CorSalud. 2022 Ene-Mar;14(1):89-93

CorSalud

Cuban Society of Cardiology

Case Report

Cardiocentro ERNESTO CHE GUEVARA SANTA CLARA CUBA

An asymptomatic right ventricular myxoma case without right ventricular outflow tract obstruction or tricuspid valve regurgitation

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Full English text of this article is also available

ARTICLE INFORMATION

Received: February 3, 2021
Accepted: March 24, 2021
Online: June 4, 2021

Competing interests
The authors declare no competing interests.

Figures
Written informed consent was obtained from the patient's legal guardians/parents and the patient itself for publication of this case report and any accompanying images.

ABSTRACT

A 24-year-old male patient, who had no complaints before, applied to our hospital for general tests in order to get a medical report due to his job application. A murmur was detected by the physician in the internal medicine clinic as a finding of listening and was referred to the cardiology clinic. The oval, well-circumscribed lobulated echogenic mass appeared in right ventricular in the research performed in the cardiology clinic and it was transferred to our clinic for emergency surgery. The right ventricular mass did not move towards either the pulmonary valve or the tricuspid valve during systole and diastole. During the operation, it was determined that the tumor originated from the right ventricular free wall. Right ventricular mass excision was performed with right atriotomy under cardiopulmonary bypass. The pathology confirmed the diagnosis of myxoma. At the sixth month of follow-up, the patient was asymptomatic and there was no evidence of myxoma recurrence.

Keywords: Right ventricular myxoma, Cardiac tumors, Surgical excision, Cardiac surgery, Emergency surgery

Mixoma ventricular derecho asintomático sin obstrucción del tracto de salida o insuficiencia valvular tricuspídea

RESUMEN

Hombre de 24 años de edad, sin síntomas previos, que solicitó la realización de exámenes generales en nuestro hospital para obtener un informe médico de aptitud preempleo. El especialista de la clínica de medicina interna le detectó un soplo, por lo que fue remitido a cardiología, debido a este hallazgo auscultatorio, donde se realizaron otros exémenes y se encontró una masa ecogénica ovalada y lobulada, de bordes bien definidos, en el ventrículo derecho; razón por la que el paciente fue transferido a la clínica de cirugía cardíaca para tratamiento quirúrgico urgente. La masa ventricular derecha era sesil, por lo que no se movía hacia la válvula pulmonar ni a la tricúspide durante la sístole y la diástole. Durante la operación, realizada mediante auriculotomía derecha y circulación extracorpórea, se extrajo el tumor que se originaba en la pared libre del ventrículo derecho. El examen histológico confirmó el diagnóstico de mixoma. Al sexto mes de seguimiento, el paciente estaba asintomático, sin evidencia de recurrencia del mixoma.

Palabras clave: Mixoma de ventrículo derecho, Tumores cardíacos, Escisión quirúrgica, Cirugía cardíaca, Cirugía urgente

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INTRODUCTION

Cardiac myxoma is the most common primary heart tumor in adulthood. They often originate in the left atrium, but can also be found in the ventricular cavities. Approximately, 75% are located in the left atrial cavity, 23% in the right atrial cavity, and about 2% in a ventricular cavity[1,2]. Typically cardiac myxomas; while solitary, pedunculated, and arising in the vicinity of the fossa ovalis, they can sometimes be multicentric, sessile or attached to other areas of the endocardium.

Cardiac tumors originating from the right ventricular outflow tract present unusual diagnostic and therapeutic challenges. Symptoms depend on the size and location of the tumor. Such tumors can have significant clinical consequences, including arrhythmias, pulmonary embolism, tricuspid valve regurgitation, pulmonary valve stenosis, and sudden death. Diagnosis at present is established most appropriately with two-dimensional echocardiography. Treatment includes surgical resection followed by continuous monitoring with echocardiography. The surgeon should perform a surgical procedure that prevents tumor rupture and intraoperative embolization and postoperative recurrence[1,2].

