

Recurrent Cardiac Myxoma with Unusual Histopathology

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Abstract

Recurrence of cardiac myxoma is rare, seen in about 2-3% of patients, in sporadic cases, although recurrence is frequently seen in familial type.

The Authors report a case of non-familial right atrial myxoma, after successful resection of left atrial myxoma with no recurrence in left atrium. The case is unusual both for its location in the right atrium and histopathology.

Keywords: Myxoma, Recurrence, Right atrial mass.

Introduction

Cardiac myxoma, even though it is benign, they tend to recur. Recurrence is rare in sporadic / non familial type, about 2-3%.^[1] Where as recurrence is frequently seen in familial type. Four possible mechanisms have been put forward to explain recurrence: Inadequate resection, Totipotent multicentricity, Familial type^[2], Metastatic recurrence. Most recurrences are attributed to multicentricity, rather than inadequate resection or seeding of tumor cell during primary resection.^[3,7]

Case Report

A 44 years old female, presented with dyspnea on exertion (NYHA-class III) diagnosed to have left atrial mass measuring 3.5 X 3.0 cm arising from interatrial septum. The mass was excised through median sternotomy, right atrial (transseptal) approach. The defect in the septum was closed with autologous pericardium. Histopathology of the mass showed features of myxoma with free tumor margins. Though the patient was asymptomatic during 18th month follow-up, trans thoracic echocardiogram showed right atrial mass measuring 3.0 cm X 2.5 cm arising from lateral wall of right atrium near inferior vena cava (IVC), right atrial (RA) junction, with no mass in the left atrium. Trans esophageal echocardiogram (Figure 1) confirmed the trans thoracic echocardiogram findings. She had no features of Carney

complex. Her family members were screened. None of them had cardiac tumors or evidence of Carney complex. This time she was re-approached through median sternotomy and through right atrium. Cannulation Strategy: Aortic, Superior vena cava and right femoral vein. Femoral cannula was placed just upto IVC and RA junction. (As the mass was very close to IVC-RA junction, hence IVC was not cannulated through RA) Right atrial mass was excised with wide margins and the defect in the right atrium was closed primarily.



Figure 1: Trans-esophageal echocardiogram (4 chamber view, 0°) showing pedunculated right atrial mass measuring 3.0 cmX2.5cm,

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Figure 2: Right atrial mass measuring 3.4 cm X 2.6 cm, with a stalk (Black arrow)

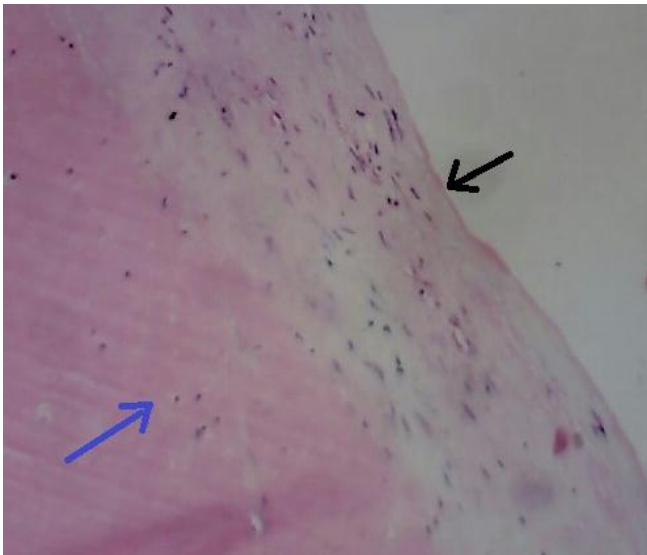


Figure 3: Histopathology: Stellate cells against myxoid background along with blood vessels were seen at the periphery of the lesion suggestive of myxoma (Black arrow). The central, major portion of the tumor is composed of red blood cells, fibrin, haemosiderin laden macrophages and cholesterol clefts, suggestive of an organized thrombus (Blue arrow).

Intraoperative findings: Pedunculated mass measuring 3.5cmX2.4cmX1.5cm was arising from right atrial lateral wall near IVC, RA junction (Figure 2).

Post operative period was un-eventful and she was discharged on fifth postoperative day.

Histopathology findings: Stellate cells against myxoid background along with blood vessels were seen at the periphery of the lesion suggestive of myxoma. The central, major portion of the tumor was composed of red blood cells, fibrin, haemosiderin laden macrophages and cholesterol clefts, suggestive of an organized thrombus (Figure 3).

During subsequent follow-ups there was no recurrence (last follow up was at 30th month, after second surgery).

Discussion

The recurrence rate of sporadic myxoma is 2-3%.^[1] They usually appear during the first four years, although they can appear within few months to several years after surgical excision. The first case of myxoma recurrence was described by Gerbode et al in 1967, several years after surgical resection. At that time incomplete surgical resection was presumed to be the cause. Since then many cases of myxoma recurrences have been reported, not only after extensive resection but also arising in other cardiac chambers.^[7] Four possible mechanism of recurrences are: Inadequate resection, Totipotent multicentricity, Familial type, Metastatic recurrence. Multifocal disease is frequent in familial type.^[2,3] Carney complex^[4] is inherited as autosomal dominant trait, it is a complex with myxomas, spotty skin pigmentation and endocrine over-reactivity.

Recurrence has been associated with abnormal DNA ploidy in upto 40% of the patients.^[5,6] Indeed DNA testing of all patients with cardiac myxoma may prove to be the best predictor of likelihood of recurrence.

The unusual nature of this case lies in its appearance in unusual location in right atrium, rare type considering this as sporadic form, peculiar histopathology i.e., features of myxoma at the periphery of the lesion with major central portion occupied by an organized thrombus.

Question of inadequate resection does not arise in this case as there was no recurrence in the left atrium. In our case possible mechanism of recurrence could be multicentricity or seeding of tumor cells during resection of left atrial myxoma.^[8] However there is little evidence to support the concept of tumor seeding, through deposition of detached fragments carried by a high velocity blood stream.

Acknowledgement:

We sincerely thank Dr. Prabhavathi, (Professor of cardiology) who helped in pre-operative evaluation of this case.

Conflict of Interest: None declared.

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