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Untreated Right Ventricle Myxoma with Pericardial Effusion in Young Men: A Case Report

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ABSTRACT

Myxomas are benign cardiac tumors that are mostly found in the left atrium. We present a case of a 20-yearold male patient who presented with worsening dyspnea and signs of right heart failure. The echocardiography shows an untreated giant intracardiac mass in a rare place in the right ventricle, obstructing the blood flow, and massive pericardial effusion. We do a pericardiocentesis first to manage the pericardial effusion. Pericardiocentesis was done in different normal locations due to ascites permagna. Nevertheless, our patient needs emergency surgical excision as soon as possible to prevent sudden death caused by obstruction, embolic, and other complications.

Keywords: Myxoma; pericardial effusion; pericardiocentesis



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Introduction

Cardiac tumors are rare diseases and mostly benign, with a 0.01-0.03% prevalence. Most of them are myxomas and lipomas. Clinical symptoms of cardiac tumors are relatively varied, from a heart murmur to heart failure due to the size of the tumors. The management of these benign tumors depends on clinical symptoms, most of them need a surgical approach. We presented a case of untreated RV myxoma with clinical right heart failure, ascites permagna, and massive pericardial effusion.

Case

A 20-year-old male was admitted due to worsening shortness of breath with low saturation and abdominal distention due to ascites permagna for over 7 months. Other than right heart failure symptoms, the patient was known to have a history of RV myxoma a year ago and was planning to undergo urgent mass extirpation, but the patient refused. Transthoracic echocardiogram (*TTE*) showed that there's an intracardiac mass occupying the whole RV with a diameter of 7.42cm x 5.17 cm, circumferential PE without sign of tamponade.

Besides fluid overload management with Furosemide drip, Pericardiocentesis was also performed via sub costae, and a 5F pig-tail catheter was inserted, guided by ultrasonography, and showed a mass at the right ventricle with massive effusion 2 cm diameter (estimated fluid 500 ml). Aspiration of pericardial effusion showed serous fluid with a total aspiration of 500cc serous fluid. After pericardiocentesis, there are not many changes in the patient's hemodynamics. So, then we refer the patient to a type-A hospital with the expectation the patient could get his definite surgical tumor excision.

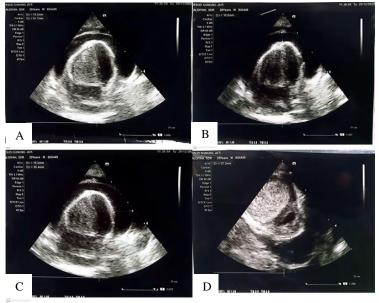


Figure 1. Four chamber views show a tumor in RV and pericardial effusion (1A, 1B, 1C), short axis view shows the tumor compresses the LV and pericardial effusion. (1D)

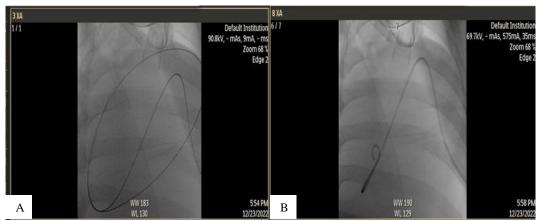


Figure 2. Subcostal sinistra location, angiographic view to know the location of the wire. (2A and 2B)

Discussion

Primary cardiac tumors are rare diseases with an approximate prevalence are 0.01-0,03% in the general population and mostly myxoma affected in middle-aged between the third and sixth decades of life and women with a case ratio of women and men 2.0:1. ^{1,2} Cardiac tumors are benign (75%) half of the tumors are myxoma and the rest are lipomas, papillary fibroelastoma, and rhabdomyoma. ² Myxoma is mostly found in the left atrium (75%) and rarely in the right ventricle (3-4%) and ventricular myxoma has a gross morbidity of less than 0.002% in the general population. ^{3,4} Predilections of myxoma are situated on the border of the fossa ovalis in the interatrial septum or the mitral annulus. Tumor size may vary ranging from 1-15cm with most sizes ranging from 1-6cm, there's no cut-off number for the definition of giant myxoma. Besides the size, myxomas can be solid polypoid and generally pedunculated more compact and have a small chance of embolization. Whereas, in one-third of cases softer papillary myxomas are less compact, and fragile and tend to undergo embolization. Most cases are sporadic, but up to 10% are inherited by the family. ^{1,3,4}