We present a 24-year-old male patient with right ventricular myxoma adherent to the free wall of the right ventricle, moving in the right ventricle, and not causing right ventricular outflow tract obstruction and tricuspid valve regurgitation.

CASE REPORT

A 24-year-old male patient, who had no complaints before, applied to our hospital for a medical report to be used in his job application. The patient was referred to the internal medicine clinic to prepare a report at the hospital. His heart rate was 77 beats/minute and the blood pressure was 122/70 mmHg at admission. Chest radiography and electrocardiography were normal. On physical examination, a grade 4/6 systolic ejection murmur was heard at the left upper sternal border. Therefore, he was sent to the cardiology clinic.

Transesophageal echocardiography revealed a large 10 × 5 mm mobile mass attached to the free wall of the right ventricle, not extending to the right ventricular outflow tract, pulmonary and tricuspid valve, and located only in the right ventricle (**Figure 1A**). Left ventricular ejection fraction was approximately 60% stable.

Primarily, it was thought that there might be a cardiac thrombus or myxoma, and cardiac tomography was performed. Cardiovascular tomographic examination revealed delayed heterogenous enhancement of the right ventricular mass, suggesting a myxoma (**Figure 1B**). There was no family history of myxoma. Due to his young age, the presence of Carney syndrome was investigated. However, no pigmentation, endocrine system pathologies or additional non-cardiac adenomas were detected in the patient.

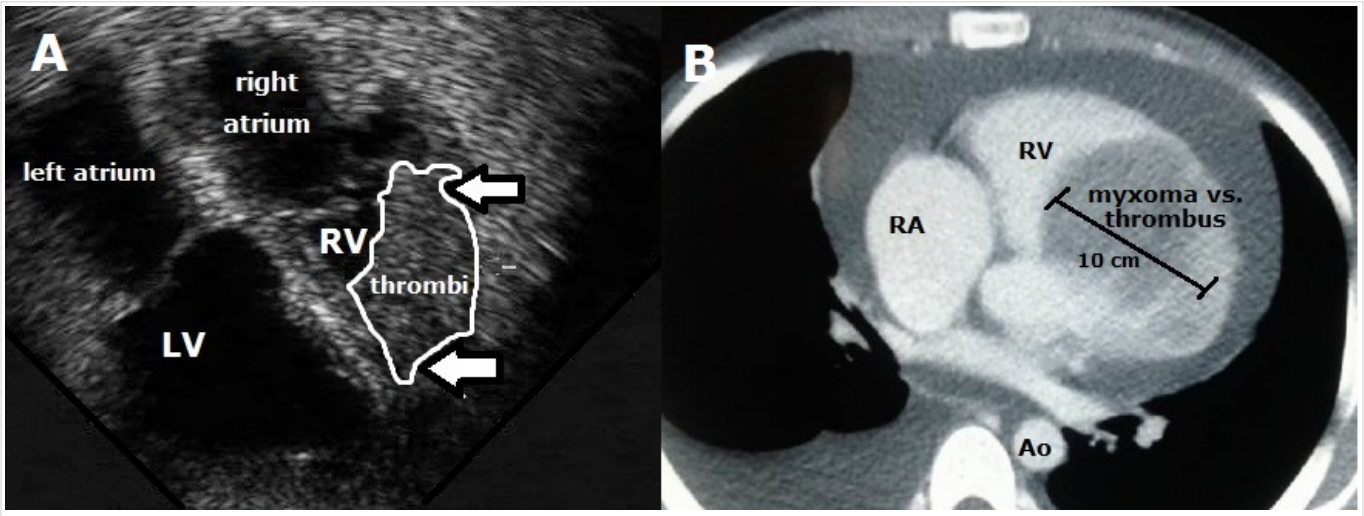

Figura 1. A. Transesophageal echocardiography 4 chamber view showing a mass in the right ventricle and adherent to the right ventricular free wall (arrows). **B.** Cardiac computed tomography showing a right ventricular mass approximately 10 cm long extending into the free wall of the right ventricle (Ao, aorta; LV, left ventricle; RA, right atrium; RV, right ventricle).

