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Case Report

Atypical size and location of a right atrial myxoma: a case report

Sushil Kumar Singhal*, Palash Aiyer, Vijay Grover, Vijay Kumar Gupta

Department of Cardiothoracic and Vascular Surgery, PGIMER and Dr. Ram Manohar Lohia Hospital, New Delhi, India

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***Correspondence:**
Dr. Sushil Kumar Singhal,
E-mail: drsinghal03@gmail.com

ABSTRACT

Primary intracardiac tumors are rare and approximately 50-55% are myxomas. The majority of myxomas are located in the left atrium. Here We report a case of a large myxoma in the right atrium, which is an uncommon location for this type of tumor who underwent operative intervention with excision of a 9x6 cm multilobulated mass. In this case report, we emphasize the rarity of large myxomas in the right atrium and the difficulty of differential diagnosis given their dimension and location.

Keywords: Right atrium, Myxoma, Uncommon

INTRODUCTION

Primary tumors of the heart are rare. They have been found in only 0.0017% to 0.19% of unselected patients at autopsy.[1,2] Seventy-five percent of these tumors are benign; 50% are myxomas. Of the myxomas, 75% to 80% are located on the left side of the interatrial septum.[3] have variable clinical presentation. We report a case of a large myxoma in the right atrium, which is an uncommon location for this type of tumor who underwent operative intervention with excision of a 9x6 cm multilobulated mass. In this case report, we emphasize the rarity of large myxomas in the right atrium and the difficulty of differential diagnosis given their dimension and location.

CASE REPORT

A 35-year-old woman had a 2-year history of palpitations, which became more frequent and intense in the last two months prior to admission and which were associated with dyspnea on exertion and weight loss of one kg per month. She was on atenolol 25 mg/day. Cardiovascular examination revealed that her heart rhythm was regular with splitting of S1 and no murmurs. The results of an examination of other systems were normal. A chest X-ray showed clear lung fields with no RA enlargement (Figure 1).

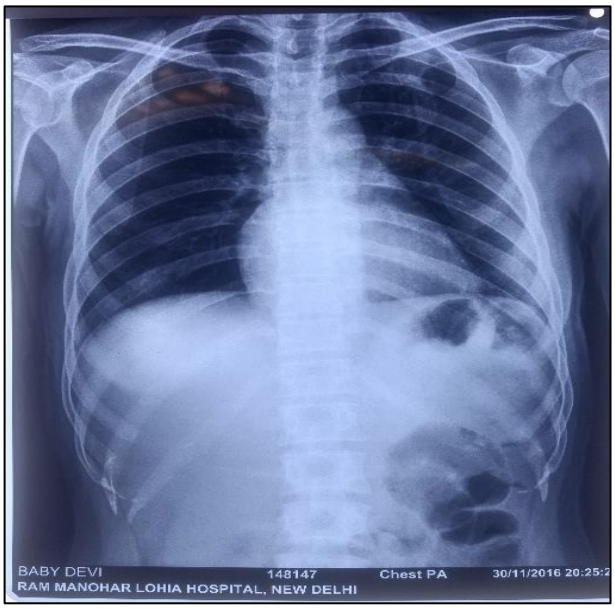

Figure 1: A chest X-ray shows no right atrial enlargement.

An electrocardiogram showed sinus rhythm with right bundle branch block. A transthoracic echocardiogram showed a moving mass 9x6cm in size in the RA, stalk origin from anterior wall of RA near IVC origin. and dilated inferior vena cava. A chest CT scan after intravenous administration of iodinated contrast showed an enlarged cardiac silhouette with an intraatrial hypodensity measuring approximately 9 × 6 cm and avoided filling of the RA with the contrast medium, suggesting the diagnosis of myxoma (Figure 2).

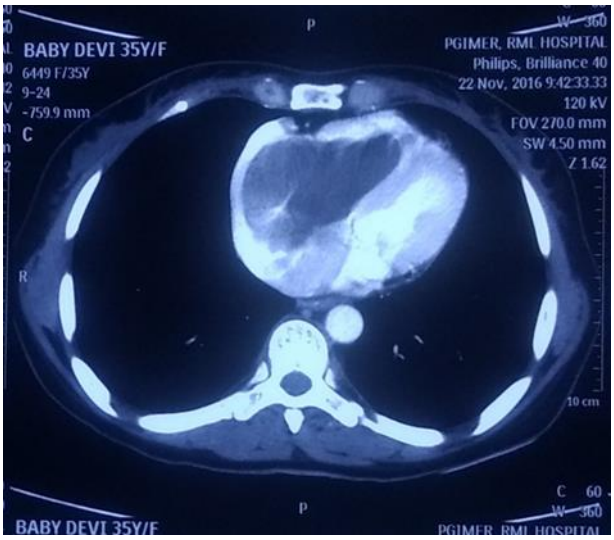

Figure 2 An image of a chest computed tomography scan with intravenous contrast shows an enlarged cardiac silhouette with intra-atrial hypo density.

After providing written informed consent, our patient underwent median sternotomy under general anesthesia. Cardiopulmonary bypass (CPB) was established with conventional mild hypothermia (34.0°C). Cardiomegaly due to RA and right ventricular enlargement was observed. During an anoxic arrest for 31 minutes with single aortic cross-clamping, the tumor was completely excised through a right longitudinal atriotomy.

