

Several diagnoses in a patient with left atrial myxoma: Apropos of a case

Rodolfo Vega Candelario¹✉, MD; Junior Vega Jiménez², MD; Roger Mirabal Rodríguez³, MD, MSc; and Luis A. Rodríguez Sánchez⁴, MD, PhD

¹ Department of Cardiology, Hospital General Docente Roberto Rodríguez Fernández. Morón, Ciego de Ávila, Cuba.

² Department of Internal Medicine, Hospital Militar Docente Dr. Mario Muñoz Monroy. Matanzas, Cuba.

³ Department of Cardiovascular Surgery, Cardiocentro Ernesto Che Guevara. Santa Clara, Villa Clara, Cuba.

⁴ Department of Intensive Medicine. Hospital General Docente Roberto Rodríguez Fernández. Morón, Ciego de Ávila, Cuba.

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CM: cardiac myxomas

CT: cardiac tumors

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✉ R Vega Candelario
Edif. 10, Apto. 11.
Microdistrito Norte.
Morón, Ciego de Ávila, Cuba.
E-mail address:
rvc_50@hgm.cav.sld.cu

ABSTRACT

Primary cardiac tumors are rare diseases in all age groups and account for 0.2% of all human neoplasms. Cardiac myxomas have an annual incidence of 0.5 per million people. We present the case of a 60-year-old man with a personal pathological history of ischemic heart disease and heart failure, diagnosed two years ago, when he began to have repeated episodes of palpitations, angina and sudden dyspnea, especially when getting up from bed. The patient presented symptoms and signs of intermittent mitral stenosis, heart rhythm disorders (blocked atrial fibrillation) and episodes of acute coronary syndrome. The echocardiogram showed the presence of a pedunculated left atrial myxoma, which was immediately referred to the Cardiovascular Surgery Department for surgical treatment.

Key words: Myxoma, Neoplasms, Cardiac tumours, Diagnosis, Treatment

Varios diagnósticos en un paciente con mixoma auricular izquierdo: A propósito de un caso

RESUMEN

Los tumores cardíacos primarios son enfermedades raras en todos los grupos de edad y representan el 0,2% del total de las neoplasias humanas. Los mixomas cardíacos tienen una incidencia anual de 0,5 por cada millón de personas. Se presenta el caso de un hombre de 60 años de edad, con antecedentes patológicos personales de cardiopatía isquémica e insuficiencia cardíaca diagnosticados desde hace dos años, momento en que comienza a presentar reiterados episodios de palpitaciones, angina de pecho y disnea súbita, sobre todo al reincorporarse del lecho. El paciente presentó síntomas y signos de estenosis mitral intermitente, trastornos del ritmo cardíaco (fibrilación auricular bloqueada) y episodios de síndrome coronario agudo. El ecocardiograma mostró la presencia de un mixoma auricular izquierdo pediculado, por lo que fue remitido de inmediato al Servicio de Cirugía Cardiovascular para su tratamiento quirúrgico.

Palabras clave: Mixoma, Neoplasias, Tumores cardíacos, Diagnóstico, Tratamiento

INTRODUCTION

Primary cardiac tumors (CT) are rare diseases in all age groups and represent 0.2% of all human neoplasms. In the large series of autopsies, they have a frequency of 0.001-0.03%. They can be primary or metastatic (secondary) and its secondary affectionation is 20 to 40 times more frequent than the primary^{1,4}.

Around 75% of these CT are benign neoplasms. Cardiac myxomas (CM) represent 30-50% of all myxomas and they have an annual incidence of 0.5 per million people. Most of the times they appear in adults aged between 30 to 50 years old, but they can turn up in practically any age group from neonates to long-lived^{1,3,5}.

A 65% of CM take place in women and about 90% are sporadic, the rest is family with autosomal dominant transmission. This variety often occurs as part of a syndromic complex (complex or Carney's syndrome) that includes: a) myxomas (cardiac, cutaneous, mammary, or both); b) efelid, pigmented nevi or both; c) endocrine hyperactivity (primary nodular adrenal cortical disease, with or without Cushing's syndrome, Sertoli cell tumors, pituitary adenomas with gigantism or acromegaly, and thyroid tumors)^{1,6}.

