG: Electrocardiogram. Sinus rhythm, left ventricular hypertrophy*

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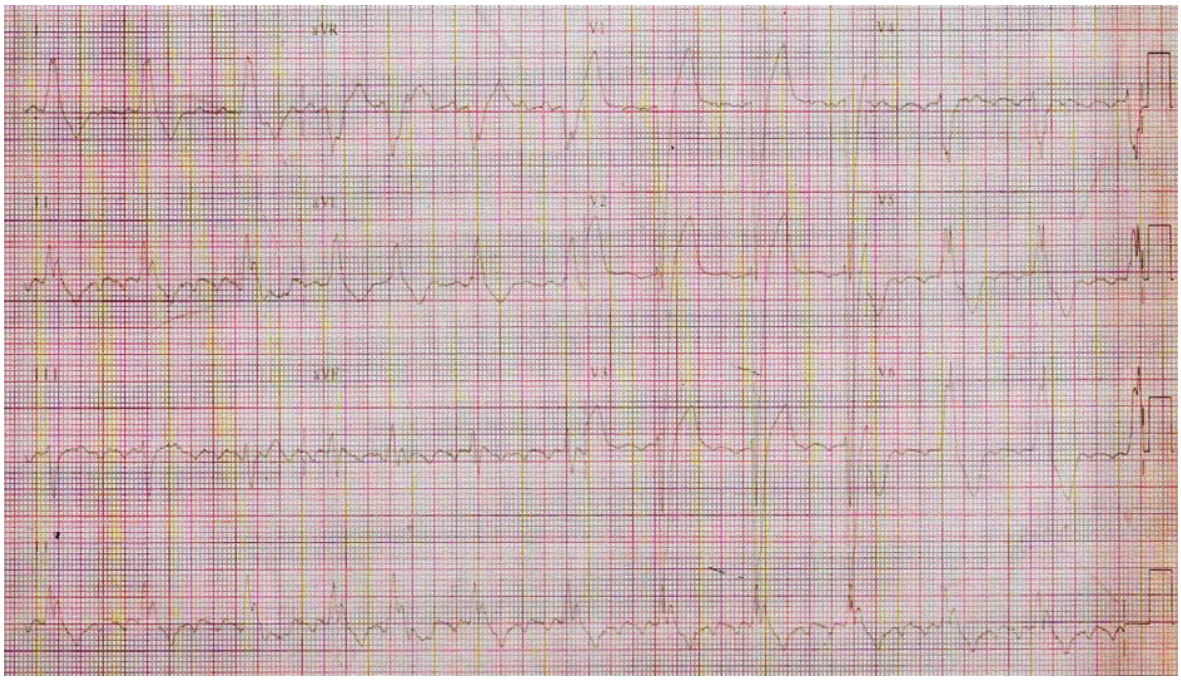

Figure 2 – *ECG: Electrocardiogram. Atrial fibrillation, left bundle branch block.*

Table 1 – Tests on admission

	September 10, 2004	October 13, 2004
Red blood cells (/mm³)	4,800,000	4,100,000
Hemoglobin (g/dL)	14.4	12
Hematocrit (%)	42	36
Leukocytes (/mm³)	14,200	14,500
Neutrophils (%)	79	85
Lymphocytes (%)	13	5
Monocytes (%)	8	10
Platelets (/ mm³)	208,000	188,000
Urea (mg/dL)	46	167
Creatinine (mg/dL)	1.0	2.9
PT (INR)	3.8	-
Potassium (mEq/L)		4.1
Sodium (mEq/L)		144
Glucose (mg/dL)		138
ALT (U/L)		27
AST (U/L)		16
Amylase (U/L)		43

Two days later, there was recurrence of tachycardia, and hemodynamic worsening after the introduction of procainamide, which was suspended and xylocaine was prescribed. The patient evolved without recurrence of arrhythmia, but there was persistent hypotension requiring orotracheal intubation for respiratory support and vasoactive amines. There was worsening of the respiratory condition with diffuse interstitial and alveolar infiltrates in the lungs. Despite the introduction of the antibiotics cefepime and vancomycin, the patient needed increasing doses of noradrenaline, acute renal failure (tests in Table 1) and developed bradycardia followed by asystole, which did not respond to resuscitation maneuvers and died (3:30 p.m., October 14, 2004).

