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Atypical Symptom and Clinical Features of Right Atrial Myxoma: A Case Report

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Abstract: Cardiac tumors are rare in medicine. Myxomas are the most common benign cardiac tumors. Most myxomas (>80%) are found in the left atrium. The site of 15-20% of cases of myxoma is the right atrium. The incidence of cardiac myxoma peaks at 40 to 60 years of age. Most atrial myxomas occur sporadically and are more common in middle-aged women. Patients with atrial myxoma usually have one of the symptoms in the following tetrad: embolic phenomena, intracardiac flow obstruction, arrhythmias and constitutional symptoms. The modality of choice for diagnosis of cardiac myxoma is an Echocardiography. Surgery should be considered promptly, because embolic complications or sudden death can be occured. This surgery is usually safe with low morbidity and mortality. Operative mortality is reported to be within 5%, and will be increased with myxoma occurring in the ventricle. Our case was a 47-year-old Iranian male with right atrial myxoma and a history of heart burn, he does not mention fever and chill, cough, chest pain or dyspnea. In CT Angiography of pulmonary artery showing pulmonary thromboembolism. After echocardiography and pulmonary CT angiography he underwent cardiac surgery and mass with adhesion to the right atrial wall near the IVC was excised. The patient was discharged after surgery with good general condition and stable vital signs.

Keywords: Myxoma, Pulmonary Thromboembolism, Cardiac Tumor

1. Introduction

The majority (>80%) of primary cardiac tumors are benign, and myxoma is by far the most common type [1, 2]. Myxomas constitute approximately 50% of all benign cardiac tumors in adults. Most myxomas (>80%) are found in the left atrium. They are also found in decreasing frequencies in the right atrium, right ventricle, and left ventricle. The incidence of cardiac myxoma peaks at 40 to 60 years of age, with a female to male ratio of approximately 3:1. Most myxomas occur sporadically, but they may be familial. Most patients with myxoma show one of these four conditions: embolic phenomena, intracardiac flow obstruction, arrhythmias and constitutional symptoms [1]. Also it was reported that about 20–30% of patients with atrial myxoma had neurological complications [3]. Dyspnea and right sided heart failure palpitation, atypical chest pain, pulmonary embolism, syncope and hemoptysis are some manifestations of myxoma [6]. Echocardiography is the best diagnostic modality for location and extent of myxoma [6] and Surgery should be considered promptly, because embolic complications or sudden death can be occurred [9].

2. Case Presentation

A 47-year-old Iranian male referred to our hospital for evaluation of heartburn was intensified with activity from a week ago, which was localized and spread to his shoulders. He does not mention fever and chill, cough, chest pain or dyspnea. The patient had no complaints of weight loss, night sweat, palpitation, dizziness, syncope or pre syncope. the patient has hypertension, diabetes mellitus and hyperlipidemia.

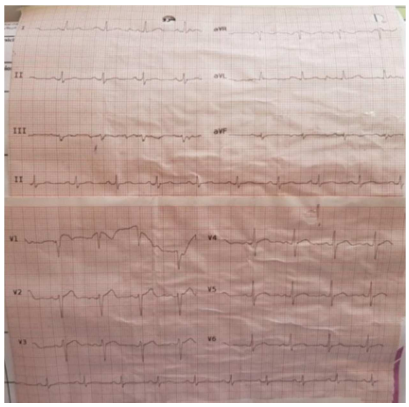
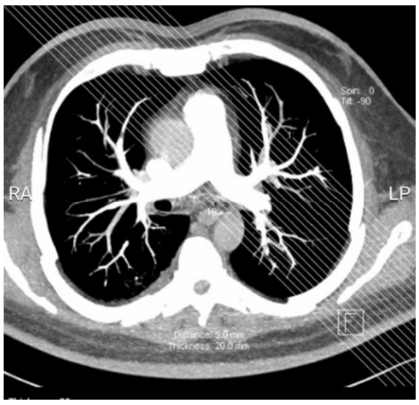
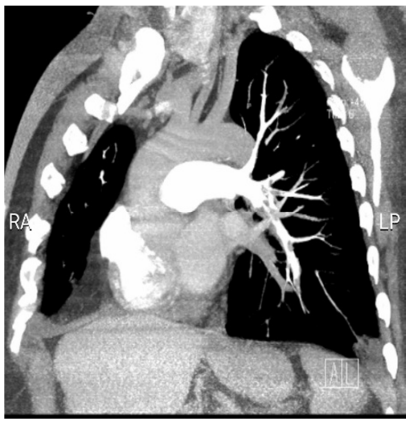


