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Case Report

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Pulmonary hypertension syndrome secondary to right atrial myxoma apropos of a case

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Abreviaturas
AM: atrial myxoma

ABSTRACT

Atrial myxoma is a benign primary cardiac tumor that is mostly found in the left atrium. This paper aims to describe the clinical presentation of a right atrial myxoma through the case report of a 54-year-old patient diagnosed with pulmonary hypertension syndrome secondary to the above-mentioned cardiac tumor. The clinical features of myxomas are determined by their location, size and mobility. In this case, intracardiac obstruction produced by the myxoma, with paradoxical movement of the septum, resulted in ventricular filling alterations, low cardiac output signs and severe pulmonary hypertension. Immediate clinical and surgical management after diagnosis successfully prevented the development of major complications. This case highlights the paramount importance of clinical judgment and the use of various cardiac imaging techniques in decision making.
Keywords: Atrial myxoma, Cardiac tumors, Pulmonary hypertension, Heart failure

Síndrome de hipertensión pulmonar secundario a mixoma auricular derecho, a propósito de un caso

RESUMEN

El mixoma auricular es un tumor cardíaco benigno de origen primario que se localiza mayormente en la aurícula izquierda. El presente trabajo tiene como objetivo describir las formas clínicas de presentación de un mixoma de aurícula derecha, a través de la presentación de un caso con el diagnóstico de síndrome de hipertensión pulmonar secundaria al mencionado tumor cardíaco, en una paciente de 54 años de edad. Las características clínicas de los mixomas están determinadas por su localización, tamaño y movilidad. En este caso, la obstrucción intracardíaca producida por el mixoma, con movimiento paradójico del septum, dió lugar a alteraciones en el llenado ventricular, signos de bajo gasto e hipertensión pulmonar grave. El abordaje clínico y quirúrgico inmediato tras el diagnóstico evitó el desarrollo de complicaciones mayores. Este caso enfatiza la vital importancia del juicio clínico y el uso de diversas técnicas de imagen cardíaca en la toma de decisiones.
Palabras clave: Mixoma auricular, Tumores cardíacos, Hipertensión pulmonar, Insuficiencia cardíaca

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INTRODUCTION

Atrial myxoma is a benign primary cardiac tumor that is mostly found in the left atrium. Primary heart tumors are quite rare (5%), 75% are benign and myxoma is the most common type, accounting for approximately 50% of them[1]. Globally, the annual incidence of AM is 0.5 per million population, and of these, 75% are located in the left atrium[2], peaking between 40 and 60 years of age, yielding a female to male ratio of approximately 3:13.

Myxomas have various presentation symptoms depending on their size, mobility and location. They may also be asymptomatic or present with non-specific symptoms such as: fatigue, cough, fever, arthralgia, myalgia, weight loss and erythematous rash. The typical symptomatic triad is embolization, non-specific condition and heart failure. Systemic thromboembolism is found in 25-50% of cases of left myxomas and approximately half of the emboli are directed to the central nervous system, where they may cause ischemia or lead to the development of aneurysms, thus resulting in intracranial hemorrhage, which may be the first manifestation in the diagnosis of myxoma[2-4].

As for the right atrial location, the most frequent cardiac manifestations of intracavitary tumors located at this level are: asthenia, peripheral edema, ascites, hepatosplenomegaly and elevated jugular venous pressure with prominent A-wave. The diagnosis is often late, with an interval of about 2.5 to 3 years from symptomatic presentation to accurate diagnosis. Patients quite often report symptoms compatible with rapidly progressive right-sided heart failure that includes new-onset heart murmurs from mechanical interference with the tricuspid valve. Pulmonary embolization is usually caused by a tumor in the right hemicardium[3].

