CorSalud 2018 Oct-Dec;10(4):330-335

CorSalud

Cuban Society of Cardiology

Case Report

Cardiocentro ERNESTO CHE GUEVARA SANTA CLARA CUBA

# Rare coincidence of two tumors: cardiac myxoma and hypernephroma. A case report

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ARTICLE INFORMATION

Recibido: June 27, 2018
Accepted: August 9, 2018

**Competing interests**
The authors declare no competing interests

**Acronyms**
**CPB:** Cardiopulmonary bypass
**LA:** Left atrium
**TTE:** transthoracic echocardiogram

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## ABSTRACT

The association of two tumors of different origin in the same patient is becoming more frequent in the current clinical practice. Here is presented a case with a rare association of cardiac myxoma and hypernephroma, previously treated. Due to the presence of neurological symptoms, there was initially set a diagnosis of brain metastasis from the renal tumor; then, with the echocardiographic finding of an intracardiac mass, the possibility of thrombus was considered, therefore a cardiac surgery was decided to be performed urgently in order to resect it. The pathologic examination confirmed the existence of a cardiac myxoma, then, we have a patient with two tumor diseases whose coincidence has been rarely described.

*Keywords:* Atrial myxoma, Renal cell carcinoma, Embolism, Multiple primary neoplasms

## *Rara coincidencia de dos tumores: mixoma cardíaco e hipernefroma. Presentación de un paciente*

## RESUMEN

*La asociación de dos tumores de diferente origen en un mismo paciente es cada vez más frecuente en la práctica clínica actual. Se presenta un caso con una rara asociación de mixoma cardíaco e hipernefroma, previamente tratado. Ante la presencia de manifestaciones neurológicas, inicialmente se planteó el diagnóstico de metástasis cerebral del tumor renal; luego, con el hallazgo ecocardiográfico de una masa intracardíaca, se pensó en la posibilidad de trombo, por lo que se decidió practicar cirugía cardíaca con carácter urgente para resecarla. El estudio anatomopatológico confirmó la existencia de un mixoma cardíaco, por lo que se trata de un enfermo con dos enfermedades tumorales cuya coincidencia ha sido pocas veces descrita.*

*Palabras clave: Mixoma auricular, Carcinoma de células renales, Embolismo, Neoplasias primarias múltiples*

## INTRODUCTION

Los mixomas son los tumores primarios más frecuentes del corazón[1-3]. Suelen tener una presentación clínica muy variada que depende de su localización y grado de obstrucción mecánica. Habitualmente sus síntomas pueden agruparse en: cardíacos, generales y embólicos[2,3]. Años atrás su reconocimiento era excepcional por la escasez de los medios diagnósticos, lo cual era una barrera difícil de sortear desde el punto de vista terapéutico. El desarrollo de la cirugía cardíaca con circulación extracorpórea (CEC) a partir de la década de los sesenta del siglo pasado ha permitido tratar muchos enfermos que han sido curados con la resección quirúrgica, convirtiéndola en la principal arma para su erradicación. Su asociación con otros tipos de tumores ha sido muy poco informada[4-6].

Recientemente hemos operado un paciente con diagnóstico previo de carcinoma de células renales y embolismos cerebrales múltiples. La presencia de lesiones a distancia del tumor primario llevó a plantear la posibilidad de metástasis del mismo tumor como primer diagnóstico; pero el estudio ecocardiográfico identificó una masa intracardíaca, lo que hizo replantear nuevas posibilidades diagnósticas y la remoción quirúrgica para identificar su histología y conducta definitiva.

## CASE REPORT

Here is presented the case of a 50-year-old mixed-race man with no medical history of interest, that seven months before was referred to our center due to high figures of blood pressure. He was studied in his health area and diagnosed a left renal tumor, thus, he was sent to the *Hospital Clínico Quirúrgico* Gustavo Aldereguía Lima of Cienfuegos, Cuba. He was taken to the operating room, a left nephrectomy was performed and the histological study concluded that it was a renal tubular papillary adenocarcinoma (hypernephroma) stage I: a tumor bigger than 7 cm growing to the organ surface without infiltrating the capsule or collecting system, with diffuse intercapillary glomerulosclerosis in the non-tumoral renal parenchyma. The tumor does not infiltrate perirenal fat.