The patient was taken to the Cardiovascular Surgery department for surgical intervention. Classical sternotomy was performed at surgery, and cardiopulmonary bypass was performed with standard bicaval and ascending aortic cannulation with moderate hypothermia. Right atrium was opened after cardioplegic arrest. As the tricuspid leaflets retracted, a gelatinous mass was visible in the right ventricle and originating from the free wall (**Figure 2A**). The mass was excised in a block with the ventricular muscle segment without any defects in the right ventricular free wall, and the tumor base was thermocoterized. Pathologically, a jelly-like mass with hemorrhagic areas was seen macroscopically (**Figure 2B**).

After tumor removal, tricuspid valve coaptation and adequacy were checked with saline wash solution, and no deficiencies were detected. The patient showed an excellent overall recovery and was discharged home on the seventh postoperative day. The pathology showed stellate cells with eosinophilic cytoplasm consistent with the final diagnosis of myxoma (**Figure 2C**). At the 6th month follow-up, the patient was asymptomatic and there is no evidence of myxoma recurrence.

COMMENT

Primary cardiac tumors are generally rare masses and most commonly occur as myxoma. While 75% of myxomas are located in the left atrium, right-sided myxomas are seen in 15-20%. Right ventricular myxomas are very rare (1-2%)[3-5]. Myxomas are thought to arise from the proliferation of multipotent mesenchymal cells. Cardiac myxoma is seen twice as often in women[3-5]. Myxomas are usually sporadic, but there are cases that are familial. The Carney complex, which was defined as a combination of myxoma, skin pigmentation, and hyperactivity of the adrenal or testicular glands, was described in 1985 and other parameters of this syndrome were investigated in each case of myxoma[6,7].

Myxomas originating from the right ventricle are extremely rare[1,2] and can cause obstructive symptoms and tricuspid valve regurgitation, usually resulting from occluding the right ventricular outflow tract and pulmonary valve. Symptoms and signs include peripheral edema, acid and shortness of breath as a result of right ventricular outflow tract obstruction (signs of right heart failure)[8]. In addition, it can cause complications such as syncope, pulmonary embolism, and sudden death. In addition, patients may experience structural symptoms such as fatigue, fever, arthralgia, and myalgia. In some cases, it may cause advanced tricuspid valve regurgitation and signs of right heart failure due to preventing the tricuspid valve from closing or the mass adhering to the tricuspid valve[2,8,9]. Tricuspid valvectomy without prosthetic valve replacement, tricuspid valvuloplasty with partial excision of the tricuspid valve and tricuspid valvectomy with prosthetic valve replacement can be performed in these patients. Right ventricular myxoma in the present case affected neither the right ventricular outflow tract nor the functions of the tricuspid valve. Therefore, excision of the mass in the right ventricular cavity alone was sufficient for treatment. No repair or correction was made for the valves of the right heart cavity.