Clinical aspects

This case deals with a young man aged 44 with a history of various rheumatic attacks since the age of 7, evolving lifelong with various secondary valve replacements and complications of the disease (carditis/valvulitis), and, at the end, presents mitral regurgitation, new decompensated heart failure (HF), complex ventricular arrhythmia, cardiogenic shock and death.

Firstly, it emphasizes the onset of symptoms, where, around age 7, the patient had repeated fever and monoarthritis of the right knee, which persisted until the age of 11. At this stage, with these clinical data alone, we cannot confirm the diagnosis of rheumatic fever, as there are no other signs and symptoms that lead to diagnosis, and we do not know exactly what this arthritis was like (was it really addictive and asymmetric

polyarthritis of the large joints?). Again, the currently known Jones criteria for the diagnosis of rheumatic fever are carditis, arthritis and Sydenham chorea, eritema marginatum and subcutaneous nodules (major criteria) and fever, arthralgia, increased inflammatory markers and prolonged PR interval (minor criteria), which are shown in Tables 2 and 3[1,2].

In view of these criteria, the patient was actually diagnosed with rheumatic fever at age 16, when double aortic lesion and rheumatic disease activity were diagnosed, since at this time he has two major criteria and one minor criterion (carditis, large joint arthritis and fever) [3,4].

During the course of the disease, at age 17, the patient had frequent palpitations (already had such symptoms at age 15) and dyspnea on minimal exertion. The presence of paroxysmal palpitations is not common when there is only aortic lesion; it is more common in cases of mitral-aortic valve involvement or mitral involvement. However, at this time, the clinical examination is consistent with double lesion aortic valve involvement with predominant failure. This case does not present a better description of systolic murmur audible in the aortic area. On physical examination, there was inconsistency in blood pressure 120/00 mmHg, corresponding to a significant widening of pulse pressure, and rapid pulse rise and descent (Corrigan or "water hammer"), which is highly suggestive of aortic regurgitation. Only after two years, the patient underwent aortic valve replacement, which was delayed due to complications consistent with rheumatic attacks and a potential endocarditis apparently ruled out[5]. After surgery, the patient remained asymptomatic.

After five years, at age 24, the patient again presents complaints similar to those prior to the surgery. The only description of the physical examination reveals a rough systolic murmur +++/4+ irradiated to the wishbone and tip, probably better heard in the aortic area. That information, in addition to others, such as the murmur characteristics, is not present in the description. The ECG remains without any new changes with respect to the previous one, just like the chest radiography (left ventricular hypertrophy – EVS — and cardiomegaly, respectively).

At this time, with the clinical data presented, stenosis of aortic valve prosthesis is suggested as a diagnosis — consistent clinical examination, young male patient with five years of implantation. However, he underwent an echocardiogram (ECHO), which ruled out prosthesis dysfunction, but revealed

Table 2 – Modified Jones criteria for a diagnosis of rheumatic fever (1992)

Major criteria	Minor criteria
Carditis	Fever
Arthritis	Arthralgia
Sydenham's chorea	Elevation of acute phase reactants (ESR, CRA)
Erythema marginatum	Prolonged PR interval on ECG
Subcutaneous nodules	

Evidence of infection with group A streptococcus by throat culture, rapid test for GABHS and high antibody titers (ASLO); Adapted from Dajani et al. Jones criteria 1992 Update – AHA22.

Table 3 – World Health Organization criteria (2004) for the diagnosis of the first onset, recurrence and chronic rheumatic heart disease (based on the modified Jones criteria)

Diagnostic categories	Criteria
Onset of rheumatic fever.*	Two major criteria or one major and two minor criteria plus evidence of previous streptococcal infection.
Recurrence of rheumatic fever in patients without rheumatic heart disease established.†	Two major criteria or one major and two minor criteria plus evidence of previous streptococcal infection.
Recurrence of rheumatic fever in patients without rheumatic heart disease established.†	Two minor criteria evidence of previous streptococcal infection.‡
Sydenham's chorea. Rheumatic carditis of insidious onset.†	The presence of another major manifestation or evidence of previous streptococcal infection is not required.
Chronic valvular lesions of CRC: initial diagnosis of pure mitral stenosis or mixed mitral lesion and/or aortic valve disease with characteristics of rheumatic involvement.§	There is no need for additional criteria for the diagnosis of CRC.

