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RESEARCH ARTICLE

MITRAL STENOSIS PRESENTATION REVEALING ALARGE LEFT ATRIAL MYXOMA

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Abstract

Introduction: Cardiac myxoma (CM) is the most frequent benign primary cardiac tumor. Around 75% of myxomas originate in the left atrium; 20% arise from the right atrium while the remaining 5% are born from both the atria and the ventricle. Atrial myxomas can result in many symptoms: heart failure, systemic or pulmonary embolism, arrhythmias, and conductive disorders, or also constitutional symptoms (such as fever, and weight loss). It largely relies on the tumor's size, location, and architecture. It can obstruct the blood flow, mimicking mitral stenosis. The surgical management is the cornerstone of this entity.

Case Report: A 63-year-old female with a past medical history of diabetes and arterial hypertension was admitted to our department for palpitations and acute heart failure symptoms.The physical examination revealed an irregular pulse with a tachycardia of 150 bpm. The blood pressure was at 118/80 mmHg. It also revealed lower limb edema, lung crackles, and ascites with a mitral stenosis murmur. EKG showed atrial fibrillation at 150HR . Transthoracic echocardiography found dilated atriums with a large mass in the left atrium causing mitral pseudo stenosis .

Discussion: Myxomas are primary cardiac tumors that originate from multipotent mesenchymal cells. They can occur at any age but mostly between 30 and 60 years. The incidence of myxoma in women is 2 to 4 times higher than in men. Clinically, atrial myxomas vary from asymptomatic especially with small tumor size, to non-specific symptoms (dyspnea, lower limb and pulmonary oedema, angina, syncope, and palpitation) and constitutional symptoms such as fever, fatigue, and weight loss. The diagnosis of atrial myxoma based on its presentation is challenging. echocardiography is usually the modality of choice which helps identify the location of the mass whether it' s intra or pericardial, its size, attachment and mobility. Furthermore, it differentiates atrial myxoma from any thrombus or vegetation. Once a presumptive diagnosis of myxoma has been made on imaging studies, prompt resection is required because of the risk of embolization or cardiovascular complications, including sudden death.

Conclusion: Myxoma continues to be a significant challenge for emergency physicians due to its non-specific symptoms and overlap with other conditions. The discrepancy between mild symptoms and the

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Introduction:-

Cardiacmyxoma(CM)isthemostfrequent benign primary cardiac tumor.Around 75% of myxomas originate in theleft atrium; 20% arise from the rightatrium while the remaining 5% are bornfrom both the atria and the ventricle.

Atrial myxomascanresult inmany symptoms: heart failure, systemic or pulmonary embolism,arrhythmias,andconductive disorders, or also constitutional symptoms(suchasfever, and weightloss). Itlargely relies on the tumor's size, location, and architecture. It can obstruct the blood flow, mimicking mitral stenosis.

The surgical management is the cornerstone of this entity.

We hereby present the case of a large left atrium Myxomapresented in a pseudo mitral presentation in a 63-year-old patient.

Case report

A 63-year-old female with a past medicalhistoryofdiabetesand arterialhypertension was admittedto our department for palpitations and acuteheartfailure symptoms.

The physical examination revealedan irregular pulse with a tachycardiaof 130 bpm.The blood pressure was at 118/80 mmHg. It also revealed lowerlimboedema, lungcrackles, and ascites with a mitral stenosis murmur.

EKG showed atrial fibrillation at 150HR (Figure 1).Transthoracic echocardiography founddilatedatriumswitha large massinthe left atrium measuring 44*33mm attached to the inter-atrial septum and causing mitral pseudo stenosis with a mean pressure gradient of 9 mmHg (Figure 2), suggesting moderate mitral stenosis. However, the mitral valve leaflets were intact with minimal mitral regurgitation.

The left ventricle was not dilated with a concentric hypertrophy and preserved function. The Right ventricle was dilated with altered function with high pulmonary arterial hypertension at 63mmhg (Figure 3). The inferior vena cava was dilated with the presence of mild pericardial effusion.