Myxomas seen in early childhood and adoles-

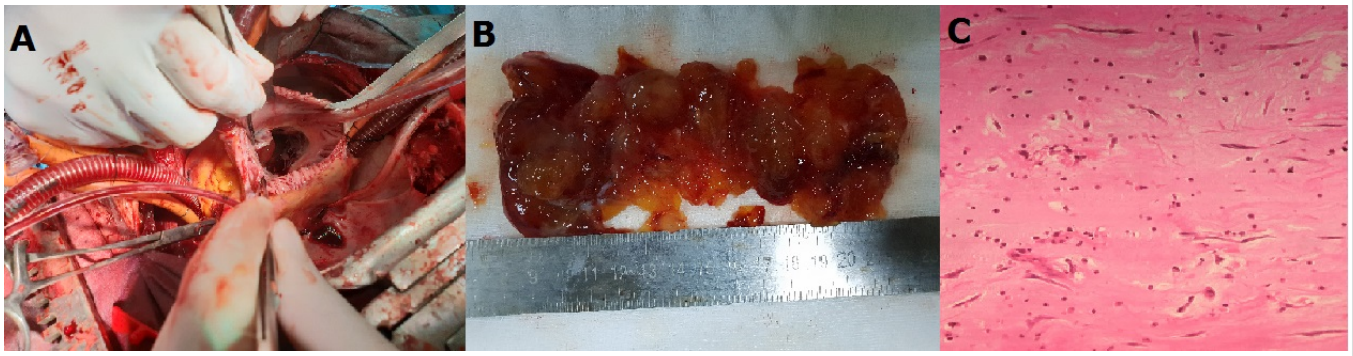

Figura 2. A. The appearance of the myxoma in the right ventricle with exclusion of tricuspid valve leaflets after right atriotomy. **B.** Gross specimen of right ventricular myxoma, consisting of multiple fragments of soft, myxoid, gelatinous tissue, with loosely adherent blood clot. **C.** Histology. Stain: Haematoxylin–Eosin 200×. Cardiac myxoma cell morphology can be variable, with irregular spindle and stellate shapes and abundant eosinophilic cytoplasm.

cence are extremely rare and this pathology may occur as a component of Carney's syndrome, more commonly known as familial myxoma syndrome. Carney complex with an autosomal dominant inheritance pattern should be considered, especially when there are multiple, multifocal masses in the ventricular chambers. In addition to cardiac myxoma in cases with this syndrome, various adenomas and mucocutaneous, visceral and endocrine disorders are frequently accompanied. Although our patient was young, he did not have a familial story and parameters of Carney syndrome.

The gold standard noninvasive diagnostic method for such tumors is transthoracic or transesophageal echocardiogram. The echocardiogram facilitates the preoperative location, size, shape, mobility of the tumor, as well as right ventricular outflow tract obstruction and the tumor attachment site and the selection of the optimal surgical approach. Transesophageal echocardiography accurately identifies other localization of myxomas[4]. Cardiac computed tomography and magnetic resonance imaging provide additional and useful information about the structure and function of cardiac tumors prior to surgical resection. They also play an important role in the differentiation of intracavitary cardiac masses (malignant mass, thrombus, myxoma and lipoma)[10,11]. The diagnosis of myxoma is confirmed only in pathology, which usually involves an acid-mucopolysaccharide-rich stroma mixed with polygonal cells. In our patient, a simple echocardiography was helpful in diagnosis, and the presence of myxoma was supported by tomography. In addition, our patient was tried to find a familial syndrome or congenital anomaly by performing additional pathological investigations on both the cardiac and the whole body, but no other additional finding was found. In addition, the final diagnosis was confirmed by postoperative histopathological findings.

Myxomas should be treated with emergency surgical resection as soon as they are diagnosed due to obstructions due to their localization and complications related to their embolism risks. In addition, great attention should be paid to hemodynamic parameters and cardiac manipulations in preparation for the operation. Although surgical procedures are generally preferred depending on the localization, different surgical methods can be applied. The root of the pedicle —the base of the tumor— must be completely removed to prevent recurrences. Surgery is usually curative and has a low operative mortality rate. Recurrence is thought to be caused by incomplete resection or de novo formation. The recurrence rate of myxoma has been reported to range from 1% to 5%[12,13]. In our patient, we closely monitored the hemodynamic parameters during anesthesia, and kept the femoral region ready for immediate cardiopulmonary bypass if needed. In addition, we decided —like other authors did[2,10,11]— that the best place to surgically reach a right ventricular mass was the right atrium and performed the surgical procedure. A curative surgical excision was performed to prevent recurrence and the excision site was additionally cauterized. There were no symptoms nor recurrence in the sixth month of follow-up.

CONCLUSIONS

This case reports our surgical experience treating a rare type of right ventricular myxoma. With this case; We wanted to emphasize that asymptomatic right ventricular myxoma can be diagnosed with simple examination methods, surgery to be performed without delay is life-saving, a curative resection during surgery is required to prevent recurrence, and adjacent heart valves should be surgically evaluated. These tumors can be excised with low morbidity and mortality rates. For patients after surgical resection, the prognosis is excellent. In addition, patients should be followed up in terms of recurrence.

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