The diagnosis is based on the clinical features and mainly on imaging techniques, such as echocardiography and chest X-ray. Also important are electrocardiography, cardiac catheterization, hemodynamic tests, computed tomography angiography and magnetic resonance imaging. Of these, the echocardiogram is an undoubtedly valuable tool for the evaluation of intracardiac masses and the best method for the detection of AM. All these techniques provide data on tumor mass, size, mobility and myocardial invasion. Diagnostic confirmation is obtained by histopathology[5,6]. It requires surgical treatment consisting of complete resection of the intracardiac tumor, which must be timely performed to avoid further complications[7]. Survival after surgical resection is high; however, long-term follow-up by echocardiography is recommended, as there is a 1-3% chance of recurrence due to inadequate resection[3,8].

The following is a case report of a 54-year-old woman with a personal pathological history of essential arterial hypertension and ischemic heart disease who presented with a pulmonary hypertension syndrome secondary to right AM requiring surgical treatment at the Department of Cardiology of the *Centro de Investigaciones Médico-Quirúrgicas* (CIMEQ) in Havana, Cuba.

CASE REPORT

A 54-year-old black woman, housewife, urban resident with a personal history of essential hypertension that had started about 10 years ago and recent diagnosis (6 months ago) of chronic ischemic heart disease, who was under treatment with aspirin (100 mg/day), atenolol (50 mg/day) chlortalidone (25 mg/day) and isosorbide dinitrate (30 mg/day), presented to our center assuring that since about 1 year ago she had started with a sharp, stabbing pain in the lateral region of the left hemithorax, radiating to the flank on the same side and towards the back, appearing on exertion and relieving on rest or when sitting; in addition, dyspnea on exertion, which sometimes worsened when lying down, and coughing with whitish expectoration occasionally accompanied by striations of blood. Within 6 months of presenting these symptoms, she was diagnosed with an ischemic heart disease that moderately improved after treatment. Her dyspnea had worsened at subsequent outpatient appointment, mostly appearing at minimal effort and at rest, chest pain was more frequent and long-lasting, and at this point, palpitations had become evident.

Physical Examination

General: Subcutaneous cellular tissue infiltrated in the distal third of the lower limbs.

Regional: Globular abdomen, which follows respiratory movements, soft, depressible. Painful on deep palpation in right hypochondrium. Liver passing through two fingers below the edge of the ribs, with normal hydro-aerial noises.

By systems:

- Respiratory system: Decreased thoracic expandability, with 19 breaths per minute, decreased

vocal vibrations, increased pulmonary sonority at the bases and globally decreased vesicular murmur, with the presence of bibasal crackling rales.

- Cardiovascular system: Downwardly displaced apex beat with parasternal retraction and heart rate of 84 beats per minute (bpm). The right ventricle is palpable at the epigastrium. Rhythmic heart sounds, with decreased S1, loud S2 pulmonary component and II-III/VI holosystolic murmur, which increases at inspiration in tricuspid foci. Jugular ingurgitation with hepatojugular reflux was observed. Parvus, rhythmic, symmetric and synchronic peripheral pulses, with a frequency equal to that of the central heart rate; normal capillary filling. Blood pressure 140/80 mmHg.

Complementary tests results

Hematology and hemochemistry analyses are shown in the **table**.

Electrocardiogram: Sinus rhythm, 74 bpm, normal electrical axis, negative T waves in V_4-V_5 and isodiphasic in V_3.

Chest X-ray (posteroanterior view): Increased cardiothoracic index, signs of pulmonary congestion and radiopaque images of ± 1 cm in the right lung (paracardiac region), which appear to be tumorous. A re-evaluation by computed tomography (CT) is thereby suggested.

Chest CT: Enlargement of the heart area to the detriment of the right atrium, over which a heterogeneous density image of approximately 8.4 × 5.7 cm in diameter can be seen. The presence of some fibroexudative-like lesions, mainly peripheral, in both lower lobes, irregular, involving the pleura, and others with a nodular tendency, larger at the base, measuring up to 14 mm, is remarkable.

Coronary angiography: Epicardial coronary arteries without angiographically significant lesions.