The tumor was mobile, pedunculated, lobulated, clear, and gelatinous, with implantation at anterior wall of RA near IVC origin. The resected mass was sent for histological assessment. CPB was discontinued without any problems and our patient remained hemodynamically stable. The procedure was concluded in the usual fashion. Our patient was transferred to the intensive care unit (ICU) in stable hemodynamic condition.

Postoperative complications included junctional ectopic tachycardia, which was reversed with intravenous amiodarone. Sinus rhythm was resumed three hours later. Our patient was discharged from the ICU on the first postoperative day and from the ward on the fourth operative day.

Macroscopically, the tumor presented as 9 × 3 × 6 cm lobulated grayish mass weighing 200 g and had an irregular surface and polypoid areas of elastic consistency and spots of loose and friable tissue (Figure 3).

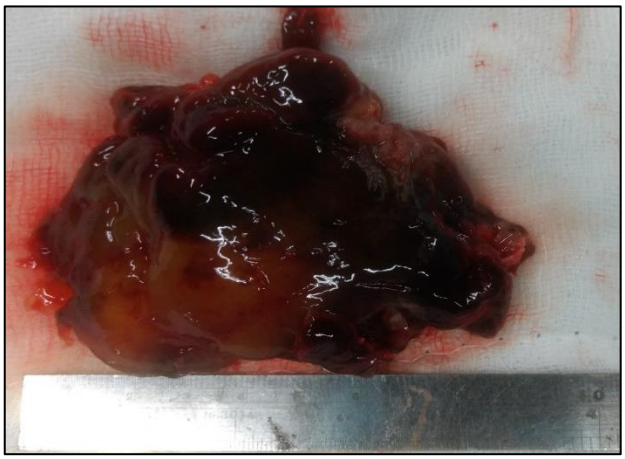

Figure 3: A lobulated mass with an irregular surface, polypoid areas, and spots of loose and friable tissue.

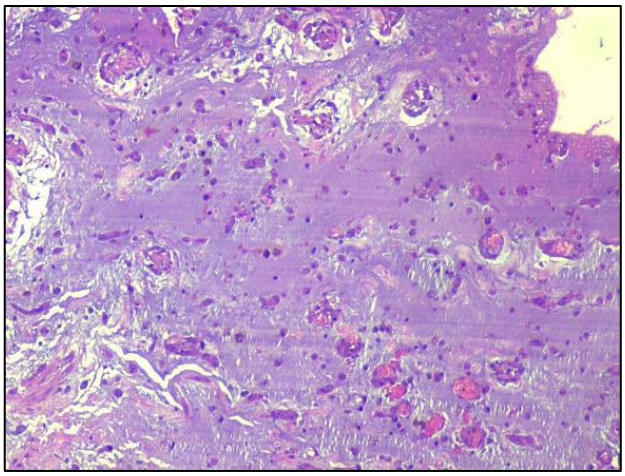

Figure 4: An optical microscopic view of myxoid stroma with stellate cells without necrosis, mitotic activity, atypia, or pleomorphism.

On microscopy, there was abundant myxoid stroma with stellate cells. mitotic activity, atypia, and pleomorphism were not detected (Figure 4).

DISCUSSION

Primary cardiac neoplasms are rare and occur with an estimated incidence of 0.0017%, representing less than 5% of all heart tumors.[4,5] 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.[6] A low incidence of RA myxoma has been reported for decades in several series of autopsy cases. Approximately 70% of affected patients are women, predominantly between the third and sixth decades of life, as was the case of the 35- year-old patient described in this report.[7,8] RA myxomas usually originate in the fossa ovalis or base of the interatrial septum, but in this case, the myxoma

was implanted in the anterior wall of RA near IVC origin.[9] 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.[10]

To the best of our knowledge, our case is one of the largest RA myxomas described in the literature. Although the clinical spectrum can be wide, most affected individuals present with one or more of a triad of symptoms, the so called "myxoma triad", which includes embolic phenomena, intracardiac flow obstruction, and constitutional symptoms.[11]

Nonspecific constitutional symptoms have been reported in 20% to 60% of individuals with cardiac myxomas.[12] Such symptoms may include fever, arthralgia, myalgia, and weight loss.[13] In this report, our patient denied fever, arthralgias, and anemia but complained of weight loss (1 kg in one months) as the only constitutional sign However, the most common manifestation is dyspnea (in 80% of patients), and right heart failure has been reported. Palpitations and dyspnea on exertion were found in this case. 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.[14] In this case, an echocardiogram suggested the hypothesis of RA myxoma, which was confirmed by a histopathological exam. CT chest in this case, showed an enlarged cardiac silhouette with an expansive ovoid mass in the RA with a density lower than that in the heart muscle.[15]

The treatment of choice for myxomas is surgical removal.[16] Adequate excision of the entire mass, along with resection of normal tissue surrounding the base, prevents recurrence.[17] In this case, the tumor was located at the anterior wall of RA near IVC origin. The recurrence rates are noted to be 1-3% in sporadic forms, 12% in familial forms, and 22% in complex myxomas. The operative mortality ranges from 0% to 3% in multiple series.[18] The surgical technique follows the basic concepts of cardiac surgery. However, some aspects should be taken into consideration in the surgical treatment of myxoma.

CONCLUSION

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.

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