**Patients may have only polyarthritis or monoarthritis + three or more minor signs + evidence of previous streptococcal infection. These cases should be considered as "probable rheumatic fever" and instructed to perform secondary prophylaxis undergoing regular cardiac evaluations; †Infective endocarditis should be excluded; ‡Some patients with recurrences do not meet these criteria; §Congenital heart disease should be excluded; Source: WHO 2004 or Adapted from the WHO Technical Report Series 923, Rheumatic Fever and Rheumatic Heart Disease, Geneva, 2004.*

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normal left ventricular dimensions (53X36 mm), left atrium of normal size (36 mm) and concentric hypertrophy of the left ventricle (LV), which are consistent with valvulopathy of aortic stenosis. We assume that, in this era (1980s), M-mode echocardiography was mostly used perhaps due to the little amount of Doppler devices (continuous), which may explain the lack of aortic transvalvular gradient measurement, which led to the probable misdiagnosis of normal structure of the prosthesis at that time, corroborating the clinical and echocardiographic dissociation.

After two years, the patient presents chest pain and dyspnea on major exertion. A new ECHO showed concentric left ventricular hypertrophy (septum and posterior wall of the LV = 14 mm), LV dilatation (66 mm) and preserved ejection fraction (EF), in addition to normal left atrium (LA); moreover, the non-coronary leaflet of the aortic prosthesis had ruptured, causing moderate aortic regurgitation. A cardiac catheterization performed at the same time showed normal right atrial filling pressure (RA); high pulmonary arterial pressure concomitant with high pulmonary capillary wedge pressure; high left ventricular filling pressure (Pd2) and LV with moderate diffused hipokynesia, and severe valve prosthesis reflux. In this examination, middle pulmonary artery pressure, average pulmonary capillary wedge pressure and mean LV end-diastolic pressure are similar (30, 31 and 37 mmHg), reflecting pulmonary hypertension (PH) secondary to high hydrostatic pressure (originating from the left chambers with good response to surgery) and non-fixed PH secondary to pulmonary arterial remodeling. Also in this examination, the transvalvular gradient is high (LV Systolic Pressure of 138 mmHg minus systolic pressure of the aorta of 90 mmHg, which corresponds to 48 mmHg), which confirms the diagnosis of prosthesis stenosis.

Operated again, presumably asymptomatic after the procedure, the patient performed a new ECHO, which showed left ventricular dilatation improvement, preservation of EF, septal thickness and LV posterior wall (LVPW) still high (18 mm) and aortic prosthesis gradient of 10 mm Hg (considered normal up to 15 mmHg in bioprosthesis); sufficient prosthesis.

At age 31, the patient demonstrates poor general condition, fever and conjunctival petechiae. There were no positive blood cultures at the time. He was treated with antibiotics for infective endocarditis for six weeks, supposedly successfully.

There is evidence that the patient had infection, such as fever, leukocytosis on complete blood count and tachycardia. On physical examination, in addition to conjunctival petechiae, there was systolic murmur without other descriptions, which can be consistent with the presence of the prosthesis only. However, a new ECHO revealed no changes, except for central aortic regurgitation, which can be found in patients with prosthetic endocarditis, which differs from the common pattern of paravalvular flow found in the prosthesis and tends to be benign and with no progression. Yet, the patient's diagnosis of endocarditis is probable, according to the Duke criteria, as he has one major criterion and three minor criteria (valvopathy, fever and embolic phenomenon — conjunctival petechiae) [6,7]. If the diagnosis of prosthetic endocarditis was confirmed, perhaps in this case the indication for surgical treatment would not be appropriate, since the onset occurred late (after 12 months); there were no signs of refractory HF class III/IV NYHA, or severe prosthesis dysfunction with perivalvular extension of the infection with abscesses, fistulas, mechanical obstruction of the prosthesis by vegetation or rupture, no clinical criteria for treatment failure, such as persistent fever and infection after 10-14 days despite antibiotic therapy, recurrent embolization in the presence of antimicrobial treatment for more than seven days, patient responded well to treatment and there was no isolation of germs known to be resistant or virulent with staphylococcus (*S.lugdunensis*), *Brucella spp*, *Coxiella burnetii* and fungi (*Candida spp and Aspergillus spp*). We do not rule out endocarditis by HACEK etiologic agents (*Haemophilus spp*, *Actinobacillus actinomycetemcomitans*, *Cardiobacterium hominis*, *Eikenella corrodens* and *Kingella kingae*) [8,9].