Family forms can be associated with other diseases, forming the "myxomatous complex" (skin myxomas, endocrine tumors and lentiginosis). Some group of findings, currently included in the Carney's syndrome, are called NAME syndrome (nevi, atrial myxoma, myxoid neurofibromas, and efelids) or LAMB syndrome (lentigos, atrial myxoma and blue nevi)^{4,6}.

The CM derived from mesenchymal multipotential cells of the subendocardium and both, the ones that take place in the Carney's syndrome and sporadic cases, are indistinguishable histologically^{1,4,6}. They are movable tumors (pedunculated) or sessile, of variable size (1-15 cm), with more usual location in the left atrium (83%), but can also be in the right (12.7%), and be biatrial (1.3%) or ventricular (1.7% in the left and 0.6% in the right). Generally, they can appear in all cardiac chambers^{4,7}.

The myxoma is the most common benign cardiac tumor, followed in order of frequency by the pericardial cyst, lipoma, papillary fibroelastoma and rhabdomyoma (in children); in the fetus, there is a greater proportion of germ cell tumors^{1,5,8}.

Patients with symptomatic CM have a wide variety of nonspecific symptoms that depend on the size, location and mobility of the tumor. Their diagnoses,

like that of the rest of the CT, represent a medical challenge, due to the different forms of clinical presentation that explain the delay in the diagnosis. They may have serious impact on the deterioration of the cardiac structure and function. That is why, the key for the early diagnosing of a CT is always to raise it as a differential diagnosis^{4,9}.

CASE REPORT

A 60-year-old white male, with history of ischemic heart disease and heart failure diagnosed for three years, with treatment of antiplatelet drugs, beta blockers, nitrates and diuretics, came to the hospital referring "lack of air and pain in the chest".

From the date of these diagnoses, the patient has had repeated episodes of intense paroxysmal dyspnea whose semiological cardinal element is the exacerbation when going from the decubitus position to sitting or standing, accompanied by intense and continued cough for several minutes. In addition, he referred irregularly irregular palpitations, when they were intense, and blackouts or loss of consciousness, generalized seizures and stertorous breathing, what was interpreted as cardiac syncope in the form of Stock-Adams crisis or possible recurrent strokes by microemboli.

He had also chest pain that radiated to the left arm and most frequently at rest, with the need to use sublingual nitroglycerin, a situation that was frequent, lasting and intense with time; thus, it was interpreted on several occasions as an acute coronary syndrome and was the cause of several hospital admissions. In the last one, the patient was admitted with anasarca, dermal lesions that were interpreted as vasculitis, changes in the skin coloration of the lower limbs, fever of 38°C, inflammation in several joints that made walking difficult, intense intermittent pain in the extremities, blurred vision and diplopia; in addition to dyspnea, orthopnea and the cough already described, all of which made him constantly complain.

Physical examination

The patient had the mucous membrane wet with normal color, generalized edema compatible with anasarca, fever, uncontrollable state of anxiety and nervousness, with the appearance of exhaustion and

frustration; polypnea of 18-20 breaths per minute (usually), with normal pulmonary auscultation alternating with presence of wet rattles; arrhythmic cardiac noises, first variable loud noise with mitral diastolic murmur II/VI, long and intermittent, changing intensity, length and tone with the position differences of the patient and the moment of auscultation, heart rate and blood pressure usually normal.

At the time of the clinical presyncopal setting or syncope, the arrhythmic arterial pulses decreased in intensity and extent to be filiform and practically imperceptible, and paleness, diaphoresis, discrete acrocyanosis, jugular engorgement, pupillary dilation and distal thermal gradient appeared. He showed occasionally atrial fibrillation with rapid ventricular response, despite the use of beta blockers.

Table. Complementary blood tests.