The clinical manifestation of myxomas can be different based on their location and size. It is categorized into classic triad including obstructive, embolic, and constitutional.^{4,5} Left-sided myxoma may lead to pulmonary congestion, unlike right-sided myxoma patients who mostly complain of peripheral edema and ascites. Myxoma can lead to peripheral embolies such as ischemic stroke, visual loss, and renal impairment. This happens mostly in left-sided myxoma, in which the fragile myxoma ruptures and causes peripheral embolism.^{3,4} On the other hand, the right-sided myxoma potentially becomes a pulmonary embolism as a risk from surgery. The risk for tumor embolization of myxoma preand postoperative regards the location of the tumors. There are a typical (tumors that arise from another cardiac chamber beside the left atrium) and typical locations (from the interatrial septum at the border of fossa ovalis in the left atrium) according to a meta-analysis study from Liu *et al* atypical location plays a

significant role.⁶

The constitutional manifestation of myxoma is mostly an inflammation disorder and connective tissue, which is pyrexia, weight loss, malaise, and arthralgia. The underlying conditions happen because the tumor itself releases cytokines IL-6 that circulate systematically and some studies said the amount of IL-6 correlates with the size of the tumor.^{2,7} Our patient shows ascites, lower limb edema, icteric sclera, and dyspnea. Those symptoms match well with right heart failure symptoms, which, in this case, are caused by the tumor's massive size that obstructs the right heart circulation and massive pericardial effusion.

This leads to patient dyspnea, mostly caused by sarcoma and its primary malignant tumors that usually cause rapid growth, local invasion, and hemorrhagic pericardial effusion. These tumors have no stalks and are usually joined in the pericardium, which leads to pericardial effusion. The pericardial cavity contains several lymphatic tissues that originate from the aorta and serve as drainage routes within the pericardium. Effusions may arise as a result of the lymphatic system being compromised by metastatic malignancies. Both acute and chronic pericarditis also contribute to drainage abnormalities in the pericardium and directly irritate pericardial tissue. But, suspicion still cannot be removed before pathologically proven by biopsy, and/or pericardial fluid analysis.

Trans echocardiography (*TEE*) is one of the essential tools, an easy and cheap way to diagnose cardiac masses, the sensitivity rate is 95% for the detection of myxomas, and with transthoracic echocardiography (*TTE*) sensitivity rate of up to 100%.² it gives us information about the size and patient hemodynamics and we need to differentiate it from thrombus, which is commonly by the presence of valvular mitral stenosis, atrial fibrillation, spontaneous echo contrast (*SEC*), and is most frequently found in the left ventricle. On myxoma, characteristics are narrow stalk followed by tumor mobility and distensibility.^{4,9,12} In this case we could not find a thrombus, even though. The definitive diagnosis requires histological confirmation.

Due to our limitations in our hospital, our focus in this admission is to manage the patient's general condition by controlling the fluids excess before having the surgery. We can't rely only upon loop diuretics, since they waste the body's electrolytes and lengthen the hospitalization because of the risks. We believe pericardiocentesis and abdominal paracentesis are the best solutions to relieve the patient's symptoms for now, before performing definite action can urgently be done, which is the tumor excision followed by the biopsy and histopathology analysis. Pericardiocentesis is a technique to drain pericardial fluid, it was a blinded or ECG-guided technique by a subxiphoid approach. But it's no longer safe, therefore. The standard technique is guided by echocardiography or fluoroscopy under local anesthesia. Several entry sites can be chosen according to the localization of the effusion. ^{13–15}

Pericardiocentesis Classical Subxiphoid Approach

Needle is inserted between xiphoid process and costal margin with a 30 to 45° angle towards the left mid-clavicle

Entry points:

A. subxiphoidal

B. apical

C. parasternal

D. dorsal

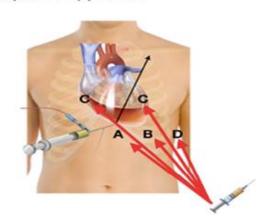


Figure 3 Classic pericardiocentesis by a subxiphoid approach.¹⁶

Our patient needs immediate surgical treatment since the prognosis rate is more than 8% of patients are dying because of obstruction, embolic, and other complications, meanwhile, the surgical mortality is less than 5%.^{3,17} Recurrency rates after surgery are rare. Nevertheless, patients with familial myxoma experience myxoma recurrence, which is more often in young males with multifocal origins or those with a family history of the tumor.^{7,18} For those, routine echocardiography is recommended. There are no guidelines on frequency and duration for monitoring. However, it would be better to monitor once a year and screen the family to find out the possibility of myxoma.²

Conflict of Interest

There is no conflict of interest

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