

## An Unfortunate Discovery of an Extremely Rare Right Ventricular Outflow Tract Myxoma in a 30 Year Old Moroccan Patient

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### Abstract

### Case Report

The most common yet rare benign cardiac neoplasms are myxomas. They usually arise from the left atrium next to the fossa ovalis of the intra-atrial septum. The treatment is generally a surgical resection due to an increased risk of embolic and cardiac complications. However, the prognosis following surgery is excellent if diagnosed early. We present a very rare case of a right ventricular outflow tract myxoma discovered in a 30 years old patient presented with dyspnea. Our patient underwent an echocardiographic evaluation and we found a myxoma located in the right ventricular outflow tract that was excised. This location is extremely rare and presents unusual diagnostic and therapeutic challenges. Although recurrence is rare, it is important to continue a follow-up for patients with a history of a cardiac myxoma.

**Keywords:** RVOT; myxoma; obstruction; syncope.

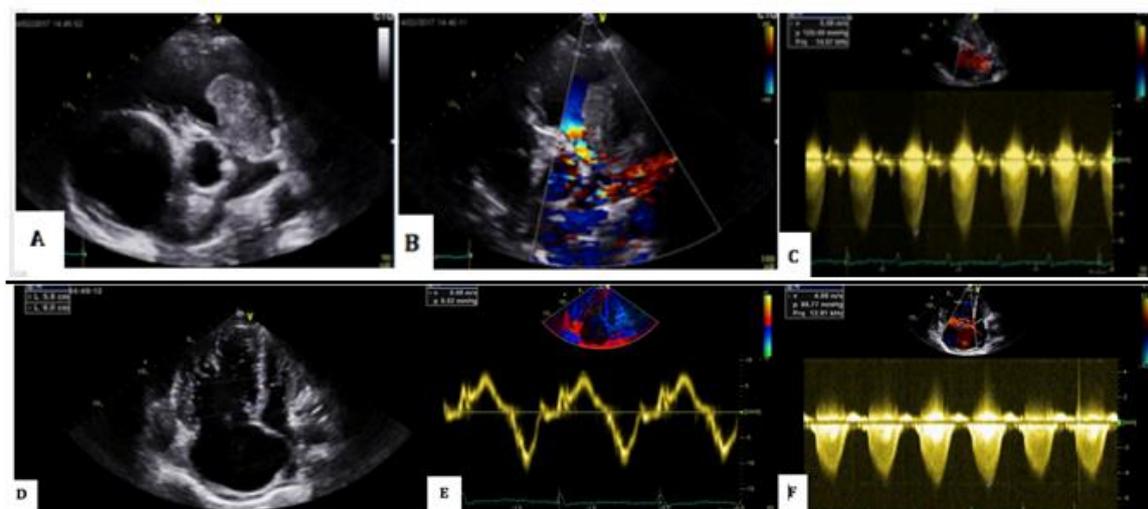
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### INTRODUCTION

In general population, the incidence of cardiac tumors is 0.2% of all tumors [1]. Myxomas are the most common cardiac primary tumors in adult population. The incidence varies from 50-70% of primary cardiac tumors, typically located in the left atrium with an incidence of 70- 80 % then right atrium 10-20% [2]. Only 5% of these tumors are located in the right ventricular [3]. They have been reported cases of ventricular myxoma and rarely valvular myxoma [4-6]. On an histological basis; these are real tumors, derived from multipotent mesenchymal cells of the subendocardium[7, 8]. They have been extremely rare right tract outflow myxomas reported in the literature and if so they generally were found in children and adolescents. These rare myxomas in the right ventricular are capable of generating multiples complications such as pulmonary emboli, arrhythmia and sudden death. If causing a right outflow tract obstruction an immediate surgery is needed after the diagnosis .We present a young man with a myxoma in the right outflow tract presenting dyspnea, without any significant hemodynamic compromise.