At age 33, dyspnea on minimal exertion reappeared. We chose a new invasive hemodynamic evaluation, which revealed, once again, normal RA pressure, increased right ventricular pressure (RV), pulmonary artery, pulmonary capillary wedge pressure and left ventricular pressure with dilated left ventricle, moderate diffuse hypokinesis without obstructive coronary artery disease. In this study, the pulmonary artery, pulmonary capillary and PD2 pressures are similar; the prosthesis gradient is high (38 mmHg), and systolic and diastolic aortic pressure present a divergent character, but with a smaller pulse pressure widening, probably due to severe left ventricular dysfunction. Again, aortic prosthesis dysfunction, stenosis and prosthesis failure are diagnosed.

A new replacement was performed at this time and this time for a mechanical prosthesis, which is better in cases like this, where the patient presents several valve replacements. Thereafter, he remained asymptomatic. According to the Brazilian Guidelines of Valvopathy in 2011, patients aged under 65 can opt for mechanical prosthesis, provided that there is no contraindication to anticoagulation. The patient can choose a bioprosthesis if he chooses a lifestyle without the use of anticoagulants (Class IIa)[10].

At age 41 (in 2002), new symptoms of dyspnea on exertion and a new finding: physical examination (PE) and ECHO consistent with moderate mitral regurgitation (MR). Right after, the patient presented cardiac decompensation and atrial arrhythmia (flutter), cardioverted without hemodynamic improvement. Then, the patient was readmitted for decompensated HF.

In 2004, at age 43, he was again hospitalized for decompensated HF with cardiomyopathy. In this admission, in addition to FE consistent with hypervolemia, the patient presented ECHO with new changes – atrial flutter and left bundle branch block — (consistent with involvement of the conduction system secondary to ventricular dilatation).

The ECHO of this admission showed marked dilatation of the left ventricle (83/73 mm), Septum 14, LVPW 13, dilated and hypokinetic RV, transvalvular gradient of 15 mmHg and PASP 65 mmHg, with moderate mitral regurgitation, mitral valve thickening and decreased mobility of the posterior leaflet.

Apparently, in this phase, the patient developed HF secondary to MI probably primary, since the condition began preceded by the diagnosis of MI and the 2004 ECHO reveals structural valve changes probably existing in 2002, information not provided by the clinical case. Possibly the etiology of such involvement is by extemporaneous development of rheumatic fever, with late involvement of this valve. Among other possible differential diagnoses, we have ischemic mitral regurgitation, but the patient had no history of coronary artery disease and no coronary angiography mentioned coronary lesions, despite being within the age range of prevalence. What draws our attention is that after the last valve replacement, at age 33, the patient apparently did not receive prophylaxis for the disease, which should be done until at least 40 years according to the guidelines[10].

When diagnosed with mitral regurgitation associated with atrial symptoms and arrhythmias, the best approach could be the indication of surgical correction of mitral valve two years earlier (in 2002) when the presence of atrial fibrillation (AF) of recent onset, severe mitral regurgitation and the patient being in CF II/III, were criteria for surgery (indication: Class IIa, according to the Brazilian Guidelines), which did not occur. Consequently, the patient developed severe valvular heart disease in which the severe ventricular dilatation, severe ventricular dysfunction (estimated at 22% by the data presented) and signs of right HF contraindicated surgical correction in subsequent years, since when there is left ventricular dilatation greater than 55 mm and LV dysfunction with EF smaller than 30% and the impossibility of preserving the subvalvular apparatus, surgical procedure is not indicated[10].