Complementary	Result	Reference values
Hemoglobin	13.0 g/L	11.0 – 15.0 g/L
Hematocrit	0.43	0.39 – 0.52
Leukogram	$8.0 \times 10^9/L$	$4.5 - 10.5 \times 10^9/L$
Erythro sedimentation	10 mm/h	< 20 mm/h
TGP	36 U/L	0 – 49 U/L
TGO	40 U/L	0 – 40 U/L
GGT	39 U/L	5 – 45 U/L
Alkaline phosphatase	180 U/L	100 – 290 U/L
Cholesterol	6.2 mmol/l	3.87 – 6.30 mmol/L
Triglycerides	1.9 mmol/L	0.46 – 1.88 mmol/L
Creatinine	85 $\mu\text{mol/L}$	47.6 – 113.4 $\mu\text{mol/L}$
Urea	5.0 mmol/L	1.80 – 6.10 mmol/L
Uric acid	349 $\mu\text{mol/L}$	155 – 428 $\mu\text{mol/L}$
Fasting glycaemia	6.2 mmol/L	3.5 – 6.5 mmol/L
Cardiac biomarkers	No alterations	
Serology (VDRL)	Negative	
HVI	Negative	
Surface Antigen	Doubtful (repeated: negative)	

Performed exams

- Complementary blood tests: Described in the **table**.
- Telecardiogram (**Figure 1**): Enlarged cardiac silhouette (cardiomegaly), without pleuropulmonary lesions.
- Electrocardiogram (**Figure 2**): Atrial fibrillation with appropriate ventricular response, QRS axis to the right ($> 90^\circ$), isodiphasic complexes in D_1 and disorders of the ventricular repolarization. In addition, signs of right bundle branch block of the His bundle (rsR' of V_1-V_4).
- Transthoracic echocardiogram (**Figure 3**): It was key to establish the diagnosis. It clearly shows the pedunculated LAM inserted in the angle between the interatrial septum and the posterior wall of the left atrium, with high mobility, which occupies much of the atrial cavity and enters in diastole in the mitral valve, with the consequent interference of the ventricular filling, what produces symptoms and intermittent signs of severe



Figure 1. Telecardiogram showing an enlarged cardiac silhouette (cardiomegaly), without pleuropulmonary lesions.



Figure 2. Electrocardiogram showing atrial fibrillation with right bundle branch block and signs of myocardial ischemia.

mitral stenosis.

Based on the polymorphism of the symptoms, the patient was considered in critical condition and an adequate interprofessional and interinstitutional dynamics was achieved, adequate for a case of this kind, with a late diagnosis and poor prognosis, and he was moved from the emergency department to the center of reference (Cardiocentro Ernesto Che Guevara, Santa Clara, Cuba) for its surgical treatment. The histological diagnosis confirmed the clinical suspicion: LAM. The patient evolved favorably, the symptoms and atrial fibrillation disappeared, and he is monitored by outpatient cardiology consulting.

COMMENT

The CT can be asymptomatic or cause a diverse and varied symptomatology. The CM is called the “great simulator”. From the clinical perspective, patients with this type of tumor may have different unspecific findings that often create a lot of confusion and hinder the diagnosis, by simulating an immune disease or presenting any cardiac symptoms^{1,3,4}.

About half of the patients present a clinical setting that simulates a mitral stenosis, and in more than 30%, the LAM causes embolic phenomena; however, most patients will consult with at least one of the elements of the classic triad of cardiac obstructive signs, embolic and constitutional or systemic^{1,2,4}. It can also look like a severe mitral valve failure, as the case published by Manzur *et al.*⁷ of a 31-year-old patient.

The cardiac obstructive findings are due to the

mechanical interference of the tumor with the mitral valve and they are common initial findings of the symptomatic triad. The LAM can cause heart failure by occupation of the tumor mass or the tumor encavating phenomenon in the mitral valve, which can lead to dizziness, paroxysmal dyspnea, cough, lung edema, syncope or sudden death. In addition, they can embolize any arterial territory^{1,4}.

In patients with LAM, the auscultation reveals a strong first noise and a second accentuated, followed by an early diastolic tone. This noise, known as “tumor plop”, is produced by the tumor prolapse through the mitral valve^{2,4}.

