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ISSN No: 2319-5886

International Journal of Medical Research & Health Sciences, 2018, 7(3): 191-195

Large Right Atrial Myxoma Associated with Unexpected Large Abdominal Ascites

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ABSTRACT

Introduction: *Myxoma accounts for about 40-50% of all primary cardiac tumors. However, the most common site for cardiac myxoma is the left atrium. In this case report, a patient with large right atrial myxoma was diagnosed, which is an unusual site for this tumor type.* ***Case presentation:*** *A 42-year-old woman presented with a history of abdominal ascites in last five months and progressive dyspnea for two weeks duration. A cardiac evaluation showed sinus tachycardia and faint systolic murmur over the left lower sternal area. An echocardiogram revealed a large mass in the right atrium, suggestive of myxoma; our patient underwent surgical excision of a multilobulated mass. Moreover, a histopathological study was done, and the diagnosis was a myxoma.* ***Conclusion:*** *In this case study we are focusing on the importance of atrial myxoma diagnosis in an uncommon location in patients presented with dyspnea and chronic ascites.*

Keywords: Sarcoma, Thrombocytopenia, Heart neoplasms, Echocardiography, Heart auscultation

Abbreviations: CT: Computed Tomography; DTPA: Diethylenetriamine Penta Acetate; ESR: Erythrocyte Sedimentation Rate; IAS: Interatrial Septum; INR: International Normalization Ratio; IVC: Inferior Vena Cava; LA: Left Atrium; MRI: Magmatic Resonance Imaging; PTFE: Polytetrafluoroethylene; RA: Right Atrium; RV: Right Ventricle; SVC: Superior Vena Cava; TEE: Transesophageal Echocardiography; TTE: Transthoracic Echocardiography; TV: Tricuspid Valve

INTRODUCTION

Cardiac tumors represent 0.2% of all tumors found in humans, and myxoma is the most common type [1]. However, 40-50% is the prevalence of myxoma among all primary cardiac tumors; 90% of them are solitary and pediculate. The most common site of occurrence is the left atrium which is represented 75-85%. Only 25% of patients have right atrial myxoma; many of those cases are sporadic with about 10% of them are familial with autosomal dominant inheritance [2].

Moreover, the incidence of biatrial myxoma is less than 2.5% [3]. Besides, it is rarely presents in the ventricles, aorta, vena cava, pulmonary artery and pulmonary veins or even other organs [1,4]. The most common attachment site is in area of fossa ovalis in the left atrium [2].

Myxoma shape can be round, oval, or polypoid; their surface can be lobulated or smooth and are usually have a white, yellowish, or brown color; however, those with a brownish color is the predominant [1,2].

As well as the consistency of such tumors is also variable either firm or sometimes gelatinous [1]. Myxoma microscopically appearance is composed of stellate to plump neoplasm, and cytologically appearance is consist from myxoid stroma with bland mesenchymal cells set in it [5]. In sporadic myxoma cases, 50 years is the mean age of presentation with female predominance [6]. In patients with intracardiac tumor; early diagnosis is so difficult because the symptoms are usually nonspecific [7].

Atrial myxoma patients may be asymptomatic, but, symptomatic cases may present with one of the following triad that include either blood flow obstruction, embolization, or constitution symptoms [8]. Generally, right atrial myxoma differential diagnosis includes thrombus, metastatic tumors, and sarcoma [4].

Additionally, cardiac auscultation of a patient with atrial myxomas is dependent on tumor size, mobility, location and its prolapse through the mitral or tricuspid valve. In 15% of cases what is called 'tumor plop' heard on auscultation; which is distinguishing of myxoma [1].

Investigations in the myxoma cases shows non-specific changes such as anemia, polycythemia, increased erythrocyte sedimentation rate, increase C-reactive protein, leukocytosis, increased levels of globulin and thrombocytopenia [1]. In the same way recent studies propose that interleukin produced and released from cardiac myxomas, which may be responsible for autoimmune or inflammatory effect [1]. Thus interleukin-6 may be elevated and it is significance that it can be used for recurrence detection [2].

Additionally, transesophageal echocardiography has approximately 100% sensitivity for detection of cardiac myxoma. while transthoracic echocardiography has less specificity than the transesophageal echocardiography. Contrast computed tomography scan shows a well-defined intracavitary ovoid or spherical mass. Also, the point of attachment can be visualized by magnetic resonance imaging (MRI) and helps to distinguish a tumor from a thrombus. Particularly with the addition of Gadolinium-DTPA contrast agent [4]. Therefore, when the diagnosis of cardiac myxoma is established, surgical excision should be done, to avoid embolization risk [9]. Additionally, careful tumor handling is mandatory; that to decreases fragmentation possibility and to reduce embolization at time of surgery [1]. Also, precise tumor excision will avoid its recurrence [9].