After one month of medical treatment, in which the patient had improvement of symptoms of heart failure, he presented an episode of syncope due to ventricular tachycardia (VT) corrected after electrical cardioversion and introduction of amiodarone – a result of severe cardiomyopathy with significant ventricular dysfunction, abnormal myocardial architecture, fibrosis and degeneration of the conduction system. Two days later, a new episode of VT with hemodynamic instability unresponsive to measures led to apparently cardiogenic shock and death despite all the intensive support employed (Dr. Danilo Guercio Fernandes, Dr. Luiz Mário Baptista Martinelli).

Diagnosis hypothesis: heart failure and cardiogenic shock due to rheumatic heart disease with multiple valve replacements for aortic valve disease treatment.

Necropsy

The heart showed large increase in volume, weighing 980 g. There were adhesions between the pericardial leaflets, probably due to previous surgeries. The aortic valve prosthesis (mechanical) was well positioned without thrombi, vegetations or valvular pannus on both sides (arterial and ventricular). In the topography of the right coronary sinus of Valsalva, there was saccular aortic root dilatation of 1.5 cm in diameter, devoid of content (Figure 3). The mitral valve presented moderate annulus dilatation and slight thickening of the cusps with slight commissural fusion. The chordae were delicate and the subvalvular apparatus was preserved (Figure 4). The ventricles were very dilated and presented hypertrophic walls. Endocardial thickening was found at the left ventricular outflow tract (Figure 5). The atria also showed dilatation and

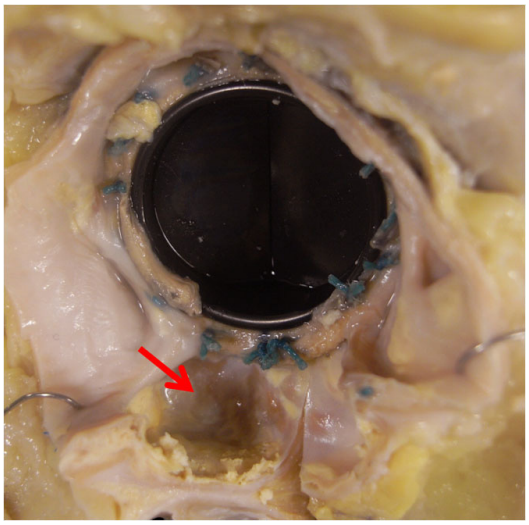

Figure 3 – *Detail of a preserved aortic valve prosthesis and aortic root. Small aneurysm without content inside (arrow).*

hypertrophy of the walls. There was no cavitary thrombi. Histologically, the myocardium showed signs of hypertrophy with extensive interstitial fibrosis without cardiomyocyte disarray (Figure 6). The mitral valve was thickened by fibrosis without inflammatory infiltrate, with thick newly formed vessels (Figure 7). Together, the lungs weighed 1900 g, were armed and presented increased consistency.

Microscopic analysis showed diffuse alveolar damage characterized by the presence of alveolar edema and hyaline membranes. There were also signs of chronic passive congestion with hypertrophy of the medial layer of the pre and intra-acinar arteries and veins, a morphological sign of passive

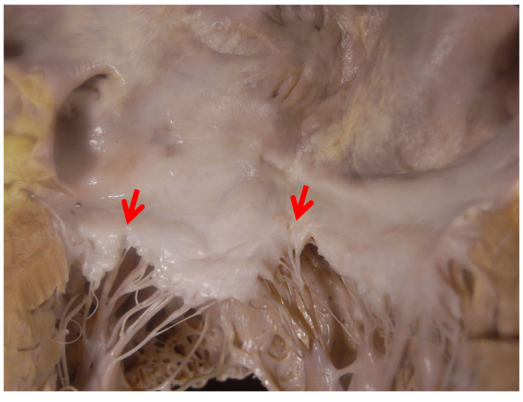

Figure 4 – *Dilated left atrium and mitral valve showing mild commissural thickenning and delicate chordae.*