This CT should also be considered in all patients in whom mitral stenosis is suspected, if there is no history of rheumatic fever or heart murmur, if the breath occurs intermittently or it is only perceived in certain positions, like if there are embolic phenomena in patients with sinus rhythm⁴.

Moreno-Ariño *et al.*¹⁰ describe a case of LAM that simulated a systemic vasculitis. Other forms of presentation have been described, such as: pulmonary thromboembolism¹¹, cardioembolic stroke¹², and obstruction of the ophthalmic artery¹³. On the other hand, Antolín *et al.*¹⁴ show a female patient with LAM that caused an acute renal failure by rhabdomyolysis, secondary to lower limb ischemia after the embolization of a tumor fragment.

Al-Fakhouri *et al.*¹⁵ and Imbalzano *et al.*¹⁶ have respective cases of patients with LAM as a cause of acute coronary syndrome, what coincides with our case; like the patient of Navarro and Fernández⁹, a 56-year-old with no known cardiovascular risk factors and medical history, who had the same diagnosis and there were found mitral valve disease and atrial fibrillation.

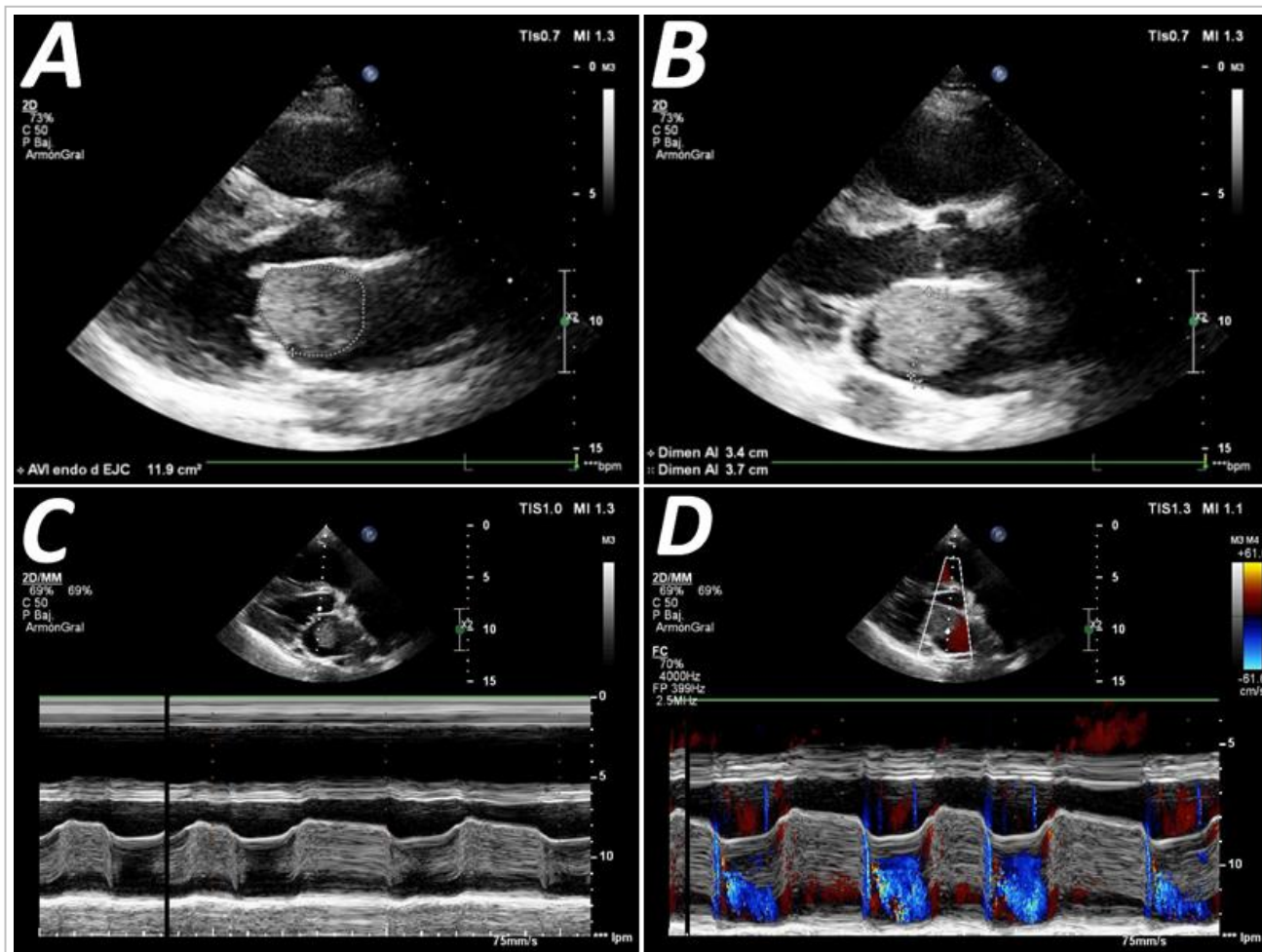


Figure 3. Transthoracic echocardiogram. **A.** Moment of diastole where the myxoma, with an area of 11.9 cm², occludes the mitral orifice. **B.** Anteroposterior diameter of the tumor (34 mm) in relation to that of the left atrium (37 mm). **C.** M mode. **D.** Doppler color in M mode.

According to Bermúdez Yera *et al.*, sometimes the pedunculated LAM can behave as a surgical emergency¹⁷, or may have binaural¹⁸ affection, and according to García Quintana¹⁹ this CT can also become infected.

Other authors have found CM of different characteristics and locations: giant bilobed myxoma of right atrium⁵ and left ventricular myxoma in a patient with human immunodeficiency syndrome²⁰.

In the case presented herein can be observed how the pedunculated LAM moves during diastole to the mitral valve and causes episodes of syncope due to obstruction of the circulation or dyspnea, orthopnea, paroxysmal nocturnal dyspnea and acute pulmonary edema, due to alteration of the ventricular filling. Furthermore, symptoms of cardiac arrhythmia (atrial fibrillation) and of an acute coro-