Moreover, 1-3% is the recurrence rate in patients with sporadic myxomas but 10-20% in patients with familial myxomas. In patients with sporadic myxomas, the most possible cause for recurrence is incomplete excision of tumor. Close clinical follow-up is recommended for patients diagnosed with familial myxomas [5].

CASE PRESENTATION

Forty-two-year-old women presented with a two weeks history of shortness of breath prior to her admission. She had been admitted five months ago with an episode of abdominal ascites, with no other significant past medical history. Cardiovascular examination revealed regular heart rhythm with faint systolic murmur heard over the left lower parasternal area and elevated JVP.

Investigations had shown normal complete blood count, random blood sugar, blood urea and serum creatinine, negative virology screen with elevated total serum bilirubin and alkaline phosphatase, elevated INR and prothrombin time and mild elevation in ESR, a chest X-ray revealed clear lung fields with global cardiomegaly. An electrocardiogram presented sinus tachycardia with right bundle branch block, and a transthoracic echocardiogram showed a large pedunculated movable mass (10 × 7 × 5) cm in the RA attached to the interatrial septum (Figures 1-4), an increased RA, RV chambers size, severe tricuspid regurgitation with mal-coopted tricuspid valve (TV) leaflets, right ventricular volume and pressure overload, impaired RV systolic function and a dilated inferior vena cava and hepatic vein. Subsequent transesophageal echocardiography (TEE) revealed a large mobile right atrial mass attached to the area of fossa ovalis of interatrial septum (Figure 5).

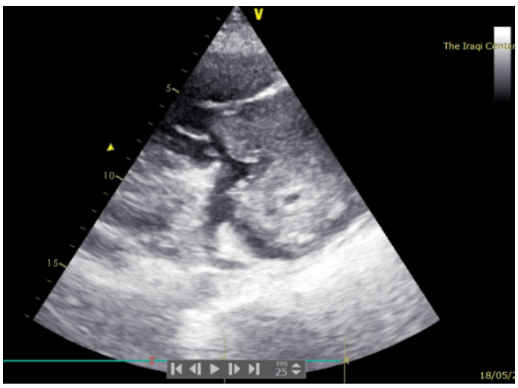

Figure 1 Transthoracic echocardiographic (TTE) image of the right atrial myxoma. The figure shows the tumor in the right atrium (RA) in the parasternal long axis view RV inflow (RA: Right Atrium, RV: Right Ventricle)

Abdominal ultrasound shows moderate to severe degree of ascites, normal liver size, fine irregular outlines, and

multiple hyperechoic masses; a picture suggesting multiple hemangiomas and CT scan of the abdomen and pelvis was done and it shows severe degree of abdominal ascites.

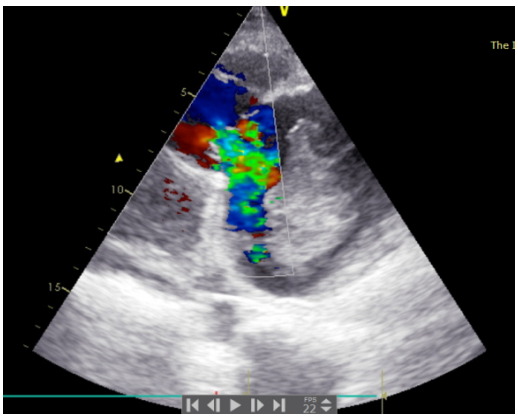

Figure 2 Transthoracic echocardiographic (TTE) image of the right atrial myxoma. The figure shows severe tricuspid regurgitation

Ascitic fluid aspiration had been done and it was sent for histopathological analysis; which shows mixed inflammatory cells, predominantly lymphocytes with reactive and benign looking mesothelial cells.

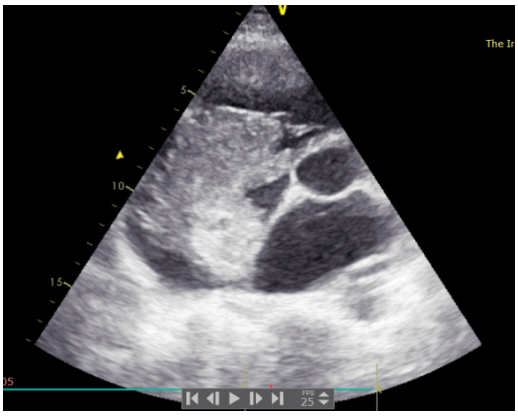

Figure 3 This figure shows the tumor in the(RA) and attached to the interatrial septum in the parasternal short axis view (RA: Right Atrium)

