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RIGHT ATRIAL MYXOMA: A RARE FINDING AND A RARELY MADE DIAGNOSIS: A CASE REPORT

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ABSTRACT **Introduction** Primary intracardiac tumors are rare and approximately 50% are myxomas. The majority of myxomas are located in the left atrium and have variable clinical presentation. We report a case of a large myxoma in the right atrium in a male patient which is uncommon for this gender, which is an uncommon location for this type of tumor.
Case presentation A 55-year-old male presented to Department of Medicine, SGMH Rewa with a history of dyspnea on exertion and generalised body swelling for the last one and a half months. A cardiac evaluation showed muffled heart sounds. He presented with features of right heart failure i.e. pedal edema, ascites with a raised JVP and palpable liver. A provisional diagnosis of COPD with cor pulmonale was made. However an echocardiogram showed a large mass in the right atrium, suggesting RA myxoma.
Conclusions In this case report, we emphasize the rarity of large myxomas in the right atrium in a male patient and the difficulty of differential diagnosis given their dimension and location.

KEYWORDS : Right Atrial Myxoma, Jugular Venous Pressure, Right heart failure

Introduction

Primary intracardiac tumors are rare. Approximately 75% are benign, and approximately 50% are myxomas, which have an incidence of 0.0017% in the general population. Myxomas are usually pedunculated, solitary, and sporadic but may be associated with familial autosomal dominant syndromes (7% of cases) [1].

Myxomas are located in the left atrium in 75% to 80% of cases and are almost always present with signs and symptoms of mitral valve disease or thromboembolic events. The differential diagnosis is performed mainly between rhabdomyoma or thrombus [2].

They may arise in other locations, such as the right atrium (RA) (18% of cases) [3], and, more rarely, in the aorta, pulmonary artery, ventricles, vena cava, or even other organs [1].

Histologically, these are real tumors, derived from multipotent mesenchymal cells of the subendocardium [2,4,5,6].

Cardiac tumors represent 0.2% of all tumors found in humans [5]. These tumors are divided into primary and secondary or metastatic. Secondary or metastatic tumors are 20 to 40 times more frequent than primary tumors. Myxomas affect patients within a wide age range (15 to 80 years), and the average age is approximately 50 years. There is a female predominance in the sporadic form [7].

According to the size, mobility, and location of the tumor as well as physical activity and body position, patients' symptoms may have an asymptomatic course or progress with thromboembolic events that may even lead to sudden death [4, 5, 8].

The classic triad found in patients with cardiac myxoma is characterized by obstruction of blood flow, constitutional symptoms, and thromboembolic events [4, 5, 7]. The obstruction of blood flow leads to intermittent heart failure, and, similar to systemic non-specific flu-like malaise symptoms, there is usually a low fever of long duration, arthralgia, anorexia, and thromboembolic events [7]. RA myxoma, in particular, can obstruct the tricuspid valve, causing signs and symptoms of right heart failure, peripheral edema, ascites, hepatic congestion, and syncope [7]. Cardiac auscultation in atrial myxomas may vary with the size, location, mobility, and prolapse of the tumor through the atrioventricular valves and even body position, and, therefore, detection of a murmur may or may not occur. An auscultation characteristic of myxoma is the 'tumor plop', which is an onomatopoeic representation of the heart sound caused by the presence of the tumor inside the atrial chamber that occurs in 15% of cases [7].

Routine laboratory assessment may show non-specific changes such as anemia, increased erythrocyte sedimentation rate, increased levels of globulin and C-reactive protein, leukocytosis, thrombocytopenia, and polycythemia [7]. Recent studies suggest that cardiac myxomas produce and release into the circulatory system an interleukin, which may be responsible for inflammatory or autoimmune phenomena.