He was evaluated by Oncology (without specific treatment because the surgical procedure was considered sufficient) and he was discharged after 15 days with follow-up by Urology. After five months of the surgery he was readmitted at the hospital of Cienfuegos due to a loss of consciousness, muscle strength and leg cramps. The physical examination showed disturbances of balance and gait, vertigo and vomiting. An MRI (magnetic resonance imaging) was performed, which reported: hypodense image of 8 mm in the right cerebellar hemisphere and poorly defined hypodense suggestive area of infiltration at that level, with another image in the left cerebellar hemisphere, which does not vary with the contrast, of approximately 9 mm and that impresses not recent ischemic lesion. The possibility of metastasis to the cerebellum of the primary tumor, although it is one of its less frequent forms of invasion to other organs, was the most logical variant and it was the first diagnosis proposed. A symptomatic treatment was imposed and the clinical study was started.

A transthoracic echocardiogram (TTE) was performed, in which a preserved ventricular function was observed, as well as a mass in the left atrium (LA) of 45 × 32 mm, with dynamic occlusion of the

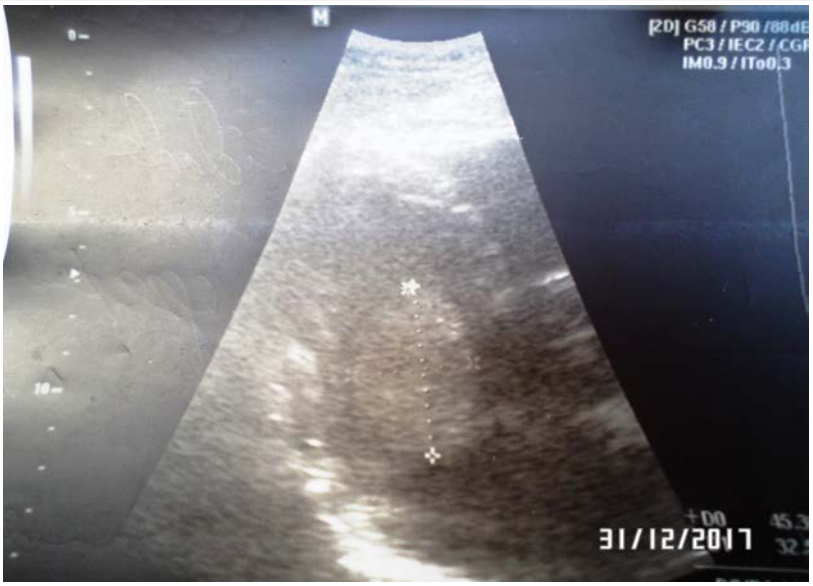


**Figure 1.** Transthoracic echocardiogram performed at the provincial hospital, where an intracardiac mass of 45 × 32 mm, diastolic protruding into the left ventricle is observed.

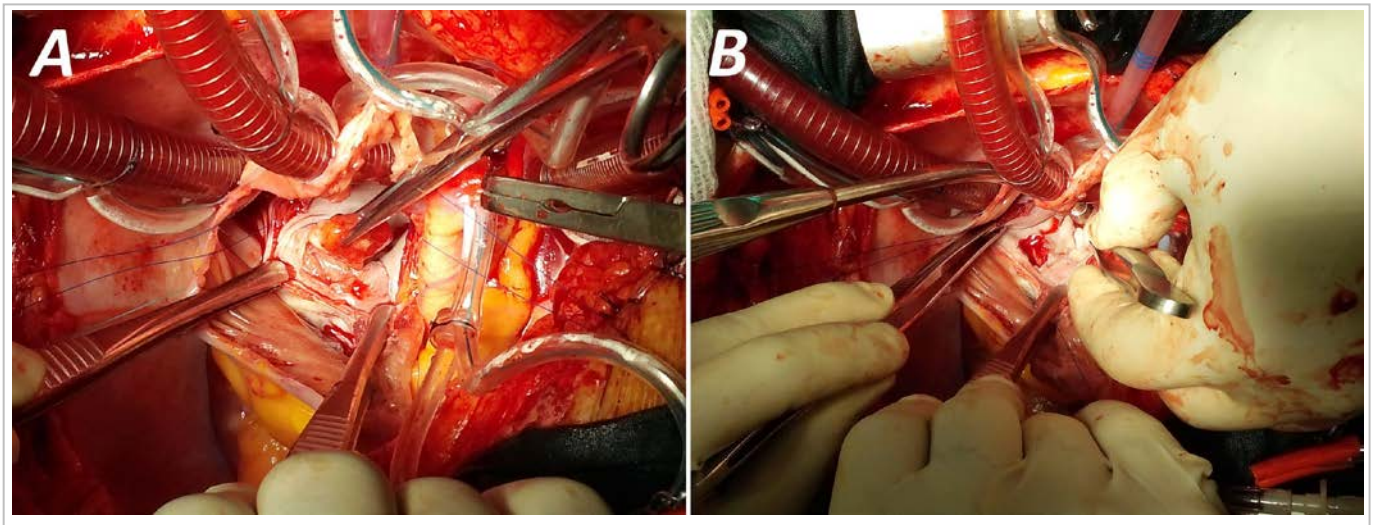


**Figure 2.** Surgical exeresis of the tumor mass. **A.** Located close to the mitral valve and fixed for dissection. **B.** Totally resected from the left atrium. The surgical edges of the tumor's base are observed.