Transthoracic echocardiography: Tumor image in right atrium, motile, hyperechogenic, of 21.97 cm^2, protruding through the tricuspid valve (**Figure 1**). Severe tricuspid regurgitation and pulmonary hypertension, with pulmonary artery systolic pressure of 77 mmHg (**Figure 2**), and preserved left ventricular systolic function (LVEF 59.7% by Simpson). The transesophageal echocardiogram confirmed the diagnosis.

The patient was discussed with the department of cardiovascular surgery for surgical treatment and –under cardiopulmonary bypass, anoxic arrest and moderate hypothermia– the tumor mass was completely removed. An incision throughout the right atrium was performed, and the tumor was resected without further complications (**Figure 3**). The results of the anatomopathological study confirmed the histological characteristics of the myxoma, which was found to be quite fragile, which facilitated the detachment of fragments towards the minor cir-

Table. Blood tests performed.

Complementary	Results
Hemoglobin	10,7 g/L
White blood cells	6,8 × 10^9/L
Platelets	368 × 10^9/L
Glycemia	4,8 mmol/L
Creatinine	119 µmol/L
Urea	5,9 mmol/L
Uric acid	302 mmol/L
GPT	21,2 U/L
GOT	29,3 U/L
GGT	34 U/L
Cholesterol	3,88 mmol/L
Triglycerides	0,82 mmol/L
Total proteins	73 g/L
Albumin	38 g/L
CK, CK-MB and troponins	Normales

CK, creatine kinase; CK-MB, isoenzyme creatine kinase MB; GGT, gamma-glutamyl- transpeptidase; GOT, glutamic oxalacetic transaminase; GPT, glutamic pyruvic transaminase

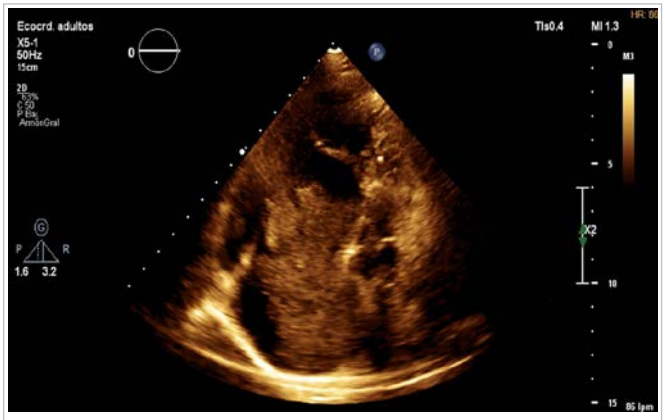

Figure 1. 4-chamber view transthoracic echocardiogram showing the mass at rigth atrial level.