Although transthoracic echocardiography is less invasive and presents an excellent sensitivity in detecting 95% of myxomas, the sensitivity increases to 100% when followed by transesophageal echocardiogr aphy [7]. Computed tomography (CT) and magnetic resonance imaging may be useful to demonstrate the point of fixation and associated complications. An electrocardiogram may be normal or show unspecific repolarization changes or even arrhythmias or heart block due to direct infiltration of cardiac conduction tissue or by irritating the myocardium itself or both. Chest X-rays and electro cardiograms are non-specific [2].

This case report describes a rare clinical case of a large RA myxoma in a male patient, highlighting the difficulty of the differential diagnosis of this tumor because of the presentation with signs of right heart failure. So it was provisionally diagnosed as a case of COPD Cor pulmonale given the history of chronic ganja smoking. However further evaluation via Echocardiography and Chest CT Scan later revealed the diagnosis to be that of a Right Atrial Myxoma which was causing a right sided heart failure.

Case presentation

A 55-year-old male, smoker presented to Department of Medicine, SGMH Rewa with a history of dyspnea on exertion and generalised body swelling since 45 days, which had become more frequent and intense in the last 14 days causing the patient to be admitted. Patient also had one episode of syncopal attack.

On General examination, patient was having clubbing, bilateral pitting edema and raised jugular venous pressure. Facial puffiness was also present.

Cardiovascular examination revealed that his heart rhythm was regular with muffled heart sounds and no murmurs. Per abdominal findings revealed palpable liver 3 cm below the right sub costal margin. A chest X-ray showed clear lung fields with RA enlargement (Figure 1). An electrocardiogram showed sinus rhythm with right axis deviation with signs of RA enlargement (Figure 2). A transthoracic echocardiogram showed a moving mass in the RA (Figure 3) attached to the atrial wall, with mild pericardial effusion and an increased RA and right ventricular volume overload. A Chest CT Scan (Figure 4) revealed a mass lesion arising from antero-superior wall of right atrium projecting through tricuspid valve into right ventricle measuring 64x61 mm in size. CT findings were suggestive of Right Atrial Myxoma. It also extended into the SVC. There was associated thrombus within it. There was also associated pericardial and bilateral pleural effusion.

An Abdominal ultrasound revealed a mild hepatomegaly and a heterogeneous enhancement of parenchyma associated with hepatic congestion and mild to moderate ascites.

Discussion

Primary cardiac neoplasms are rare and occur with an estimated incidence of 0.0017% to 0.19%, representing less than 5% of all heart tumors [9]. Myxomas are usually polypoid and pedunculated tumors (approximately 83% of cases) [10].

Approximately 70% of affected patients are women [2] predominantly between the third and sixth decades of life [10]. As our case was of 55 year old male which was unusual for this gender.

Myxoma is the most prevalent primary cardiac tumor. The RA is an unusual location and is the site of 15% to 20% of cases of myxoma [1]. A low incidence of RA myxoma has been reported for decades in several series of autopsy cases. Echocardiography remains the best diagnostic method for locating and assessing the extent of myxomas and for detecting their recurrence, with a sensitivity of up to 100%. However, transthoracic echocardiogram may not identify tumors smaller than 5 mm in diameter, and a transesophageal echocardiogram is required when there is suspicion of a very small tumor [11].

RA myxomas may remain asymptomatic [12] or eventually cause constitutional signs and symptoms, including fever, weight loss, arthralgias, Raynaud phenomenon, anemia, hypergammaglobuli nemia and an increased erythrocyte sedimentation rate due to the production of interleukin-6 [10].

In a recent publication reporting 19 years of experience with surgical treatment of primary intracardiac myxoma, seven (17%) cases out of 41 originated from the RA. However, in this series, the mean maximal diameter of the tumors was 5.1 ± 1.8 cm [13]. In our case the tumor size was 6.4 x 6.1 cm.

The signs and symptoms of RA myxomas are atypical and highly variable, depending on the size, position, and mobility of the tumor and are modified according to physical activity and body position of the patient [7]. In our case none of the constitutional symptoms were present making it an even difficult case to diagnose.