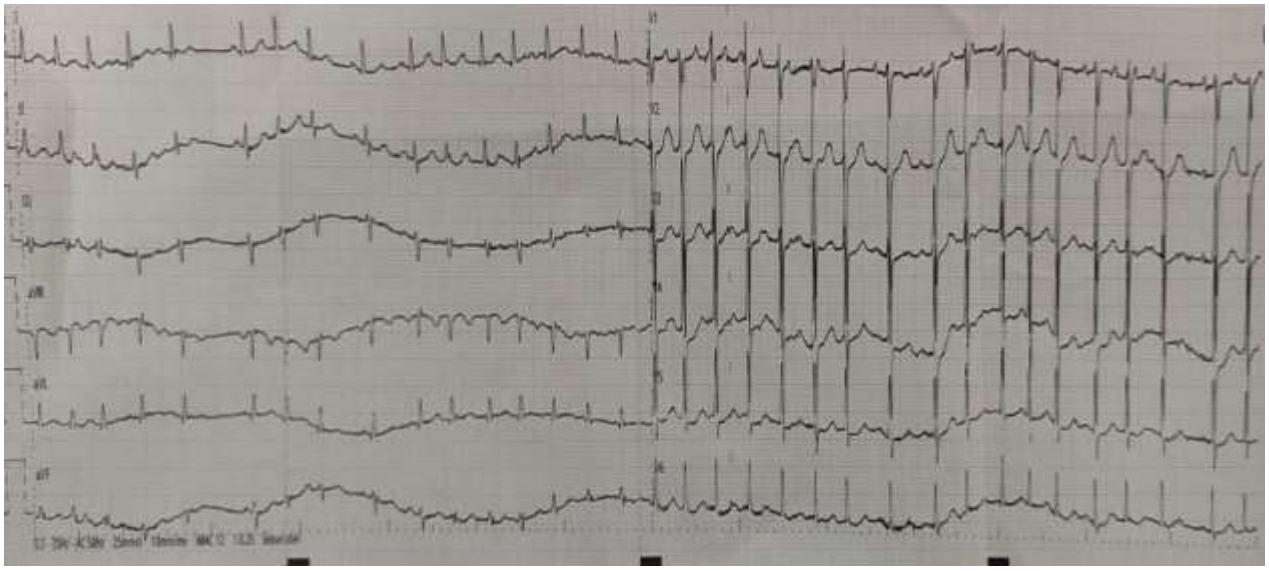

Figure 1:- EKG showing an atrial fibrillation at 150 HR.

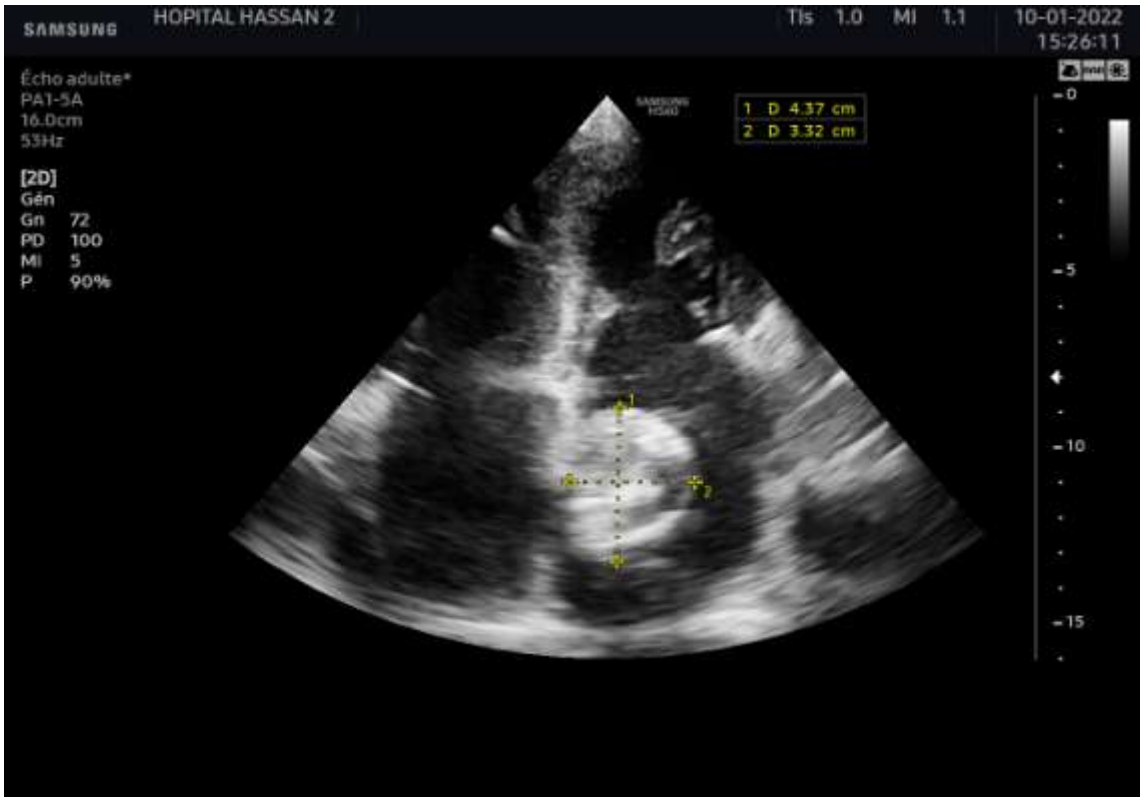

Figure 2:- Apical Four cardiac chambers view showing a large left atrial tumor.

