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Left atrial myxoma with chondroid differentiation – A rare case

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Abstract

Primary cardiac tumors are rare and are seen in 0.3% of cardiac surgeries, amongst these left atrial myxoma is the most commonly seen. 10% of left atrial myxomas are seen in context of Carney syndrome. Cartilaginous differentiation is very uncommon in cardiac myxomas. We aim to present a case of left atrial myxoma in a 50 years old female patient who presented with breathlessness and gradually progressive chest pain since 4 months. Histopathological examination revealed left atrial myoxoma with chondroid differentiation.

Keywords: Myxoma, left atrium, chondroid differentiation.

Introduction

Primary cardiac tumors are rare and account for 0.3% of cardiac surgeries. Amongst these tumors, the commonest is left atrial myxoma, which occur as a solitary mass in the left atrium. They are commonly seen in the age group of 30 to 60 years. 90% of these cases are sporadic while 10% present as a familial disease with Carney syndrome. Chondroid differentiation is very uncommon in left atrial myxoma. We present a case of left atrial myxoma with chondroid differentiation to highlight its rare occurrence.

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Case report

A 50 years old female patient presented with dyspnea on exertion and gradually progressive chest pain since 4 months. The patient had no past history of diabetes mellitus, hypertension or any other major illness. There was no evidence of skin pigmentation or endocrine over activity. Family history was not significant.

Investigation: 2 D echo was done which revealed a large homogeneous mass in the left atrium involving mitral valve opening. It was 36 x 32 cm with left ventricular ejection fraction as 60%. (Fig. 1)

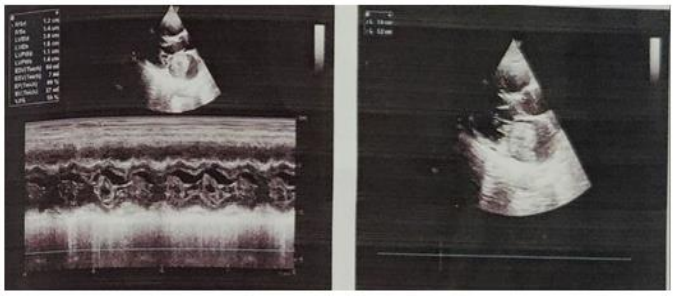

Fig. 1: 2D Echocardiograph showing a large mass in left atrium extending into left pulmonary vein.

Median sternotomy was done and the mass was excised. The tumor was seen in the left atrium and extending into

left superior pulmonary vein. Excised tumor was sent for Histopathological examination.

Histopathological examination

Gross examination revealed a gelatinous, grey white, soft tissue mass measuring 10.5 x 3.5 x 2 cm, cut section was grey white and gelatinous. (Fig. 2)

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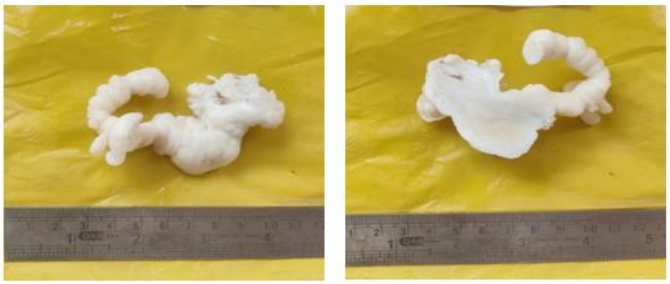

Fig. 2: Gross features: A gelatinous, grey white, soft tissue mass, cut section was grey white, gelatinous.