left ventricular outflow tract (**Figure 1**). The patient was moved to the Cardiocentro Ernesto Guevara of Santa Clara, Villa Clara, Cuba, to conclude the study, where it was found that he had a preserved general condition; he had great difficulties in walking and incoherent language, as well as a discrete dyspnea. The heart rate and blood pressure values were normal and a diastolic murmur in the mitral focus was auscultated. The rest of the physical examination was normal. In blood tests, there was only a remarkable normocytic and normochromic anemia and elevation of acute phase reactants. The electrocardiogram and the postero-anterior thorax radiography were normal.

The transesophageal echocardiography confirmed the findings previously described, with a larger mass in the LA (54 × 35 × 22 mm) adhered to the atrial wall and mild mitral stenosis. The left ventricular function was preserved. The coronary angiography showed no significant lesions, and the abdominal ultrasound and the computerized axial tomography with contrast showed no evidence of re-

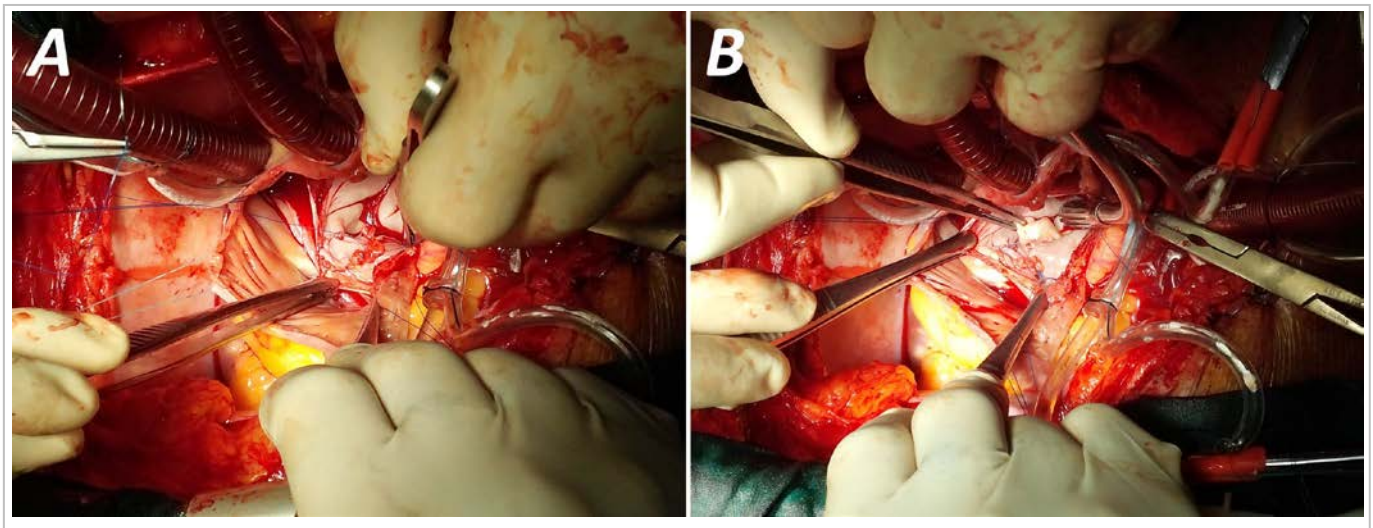


**Figure 3. A.** Autologous pericardium patch implant. **B.** Implanted patch and examination of the mitral anterior leaflet (dissecting forceps).