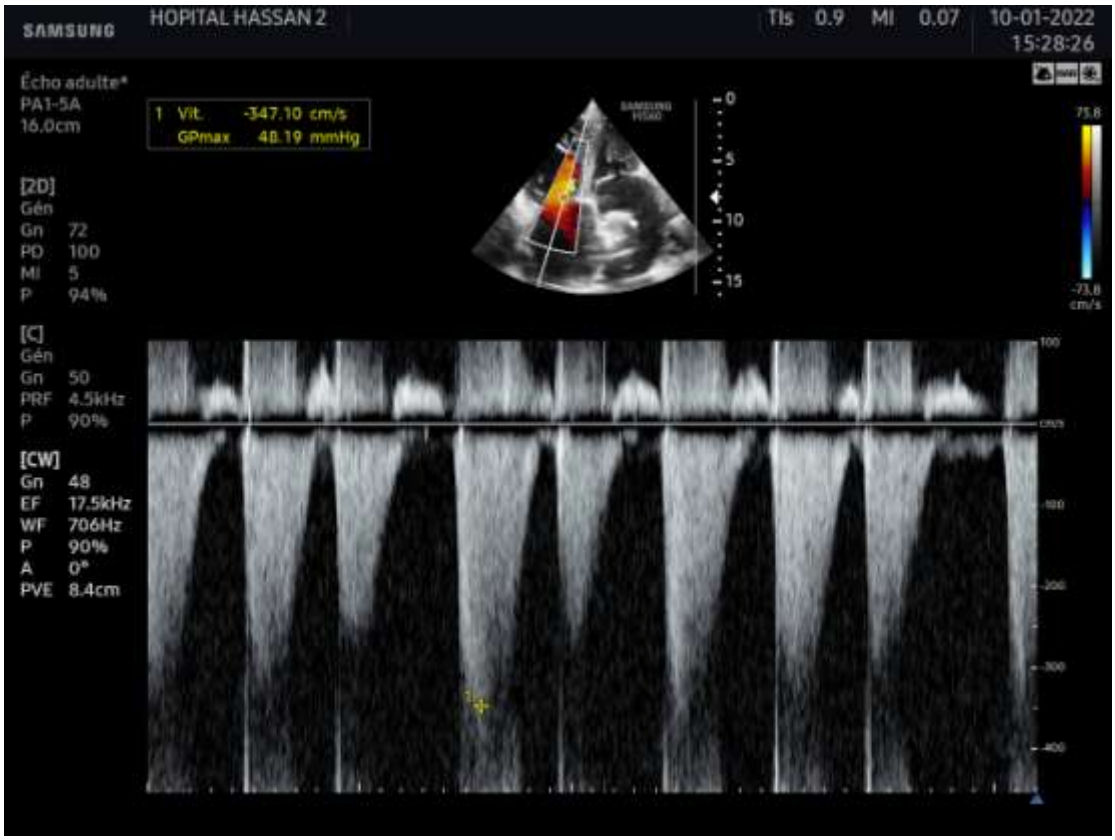

Figure 3:- Continuous wave Doppler showing high pulmonary arterial hypertension

The diagnosis of a probably left atrium myxoma with a functional mitral stenosis was made.

The patient deniedany complementary exploration or surgical option.

Discussion:

Myxomas are primary cardiac tumors that originate from multipotentmesenchymal cells. They can occur at any age but mostly between 30 and 60years. The incidence of myxoma in women is 2 to 4 times higher than in men.

Clinically, atrial myxomas vary from asymptomatic, especially with small tumor size (<4 cm), to non-specific symptoms (dyspnea, lower limb and pulmonary oedema, angina, syncope, and palpitation), and constitutional symptoms (20–60%) such as fever, fatigue, and weight loss. In severe cases, symptoms might include systemic embolization (30–40%), obstructive symptoms (67%) and unexpected sudden cardiac death. (1)

Commonly observed symptoms and signs are dyspnea, orthopnea, paroxysmal nocturnal dyspnea, pulmonary oedema, cough, hemoptysis, oedema, and fatigue. Symptoms may be worsened in certain body positions, due to motion of the tumor within the atrium. On physical examination, a characteristic “tumor plop” may be heard early in diastole [2, 3]. Constitutional symptoms (e.g., fever, weight loss) are seen in around 30 percent of patients. Laboratory abnormalities (e.g., anemia and elevations in the erythrocyte sedimentation rate, C-reactive protein, or globulin level) are present in 35 percent, usually those with systemic symptoms [3].