### CASE DESCRIPTION

We present the case of our young 30 years old male patient with no medical history nor family myxoma that was referred to our department in cardiology B for the exploration of a fatigue and dyspnea. On admission; he was hemodynamically stable with 80 pulses per minute and 110-60 mmhg on his blood pressure. In the physical exam, he had no signs of congestive heart failure. We discovered a 4/6 grade systolic ejection murmur at the left upper sternal border. His electrocardiogram showed a right ventricular hypertrophy and right axis deviation. Chest X-ray was normal and blood samples didn't reveal any abnormalities. Transthoracic echocardiography revealed a mass located in the right ventricular outflow tract (RVOT) measuring 4cm/3cm, mobile and homogenous. The pulmonary valve seemed spared .The pressure gradient between RV and pulmonary was 120mmhg. We found a dilatation of the right ventricular and the right atrium associated with a RV dysfunction. There was also a mild tricuspid regurgitation with right ventricular systolic pressure at 90mmhg (figure 1).



**Fig-1: Transthoracic echocardiography. Parasternal short-axis view in the same level of the aortic valve showing a large right ventricular outflow tract mass (myxoma) (panel A). This mass causes obstruction demonstrated in the same view by color Doppler (panel B) and the presence of a gradient 120 mmHg (panel C). Four chamber view showing an enlargement of the right ventricle (panel D) and systolic dysfunction of the right ventricular (panel E) and the presence of a tricuspid insufficiency associated with pulmonary artery pressure at 99 mmHg**



**Fig-2: CT scan image showing the right ventricular outflow tract myxoma**

The patient underwent a computed tomography scan (figure 2) and was transferred to surgery. The mass was excised, originating below the pulmonary valve and attached to the RVOT. The mass was completely removed. The diagnosis of myxoma was confirmed by histology. The postoperative period was good without any complications and the patient was discharged a few days later.

## DISCUSSION

Myxomas are the most common primary tumors, thus, they are benign but sometimes their location can be challenging for the diagnosis and therapy.

Statistically, the incidence of primary cardiac tumors is less than one-tenth of a percent. Whereas, secondary cardiac tumors are more common. The majority of primary cardiac tumors are benign and myxomas are the most common [6]. The location of myxomas varies in literature; a meta-analysis which

reviewed 1029 patients found that 83% of myxomas arise from the left atrial cavity and 12.7% were found within the right atrial cavity. 1–5% originate from the mitral valve leaflets [9].

The most common cardiovascular symptoms associated with myxomas are dyspnea, constitutional symptoms like anemia and asthenia and embolic symptoms that can reveal the myxoma sometimes. An incidental echocardiography can be also a way of diagnosis. There are very few case reports of right ventricular myxoma obstructing the outflow tract in adults and children. Clinical symptoms in this rare localization are variable, depending on the size, location and shape. A myxoma located in the RVOT or artery pulmonary can lead to complications by its localization that can be lethal by sudden death or less pejorative consequence such as syncope and pulmonary embolism. It can also lead to heart failure which shows as dyspnea, peripheral edema, ascites due to RVOT obstruction.

The first exam to run in order to have a diagnosis is transthoracic echocardiography (TTE) which is the gold standard as being non-invasive and giving major information. The TTE gives crucial precisions guiding the preoperative time: the shape, size, location relative to RVOT and pulmonary valve, gradient and other possible localizations it also gives instruction to surgeon toward the best surgical approach. Transesophageal echocardiogram can also help in the diagnosis and provide the same information as TTE with more specific details concerning other localizations. Magnetic resonance imaging, and cardiac gated computed tomography can offer additional precision about the structure and the function of the heart [10].

At the end, it's a histological diagnosis so with the ETT images we can recall all the intra-cardiac masses for instance: lipoma; thrombus other tumors.

Although RVOT and pulmonary artery myxomas are extremely rare, it is a cause of sudden death before the surgery and in the peri-operative time especially in the induction of anesthesia and during the surgery due to the high risk of embolisation and pulmonary obstruction. It can also occur in the manipulation of the tumor, the valves or the heart. Several surgical approaches may be valid depending on the size and localization although, surgeons should always be very gentle to avoid any kind of catastrophe. A follow-up is justified to watch for recurrences.

## CONCLUSION

RVOT myxomas are very rare, the diagnosis can be suspected in a patient presenting dyspnea, constitutional symptoms, or embolism. Although, it's extremely rare, it's urgent to have a diagnosis and as soon as possible a resection to avoid deadly complications. A follow-up to watch for recurrence is necessary.

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