Figure 1. *His electrocardiogram (ECG) showed sinus rhythm without any pathologic changes.*





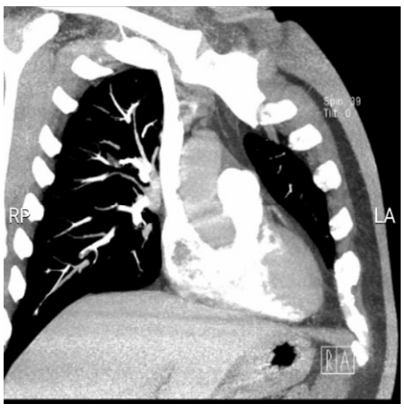
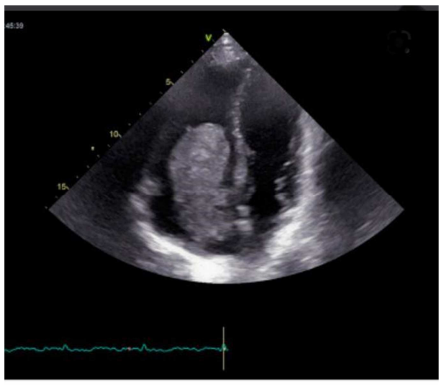


Figure 2. *CT Angiography of pulmonary artery showing filling defect in RLL, LLL and RML artery suggests pulmonary thromboembolism.*



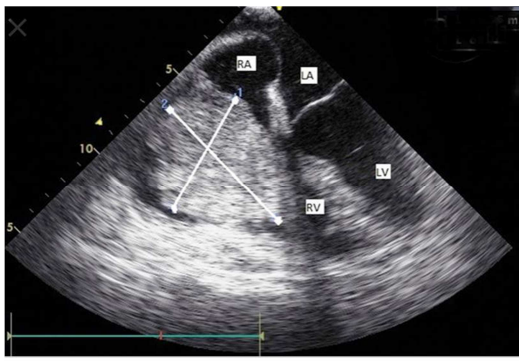


Figure 3. *Echocardiography showed normal LV size and function, mild RV enlargement with mild systolic dysfunction, moderate RA enlargement, mobile multilobulated homogeneous large mass (45*55mm) in RA with out attachment to interatrial septum with protrude to RV in systole, moderate TR, moderate PAH, no PE, no mass in other cardiac chambers.*

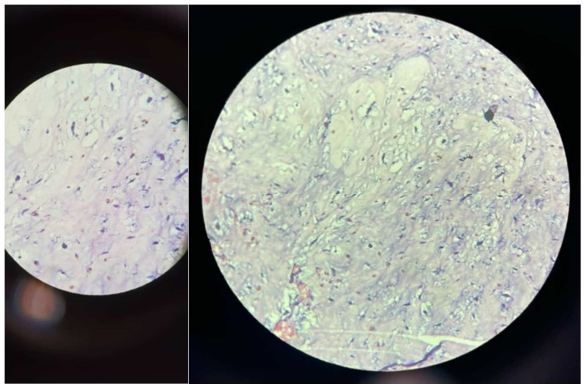


Figure 4. *A very fragile 2*2 cm mass with adhesion to the right atrial wall near the IVC was evident in the surgery report. In histopathology serial section show a benign hypo cellular neoplasm revealing myxoid background and few stellate like cell abundant cytoplasm and indistinct cell border and oval nuclei.*

The patient was discharged after surgery with good general condition and stable vital signs.

3. Discussion

Cardiac tumors are rare in medicine. Primary cardiac tumors are rare findings with an estimated incidence of less than 0.03, 75% of which are benign and 50% of these benign tumors are myxoma [1].

Myxomas are the most common benign cardiac tumors.

These benign tumors mimic the aggressive behavior of metastatic malignant tumors because of embolization that have a poor prognosis in these patients [2].

Myxoma may present with many clinical syndromes such as neurological events and PTE. It was reported that about 20–30% of patients with atrial myxoma had neurological complications [3-18].

The CNS events include progressive headache, nausea or vomiting due to the increase of intracranial pressure, limbs or body weakness or numbness and new onset of seizure [3-19].

A familial cardiac myxoma is rare, approximately <10% of all cardiac myxomas. Syndrome myxoma or Carney's syndrome also consists of extra cardiac myxomas in the skin or brest, spotty pigmentation, and endocrine over activity. Patients with Carney's syndrome are often younger (20 years old) and the cardiac myxomas are usually recurrent and multiple, involving more than 1 cardiac chamber and unusual locations [4-17].