There are several mechanisms by which cardiac tumors may cause symptoms. The obstruction of the circulation through the heart or heart valves produce symptoms of heart failure. Atrial myxoma may interfere with heart valves causing regurgitation. The direct invasion of the myocardium may result in impaired contractility, arrhythmias, heart block, or pericardial effusion with or without tamponade. The invasion of the adjacent lung may cause pulmonary symptoms and may mimic bronchogenic carcinoma. Finally, left atrial tumors may release tumor fragments or thrombi into the systemic circulation, leading to embolization which is usually systemic but can be pulmonic. The most serious complications of such embolization are neurologic.

Pinede et al. (4) studied 112 cases of left atrial myxoma, examined the tumors macroscopically and correlated the size with the symptoms. The tumor diameter ranged from 1 to 15 cm, and weighed between 15 and 180 g, with a mean of 37g. They discovered a statistically significant association ($P = 0.009$) between having a large atrial myxoma (>5 cm) and developing cardiac symptoms. On the other hand, no significant association was found between the size of myxoma and presence of embolic or systemic symptoms. Paradoxically, larger myxomas went undiagnosed for a longer period of time since the onset of symptoms ($P = 0.004$).

The diagnosis of atrial myxoma based on its presentation is challenging. echocardiography is usually the modality of choice which helps identify the location of the mass whether it’s intra- or pericardial, its size, attachment and mobility. Furthermore, it differentiates atrial myxoma from any thrombus or vegetation. (5)

Transesophageal echocardiography is superior to transthoracic echocardiography in profiling atrial mass.

A study conducted by Shyuetal.(6)where 17 patients were investigated for intracardiac tumor using both TTE and TOE, 14 of whom underwent cardiac surgery, found that TTE had 4 false- positives and 2 false-negatives when compared with TOE which had only one false-positive and no false-negatives. The superior diagnostic utility of TEE is due to the proximity of the esophagus to the heart, the lack of intervening lung and bone, andthe ability to use high-frequency imaging transducers that afford superior spatial.

Other potentially useful diagnostic imaging modalities include computed tomography scan and magnetic resonance imaging.

In our patient, transthoracic echocardiography was sufficient in confirming the diagnosis.

Histopathological examination is considered the definitive confirmatory test for atrial myxomas.

The need for a transvenous biopsy is determined on a case-by-case basis. Generally, noninvasive imaging is sufficient to identify and characterize atrial myxoma. Transvenous biopsy carries the risk of mobilizing emboli associated with the tumor.

Myxomas are generally slow growing cardiac tumors. Studies have made attempts to provide growth rate estimates as shown in Table 1. (7)

Table 1:- Previous studies documenting growth rates of atrial myxomas.

Table 1. Previous studies documenting growth rates of atrial myxomas.

Study	Age, sex	Size on initial echo/cardiac MRI (mm)	Size on ultimate echo (mm)	Growth rate (mm)	Time interval (months)
Alvarez et al.[12]	60, F	Absent	10 × 10	6.6	3
Iga et al.[13]	57, M	15 × 13	38 × 36	1.3 × 1.3	18
Ullah and MaGovern[14]	89, M	140.6	160	0.18	79
Karlof et al.[15]	58, M	Absent	150 × 30	13.6 × 3	11
Marinissen et al.[16]	65, M	No echo done	60 × 40	3.3 ×2.2	18
Roudaut et al.[17]		No echo done	5.05 × 40	6.9 × 5	8
Present case	39, F	Absent	60 × 32	7.5	8

MRI: magnetic resonance imaging; F: female; M: male.

In general, myxoma growth rate estimates are made with the assumption that it develops linearly, while in fact, growth may be exponential.

Once a presumptive diagnosis of myxoma has been made on imaging studies, prompt resection is required because of the risk of embolization or cardiovascular complications, including sudden death.

The results of surgical resection are generally very good, with most series reporting an operative mortality rate under 5 percent (8)

The best technique to excise left atrial myxomas is known as inverted T- shaped biatrial incision, which provides an optimal exposure of all four chambers enabling radical excision and sufficient surgical correction of the mitral valve incompetency, if present. (9)

Cardiac transplantation has been reported for other tumors and might be considered for multiple, recurrent atrial myxomas [9].

The mortality rate from the surgical intervention is low, and the long-term prognosis is satisfactory.

Postoperative recovery is generally rapid. However, atrial arrhythmias or atrioventricular conduction abnormalities were present postoperatively in 26 percent of patients in one series [4]. In addition, patients are at risk for recurrence of the myxoma or the development of additional lesions. In one large series, 5 percent developed recurrent myxoma, suggesting the need for careful follow-up. Development of a second primary myxoma may be more common in patients with a family history of myxoma (10) (11)

Conclusion:-

To conclude, we described a case of a left atrial myxoma. It continues to be a significant challenge for emergency physicians due to its non-specific symptoms and overlap with other conditions. The discrepancy between mild symptoms and the serious consequences the condition carries, necessitates high index of suspicion and thorough investigation to timely diagnose it among a long list of differentials.

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