Patients may also present with atypical chest pain, syncope, lethargy, malaise, palpitation, peripheral edema, pulmonary embolism, and hemoptysis. However, the most common manifestation is dyspnea (in 80% of patients), and right heart failure has been reported.

Dyspnea on exertion and generalised body swelling along with one episode of syncopal attack were present in our case. Our patient presented with the signs of right heart failure i.e. ascites, bilateral pitting edema in the lower extremities with raised JVP.

In this case, an echocardiogram suggested the diagnosis of RA myxoma. Although echocardiography is the modality of choice for screening cardiac masses, magnetic resonance imaging and CT provide information regarding tissue characteristics and allow an excellent overview of cardiac and paracardiac morphology. CT scan in this case supported the diagnosis of a Right Atrial Myxoma along with the site of attachment and its extension into SVC.

The recurrence rate of sporadic tumors is very low: between 1% and 3% [7]. The operative mortality ranges from 0% to 3% in multiple series [7].

Conclusions

Though a rare location for a large myxoma, the RA should always be considered in the differential diagnosis of a right-sided heart mass, especially when the patient shows signs and symptoms of heart failure with uncertain etiology. The findings in our case report suggest that cardiologists and surgeons need to make an early diagnosis and treat patients with these tumors to improve the prognosis. In our case the signs of right heart failure with a history of smoking were misleading but the probability of a right atrial myxoma should always be considered in a list of differential diagnosis.

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

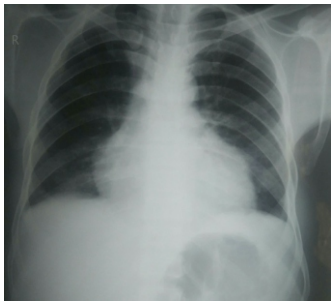

Figure 1: Chest X ray PA View showing Right Atrial Enlargement

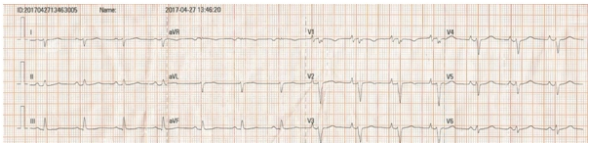

Figure 2: ECG showing sinus rhythm with signs of Right Atrial enlargement with Right axis deviation

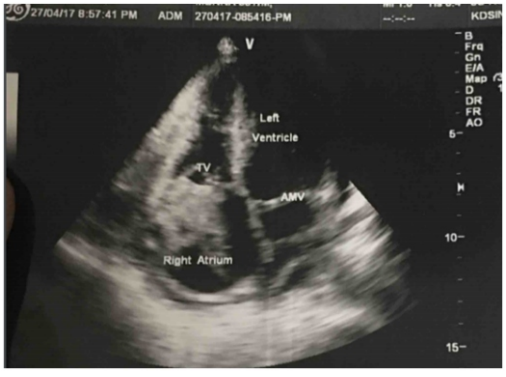

Figure 3: A transthoracic echocardiogram (Apical 4 chambered view) shows a mass in the Right Atrium

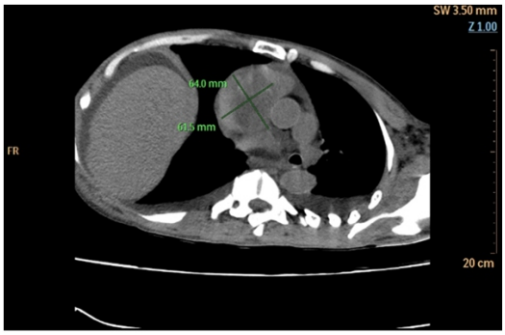

Figure 4: An image of Chest CT scan showing mass lesion in right atrium projecting through tricuspid valve into right ventricle measuring 64x61 mm in size.

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