nary syndrome as a result of the deleterious effects of LAM in hemodynamics and the cardiac cycle that produces coronary flow disturbances, because in the coronary angiography was not found evidence of embolism or other obstructive lesions at that level.

Another aspect of interest was the presence of transitory pulmonary hypertension that appeared when the LAM enclaved in the mitral hole and produced blood reflux into the pulmonary veins, or when an acute edema of one lung (disconcerting element for medical assistance) was detected, that can appear in patients with pedunculated LAM by obstructing –during its movement– the two pulmonary veins on the same side.

The diagnostic imaging method of choice is the echocardiogram, because it is accurate, reliable, inexpensive, bloodless and does not imply any risk

of tumor fragmentation and subsequent embolization. The first was made in 1959 through the M mode. The sensitivity of the transthoracic echocardiography is 93% and 97% for the transesophageal echocardiography^{1,4,5}.

The first surgical resection of a LAM was described by Clarence Crafford 1954⁴, and since then, the surgical nature of this disease was established, and that this is the treatment of choice in selected cases with high risk of valve obstruction or systemic embolization. A broad resection of the tumor implantation base should be performed to avoid recidivism. The long-term prognosis is very good and an annual control echocardiogram is recommended^{4,5,7}.

The diagnosis of CM is based on the clinical suspicion and it is confirmed by a two-dimensional echocardiography. The medical group attending these patients must undertake the most appropriate behavior in a timely way and maintain and optimal echocardiographic monitoring for the rest of her/his life.

CONCLUSIONS

Los mixomas son los tumores cardíacos primarios más frecuentes. A pesar de su benignidad, pueden tener una repercusión potencialmente grave en dependencia de su tamaño, localización anatómica y relación con las estructuras vecinas. Pueden ser asintomáticos, producir múltiples síntomas cardíacos y sistémicos, o tener consecuencias mortales. En la práctica clínica se debe pensar en su diagnóstico ante un paciente con síntomas cardiovasculares inespecíficos. Es necesario el pequizaje para detectar formas familiares. Son tumores que por su rareza y formas de presentación suponen un desafío para el médico.

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