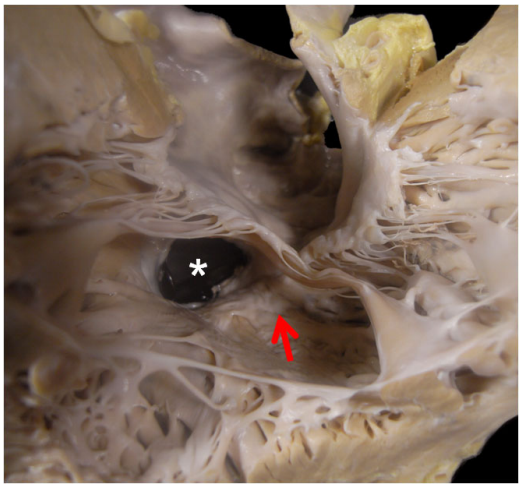

Figure 5 – *View of the left ventricular cavity toward the outflow tract, whith endocardial septal thickening (arrow). The asterisk shows the ventricular aspect of the aortic prosthesis.*

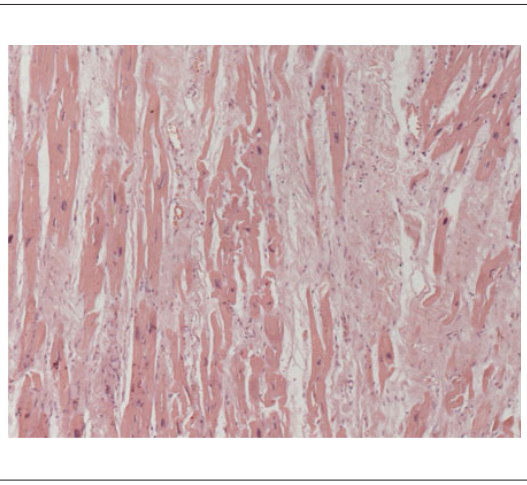

Figure 6 – *Photomicrograph of the ventricular myocardium showing severe interstitial fibrosis and hypertrophy of cardiomyocytes. Hematoxylin-eosin staining, objective lens magnification = 10X.*

pulmonary hypertension. We also found signs of low systemic output, such as acute renal tubular necrosis and hepatic centrolobular sinusoidal dilatation. There was no evidence of active infection in the various organs analyzed. The gallbladder had multiple mixed calculi inside, but there were no signs of acute cholecystitis (Dra. Vera Demarchi Aiello).

Clinicopathological diagnoses: well-preserved aortic valve prosthesis; saccular aneurysm of the aortic root (1.5 cm in diameter), without content; chronic mitral valve disease; acute interstitial fibrosis and cardiomyocyte hypertrophy; systemic signs of shock with pulmonary alveolar damage in the hyaline membrane phase (cause of death) (Dra. Vera Demarchi Aiello).

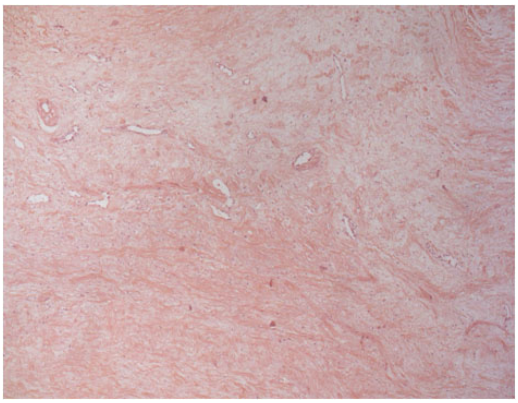

Figure 7 – *Photomicrograph showing mitral valve fibrosis, newly formed vessels without significant inflammation. Hematoxylin-eosin staining, objective lens magnification = 5X.*