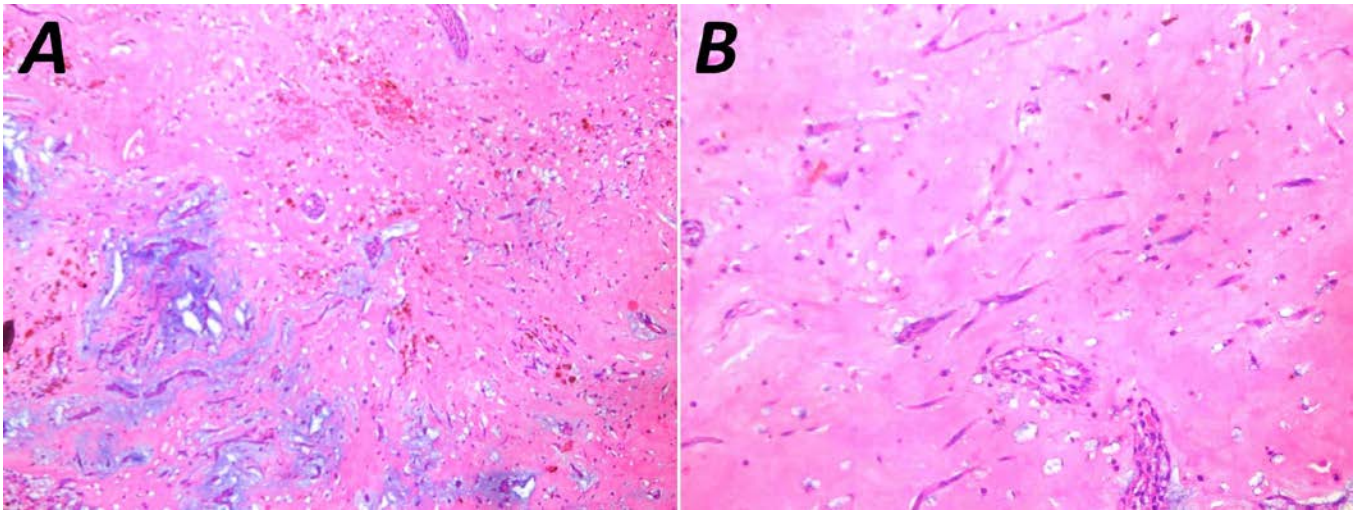


**Figure 4.** Microscopic images of the anatomical specimen as corresponding to a cardiac myxoma. **A.** Note the abundant myxoid matrix of the stroma (Hematoxylin-Eosin × 20). **B.** Stellate cells and pseudoglandular formations are observed (Hematoxylin-Eosin × 40).

currence of the renal tumor previously resected, no adenopathies, or infiltration of organs of the abdominal cavity.

The case was discussed with the cardiovascular surgery department and there was decided to perform an emergency surgery to resect the intracardiac mass, given the possibility of a secondary or metastatic tumor of the heart. A longitudinal median sternotomy and an bicaval aortic and venous arterial cannulation were performed in order to enter in cardiopulmonary bypass, and after the aortic clamping and anoxic arrest with isothermal blood cardioplegia, the opening of the interatrial septum and the LA roof was also performed. A gelatinous, friable tumor mass of about 6-8 cm in the LA was exposed, with wide implantation base close to the anterior mitral veil without damaging the valve apparatus (**Figure 2**). It was completely resected including its implantation base (**Figure 2B**) and an autologous pericardial patch was placed to repair the atrial wall (**Figure 3**). The adequacy of the mitral valve apparatus was checked and it was decided not to act on it. After closing the LA roof and SIA, the next step was the aortic unclamping, closing the right atrium, and deaeration maneuvers and cardiopulmonary bypass output with progressive decannulation (54 minutes of anoxic arrest and 86 minutes of CPB).

In the Intensive Care Unit, the patient presented rhythm disorders (atrial fibrillation with rapid ventricular response), therefore he needed electrical cardioversion twice; he was extubated in the first four hours of the postoperative period, transferred to the conventional ward in 48 hours and discharged from the hospital ten days after surgery.

The histology of the tumor was compatible with a cardiac myxoma (**Figure 4**). At 6 months postoperatively the patient was fully recovered, free of symptoms and the monitoring TTE showed no evidence of thrombus, stroke, or intracardiac mass; the cardiac chambers were of normal size, there was no mitral regurgitation and the left ventricle ejection fraction was 58.6%.

## COMMENT

The description of multiple primary neoplasms dates from the late nineteenth century. Warren and Gates established the clinicopathological criteria for its diagnosis[4]. The frequency of clinical presentation is from 1.5 to 5.4% of cancers and from 5 to 11% in autopsies. In recent years, there has been an increase in early diagnoses of second tumors[5], perhaps because of their state, at better monitoring of patients, advances in diagnostic procedures and improved survival from the diagnosis of a primary tumor.