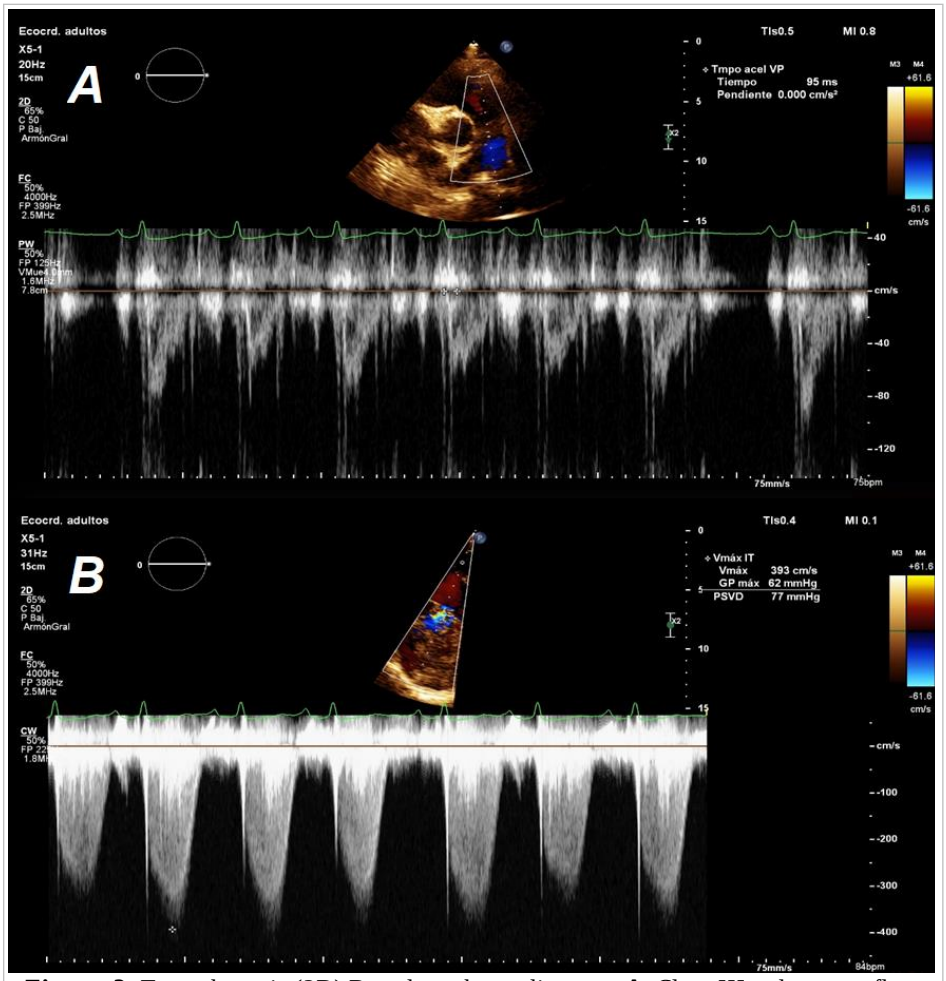

Figure 2. Transthoracic (2D) Doppler echocardiogram. **A.** Class III pulmonary flow curve. **B.** Tricuspid regurgitation with estimated right ventricular systolic pressure of 77 mmHg.

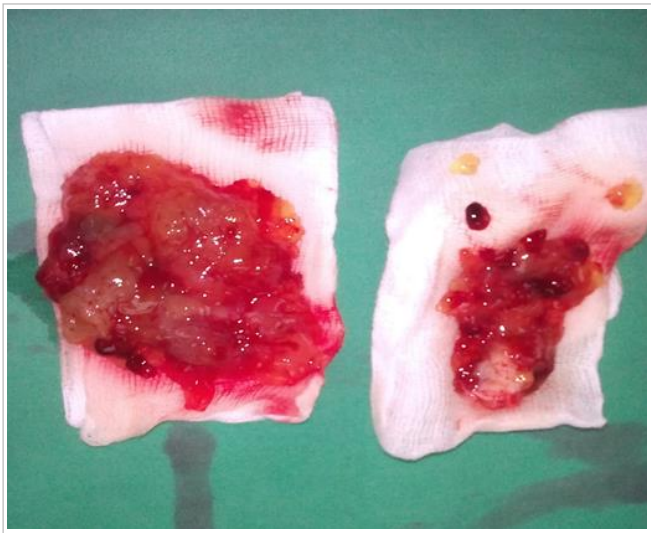

Figure 3. Anatomical piece of the myxoma extracted from the right atrium.

culation that caused the pulmonary hypertension due to embolic phenomena.

The diagnosis of AM and its specific presentation with severe pulmonary hypertension and decompensated right heart failure was based on the clinical findings, the course of the disease, the echocardiographic study, and mainly the anatomopathological study that definitely led to the diagnosis of myxoma. Other possible causes implying a probable etiology of pulmonary arterial hypertension were ruled out and therefore the chronic thromboembolic origin was confirmed. The patient had a favorable postoperative outcome and was discharged after 7 days under medical treatment.