Lipomas, fibromas, hemangiomas, teratomas, and rhabdomyomas are other types of primary benign tumors of the heart [5].

Myxomas most commonly arise from left atrium, usually from a stalk attached to the atrial septum, left atrium (75%), right atrium (up to 20%,) and around 8% in the ventricles [5].

The site of 15-20% of cases of myxoma is the right atrium and Common site of origin is fossa ovalis or base of interatrial septum [6].

Etiology of CM remains unclear but we know that this endocardial-based mass originates from undifferentiated mesenchymal cells [7].

In Histology of mixoma, it consist of an acid-mucopolysaccharide rich stroma. Polygonal cells arranged in a single layer or small clusters are scattered among the matrix [8].

Most atrial myxomas occur sporadically and are more common in middle-aged women [9].

Patients with atrial myxoma usually have one of the symptoms in the following tetrad: embolic phenomena, intracardiac flow obstruction, arrhythmias and constitutional symptoms [1]. Dyspnea and right sided heart failure is the most common manifestation (80%) but patients may also present with palpitation, atypical chest pain, pulmonary embolism, syncope and hemoptysis [6].

The most common first symptom of cardiac myxoma is dyspnea [5].

The modality of choice for diagnosis of cardiac myxoma is an Echocardiography. The sensitivity of the Transthoracic echocardiography for the detection of cardiac myxoma is 93% and Trans esophageal Echocardiogram increases the sensitivity of it to 97% [9]. Echocardiography is the best diagnostic modality for location and extent of myxoma and for detecting the recurrence of it, TTE may not be useful for detection of tumor mass less than 5mm in diameter so TEE is required in small size tumor or in suspicion [6].

TEE is a semi-invasive diagnostic test and its complications is a very low but lethal pulmonary embolisms during the TEE procedure have been reported [10].

Multi detector computed tomography (MSCT) and cardiac magnetic resonance imaging (CMR) are more accurate in determining the relationship of the myxoma to normal intracardiac structures, tumor infiltration into the pericardium, extension to adjacent, and presence of pulmonary arteries emboli and aiding in surgical planning vasculature and mediastinal structures in Compared with echocardiography [11].

Differential diagnoses of cardiac myxoma in pro operative condition pose important clinical implications for appropriate treatment for the underlying diseases. Imaging features could reliably predict primary versus secondary and benign versus malignant cardiac tumors [4].

Surgery should be considered promptly, because embolic complications or sudden death can be occurred. This surgery is usually safe with low morbidity and mortality [9].

Irregular tumor surface, atrial fibrillation, increased tumor size, and increased left atrial diameter are associated with increased risk of embolism in patients with left atrial myxoma [12].

Operative mortality is reported to be within 5%, and will be increased with myxoma occurring in the ventricle [13, 14].

CM surgery and excision was depending from the tumor location. In all patients standard sternotomy and cardiopulmonary bypass (CPB) were established. In addition, in all cases, superior and inferior vena cava cannulation and cold blood cardioplegic heart arrest was applied. In cases with other cardiac pathologies, cardiac myxoma was resected firstly. After 2005, the patients who underwent of CM resection perioperative TEE had used in all cases [15].

The survival after surgery is excellent and recurrence is rare. The risk of recurrence is generally 12%. it is only 1–3% for sporadic tumors and in familial and complex cases is 22% [9].

Resection of CMs by surgery contributes in an excellent prognosis and associated with low complications and recurrences rate [15].

Regular follow-up tests and echocardiography are indicated [9].

Long-term survival after this surgery was excellent and recurrence was rare and surgical method did not affect the outcome [13].

The recurrence rate after this surgery has been reported to be less than 5% [16].

For observations after surgery annual TTE and V/Q (ventilation perfusion scan) scans should be performed to detect the eventual recurrence of new myxomas and pulmonary embolisms. Excision of the recurrent lesions may be the only treatment choice because of the poor effects of chemotherapy and radiation [8-11].

4. Conclusion

These patient present some atypical symptoms such as dyspnea, chest pain, abdominal pain or syncope.

Embolization and cerebrovascular events can be occurred in these cases.

Myxomas are easily misdiagnosed.

In healthy patients presenting with systemic thromboembolism.

Myxomas should always be considered. To prevent further embolization early diagnosis and surgical resection is essential as soon as possible.

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