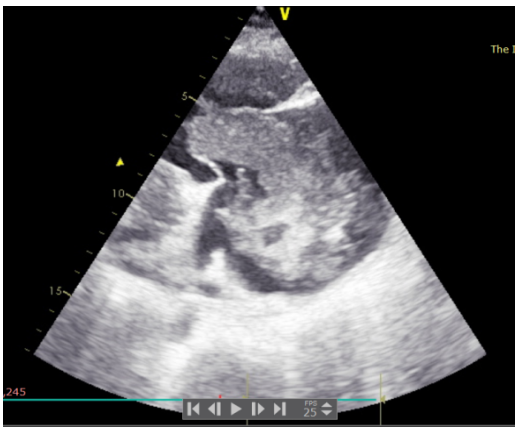

Figure 4 Transthoracic echocardiography TTE showing the mass in the RA and protruding to the RV through the TV. (RA: Right Atrium, RV: Right Ventricle, TV Tricuspid Valve)

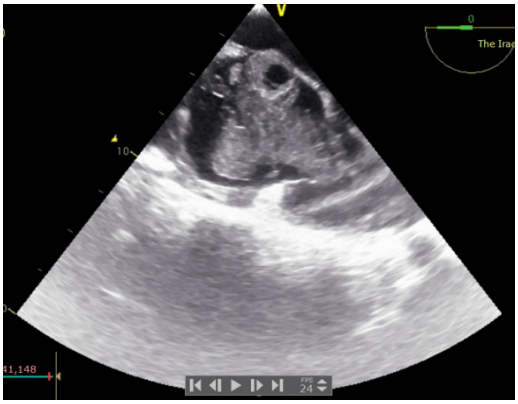

Figure 5 Trans-oesophageal echocardiographic (TOE) image of the right atrial myxoma. The four-chamber view shows the tumor attached to the interatrial septum and prolapsing through the tricuspid valve in diastole

However, after providing informed consent, the patient underwent median sternotomy under general anesthesia, the base of the tumor mass was attached to the center of fossa ovalis, total excision was done with a safe margin of about 2 cm × 3 cm of the apparently (grossly) healthy septum done with closure of the defect by an artificial PTFE patch (Figure 6). Over this time, the resected mass was sent for histological assessment that shown single piece of tissue measured (10 × 7 × 5) cm with section show myxoid tissue and stellate stromal cells a picture consistent with atrial myxoma and no malignant changes was seen.

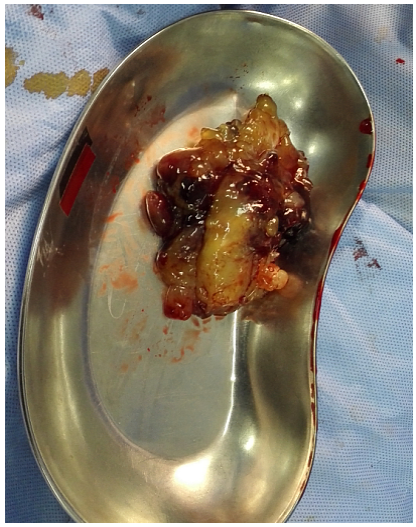

Figure 6 Postoperative excised atrial myxoma

The echocardiogram done in the postoperative period, showed no residual myxoma with an intact interatrial septal patch.

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, myxoma is the most prevalent primary cardiac tumor [1]. However, right atrial myxomas are rare tumors and it accounts for about 15%, and it should be expected in patients, especially women 35 to 55 years old as in this case of the 42-year-old [1]. It is mainly originated from an area in the atrial septum near the fossa ovalis [7] that completely agree with our case as the myxoma originates from inter-atrial septum in the fossa ovalis area. Although symptoms are said to be more variable when a tumor originate in the right side of the heart [1], hence that may explain why our patient major problem was abdominal ascites primarily then with the progression of the condition shortness of breath supervene.

Other symptoms might be occurring like atypical chest pain, syncope, lethargy, malaise, palpitation, peripheral edema, pulmonary embolism, and hemoptysis. However, the most common manifestation is dyspnea that happens in 80% of patients, and right heart failure has been reported [4]. Accordingly, our case presents with shortness of breath which is one of the major causes for patient referral for echocardiography. Figure 3 This figure shows the tumor in the (RA) and attached to the interatrial septum in the parasternal short axis view, (RA: right atrium).

Echocardiography shows a large pedunculated moving mass (10 × 7 × 5) cm in the RA attached to the atrial septum and prolapsing (Figure 4) through TV without causing functional tricuspid stenosis. After providing written informed consent, the patient underwent mass resection surgery under the cardiopulmonary bypass, then resected mass was sent for histological assessment and it results revealed features of benign cardiac myxoma. Post-operative echocardiography had been done to her with no residual mass appear in the right atrium.

CONCLUSION

Although right atrium is a rare location for a large myxoma, so should always kept in mind as one of the differential diagnosis of a right-sided heart mass. The diagnosis of such a large RA myxoma highlights the importance of early detection and removal to avoid subsequent complications.

DECLARATIONS

Competing Interests

The authors declare that they have no competing interests.

Consent

Informed consent was obtained from the patient for publication of this case report and any accompanying images.

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