COMMENTS

Cardiac tumors are relatively unusual, with a prevalence of only 0.002-0.3%; the AM has a presentation age ranging from 40 - 60 years[3,8]. Only three studies have been published in Cuba that reflect statistics on myxomas. At *Hermanos Ameijeiras Hospital*, between January 1986 and January 2006, it was found that 1.02% of the 2925 patients operated on using extracorporeal circulation were AM, which is similar to the results published in the international literature[3]. The incidence in females predominated, with a ratio of 2:1, with respect to males and an average adult age between 51-60 years[9]. Other series, enrolling 38 and 15 patients from the Santiago de Cuba[10] and Villa Clara[11] Cardiocentros respectively, who were treated between 1986 and 2010, exhibit similar results.

The cardiac myxoma is normally located in the left atrium (83%), where it is usually attached to the oval fossa, the anterior mitral leaflet or the posterior wall, and successively occurs in the right atrium

(12.7%). Nearly 1.3% are biauricular[12,13]. The presentation in Cuba is similar[9-11].

Cardiac myxoma is a neoplasm of uncertain and controversial histogenesis that occurs only on the endocardial surface and is usually located in the atria. Its histological diagnosis is based on the finding of typical cells in a mucopolysaccharide-rich matrix, containing cells that are histologically and histogenetically different from the spindle cells of soft tissue myxomas. Those that give rise to this tumour are the so-called "subendothelial reserve cells", which are totipotent and have the capacity to form vascular structures, expressing endothelial and neural markers[14,15]. In the case presented here, the results of the anatomopathological study were confirmatory.

Cardiac myxoma may be asymptomatic and be found incidentally by means of an echocardiogram; however, on other occasions it may present with systemic or constitutional symptoms due to the release of inflammatory cytokines; cardiovascular symptoms, such as myocardial infarction, acute or chronic heart failure, arrhythmias or mimic mitral stenosis (considered as a differential diagnosis). It may also cause neurological symptoms and cardioembolic manifestations in the systemic or pulmonary circuit[12].

In this case, the patient presented a clinical picture of chronic thromboembolic pulmonary hypertension secondary to AM in the right atrium, protruding through the tricuspid valve; all of which explains the symptomatology of chest pain and the manifestations of heart failure: shortness of breath on exertion, jugular ingurgitation and edema in the lower limbs, which coincides with the national[9,10,13] and international scientific literature[16-19].

Chronic thromboembolic pulmonary hypertension–classification provided at the IV World Symposium on Pulmonary Hypertension held in Dana Point, United States, the V World Symposium on Pulmonary Hypertension held in Nice, France, and the recent update by the European Society of Cardiology (ESC) and European Respiratory Society (ERS) scientific societies–occurs as a result of unresolved thromboembolic occlusion of the pulmonary vessels and by vascular remodeling. It mainly affects large vessels which makes it generally susceptible to surgical treatment. While the exact data are unknown, the cumulative incidence of this disease is 0.1-9.1%, 24 months after the first event of acute pulmonary thromboembolism[17]. It is estimated that nearly 2500 cases take place in the United States each year and some data suggests that such an event may occur in approximately 5 individuals per million population a year[18].

About 40% of patients with pulmonary thromboembolism may be completely asymptomatic, which entails a difficult and often challenging diagnosis[20]. Approximately 3.8% of patients who manage to survive the first event relapse within the following two years[19]. The symptoms– dyspnea, fatigue, syncope, chest pain, hemoptysis, and right-sided heart failure– flare up when 40 to 50% of the pulmonary circulation is compromised. Bilateral pulmonary endarterectomy via the pulmonary artery, under deep hypothermia and circulatory arrest, without the need for cerebral perfusion, still remains the surgical technique of choice for the treatment of chronic thromboembolic pulmonary hypertension[21].

The importance of the present case lies in pointing out that the complications associated with this intracardiac tumor and its particular form of presentation can be avoided with early diagnosis and timely surgical resection of the AM; thus, early clinical judgment and the use of various cardiac imaging techniques is critical in decision making; which, in this particular case, was surgical management.

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