The development of a second malignant tumor after the treatment of the first with radiotherapy or chemotherapy is not uncommon; however, other factors such as age, genetic alterations, the type of primary tumor, exposure to certain substances or

pathogens, the patient's inheritance and immunological status have been related to the risk of presenting multiple tumors either synchronously or sequentially[6]. It is understood by synchronous neoplasia the one diagnosed at the same time that the neoplasia index, and metachronous neoplasia, that which appears after the diagnosis of the first, according to that the interval between one and another is less than or greater than 6 months, respectively. Although there is no unanimity of criteria in this time interval, some authors claim that, at lower intervals, the percentage of synchronous neoplasms that have gone unnoticed may be high[7].

The genitourinary system is frequently involved in cases of multiple primary neoplasms. The urological organs are one of the settlement sites of some of the primary tumors in almost a quarter part of the described cases[8], suggesting a susceptibility/target increased of this system to the neoplastic disease. That is why the clinical follow-up of this subgroup of patients should be narrow and screenings of the second most common primary neoplasms should be done, as the risk of another independent tumor after a urothelial tumor appears to be considerably increased.

The renal cell carcinoma, hypernephroma or Grawitz tumor, represents 90-95% of primary renal malignant tumors, it affects more men than women, in a 2:1 ratio and it is most common after the age of 50, although in the literature cases have been reported at any age[9-11]. Its classic urological presentation triad, described by Guyon (hematuria, abdominal pain and flank tumor), is rarely observed in practice. The heterogeneity of its clinical presentation has caused it to be called the "internist tumor". It is a malignant tumor that metastasized early, mainly to lymph nodes, lungs, adrenal glands, liver and bones[12]. It can present metastatic disseminations to rare sites, among them, the heart[13], although, when it takes places, it is usually to the right atrium through the inferior vena cava. Apart from this route of spread, cardiac metastases of the renal carcinoma are rare[14,15]. It is associated with tumor thrombosis that can extend to the renal vein, inferior cava and reach the right atrium. There are several reports of combined surgery for exeresis of renal tumors and right atrial thrombi, where most authors recommend CPB when the thrombus settles in the right atrium[16-18].

Brain tumors can be asymptomatic or cause a diverse and varied clinical symptomatology, according to their location, mobility and histological type. They have been called the great simulators[1], as they may take place almost with any cardiac symptom. In the presence of a primary cardiac tumor, initially it should be thought to be a benign tumor, the myxoma, since this is the most common and accounts for more than half of the cases[15]. The myxoma is a mass of intracardiac growth that develops, in 75% of cases, in the left atrium, 20% in the right atrium and the rest in ventricles and occasionally, in valves. It usually affects middle-aged patients, although there is a wide range (11-82 years) and a predominance of females. Its clinical presentation will depend on the location, size and tendency to embolize, although 20% of them are asymptomatic at the time of diagnosis.

Classically, systemic symptoms are described, such as fever, petechiae, arthralgia or lethargy. The physical examination may reveal the existence of an obstructive mitral murmur and, rarely, the auscultatory sign of a tumor "plop" in cases where the myxoma occupies all the mitral valve plane. The clinical presentation is dyspnea or systemic embolization (not metastasis), in general to the central nervous system (letter of presentation of our case), coronary arteries, aorta, kidney, spleen or limbs. Rarely, they can be associated with breast or cutaneous myxomas, testicular tumors, ovarian hyperactivity, pigmentation or *schwannomas* forming Carney syndrome[2,3].

The echocardiography is the diagnostic method par excellence, the sensitivity of TTE for diagnosing the myxoma is approximately 95%, reaching up to 100% with transesophageal echocardiography[19]; and surgery provides high cure rates with low associated morbidity and mortality, thus, that the emergency treatment should be offered, in selected cases, to prevent embolic complications.

The association of a cardiac myxoma with a hypernephroma is rare, with very few bibliography reports[20] and in the series describing the multiple primary tumors. They are two complex clinical diseases to diagnose because of the variety of clinical presentations, not as uncommon as it was thought until recently, and that the development of imaging techniques and surgical procedures have completely revolutionized them; this allows to incorporate them into the lists of treatable neoplastic diseases and with high cure rates, when discovered in the early stages.

## CONCLUSIONS

The increasing frequency of occurrence of second

tumors should be kept into account in the diagnosis and monitoring of patients with oncological diseases, with emphasis on tumors of the genitourinary system. Patients should have a close follow-up with high sensitivity imaging techniques. The surgical resection is the only effective therapeutic form for patients with cardiac myxoma, and the emergency treatment should be offered, in selected cases, in centers with equipment and trained personnel in cardiac surgery.

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