Comments

Although there is strong clinical evidence that the patient had rheumatic disease, the clinicopathological examination was not able to definitively confirm this diagnosis. In particular, it would be essential to have assessed the native aortic valve, but the patient was operated in another institution and we have not had access to this material. We could only analyze the mitral valve, as described above, which showed fibrosis, newly formed vessels, without significant inflammation. In addition, its macroscopic appearance was not typical of rheumatic disease, since commissural fusion was very mild and the both the chordae and subvalvular apparatus were preserved, which usually does not usually happen in this disease, particularly of very early onset in childhood, as it is the case of the patient discussed. We could not, however, definitively rule out the possibility of an a typical morphological presentation of rheumatic disease, but it we hypothesize that the aortic valve disease had another etiology (congenital, for example), and that the mitral involvement has been late and secondary to the annulus dilatation. The myocardium showed severe hypertrophy and fibrosis consistent with several episodes of decompensation of heart failure. Importantly, one should not speak of "cardiomyopathy" in this case and in others with these characteristics, since the use of this term should be reserved to cases of primary myocardial impairment without any valvular disease associated. When, on the other hand, the valve impairment is disproportionately mild to explain myocardial dysfunction, some authors have suggested using the term "valvular cardiomyopathy"11. The lungs, in particular, showed signs of passive congestion of long evolution with evidence of secondary vascular disease. This passive pulmonary hypertension condition is also present in cardiac decompensation of other etiologies and can affect both indication and outcome after cardiac transplantation[12,13].

References

1. Guidelines for the diagnosis of rheumatic fever. Jones Criteria, 1992 update. Special Writing Group of the Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young of the American Heart Association. JAMA. 1992;268(15):2069-73.
2. Barbosa PJ, Müller RE, Latado AL, Achutti AC, Ramos AI, Weksler C, et al.; Sociedade Brasileira de Cardiologia / Sociedade Brasileira de Pediatria / Sociedade Brasileira de Reumatologia. Diretrizes brasileiras para o diagnóstico, tratamento e prevenção da febre reumática. Arq Bras Cardiol. 2009;93(3 supl.4):1-18.
3. World Health Organization (WHO). Rheumatic fever and rheumatic heart disease: report of a WHO expert Consultation. 29 Oct 29. Geneva (Switzerland); 2001. (WHO Technical Report Series 923).
4. Carapetis JR, McDonald M, J Wilson NJ. Acute rheumatic fever. Lancet. 2005;366(9480):155-68.
5. Raju BS, Turi ZG. Rheumatic fever. In: Bonow O, Mann DL, Zipes DP, Libby P. Braunwald's heart disease: a textbook of cardiovascular medicine. 9th edition. Philadelphia: Elsevier-Saunders; 2012. p. 1869-74.
6. Sexton DJ, Otto CM, L Baron EL. Diagnostic approach to infective endocarditis. © 2012 UpToDate, Inc. Mar 19, 2012.
7. Li JS, Sexton DJ, Mick N, Nettles R, Fowler VG Jr, Ryan T, et al. Proposed modifications to the Duke criteria for the diagnosis of infective endocarditis. Clin Infect Dis. 2000;30(4):633-8.
8. Durack DT, Lukes AS, Bright DK. New criteria for diagnosis of infective endocarditis: utilization of specific echocardiographic findings. Duke Endocarditis Service. Am J Med. 1994;96(3):200-9.
9. Karchmer AW. Infective endocarditis. In: Bonow O, Mann DL, Zipes DP, Libby P. Braunwald's heart disease: a textbook of cardiovascular medicine. 9th edition. Philadelphia: Elsevier-Saunders; 2012. p. 1549-57.
10. Tarasoutchi F, Montera MW, Barbosa MR, Piñero DJ, Sanchez CR, Barbosa MM, et al. Diretriz brasileira de valvopatias - SBC 2011 / I Diretriz Interamericana de Valvopatias – SIAC 2011. Arq Bras Cardiol. 2011;97(5 supl.1):1-67.
11. Thiene G, Basso C, Calabrese F, Angelini A, Valente M. Twenty years of progress and beckoning frontiers in cardiovascular pathology: cardiomyopathies. Cardiovasc Pathol. 2005;14(4):165-9.
12. Guazzi M, Arena R. Pulmonary hypertension with left-sided heart disease, Nat Rev Cardiol. 2010;7(11):648-59.
13. Delgado JF, Gómez-Sánchez MA, Sáenz de la Calzada C, Sánchez V, Escribano P, Hernández-Afonso J, et al. Impact of mild pulmonary hypertension on mortality and pulmonary artery pressure profile after heart transplantation. J Heart Lung Transplant. 2001;20(9):942-8.