Microscopic examination revealed a tumor composed of stellate cells with uniform nuclei dispersed in a myxoid stroma. The tumor showed areas of chondroid differentiation. (Fig. 3 and 4)

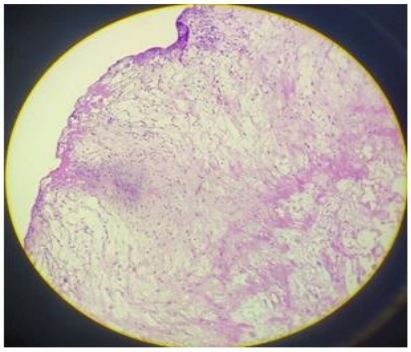

Fig. 3a: Photomicrograph of Left Atrial Myxoma – tumor composed of stellate cells with uniform nuclei dispersed in myxoid stroma, 100x H and E

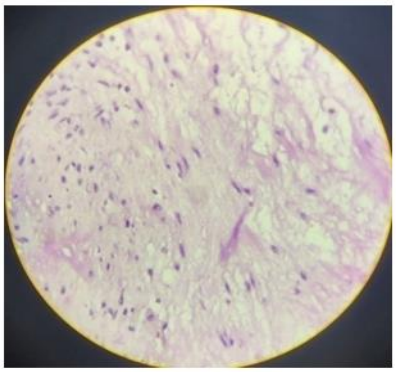

Fig. 3b: Photomicrograph of Left Atrial Myxoma – tumor composed of stellate cells with uniform nuclei dispersed in myxoid stroma, 400x H and E

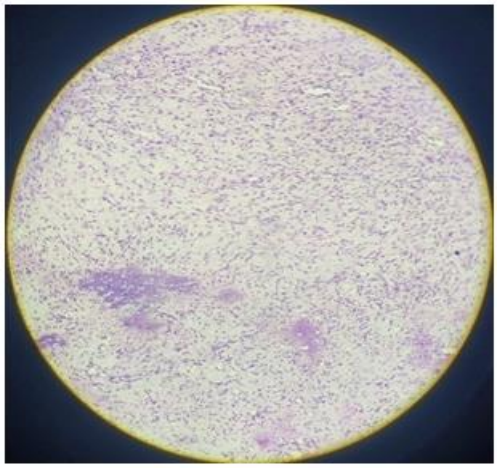

Fig. 4a: Photomicrograph of Left Atrial Myxoma - The tumor shows areas of chondroid differentiation, 100x H and E

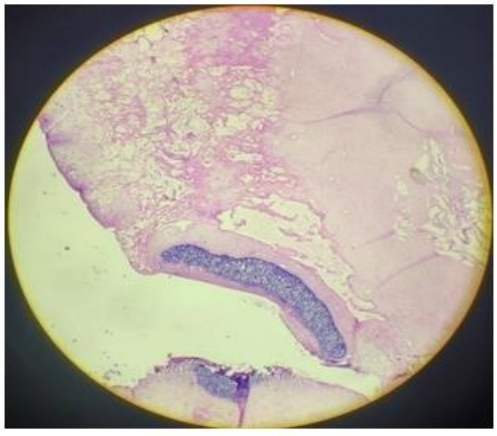

Fig. 4b: Photomicrograph of Left Atrial Myxoma - The tumor shows areas of chondroid differentiation, 400x H and E

Discussion

Primary cardiac tumors are rare with an autopsy incidence of 0.01 to 0.03% [1]. Primary cardiac myxoma accounts for 50% of cardiac tumors; commonest age group is 30 to 60 years [2] and is common in female patients. Cardiac myxoma was first described in 1845.[2] They can be familial or sporadic. Sporadic tumors are seen frequently in middle aged female patients, as seen in our case. Those with Carney complex present in young age and are seen as metacentric tumor masses. They can be seen at sites other than left atrium along with extra cardiac abnormalities such as skin pigmentation, endocrine over activity, schwannomas etc. In sporadic cases, tumor is found in left atrium attached to inter-atrial septum.[3]

Cardiac myxomas have uncertain origin and histogenesis. Many hypotheses suggest an origin from multipotent mesenchyme cells. These tumors are highly vascular and produce growth factors and inflammatory cytokines.[4,5]

Diagnosis of left atrial myxoma is done with 2D echo; MRI may be required to rule out embolism.

Clinical presentation of this tumor depends on size and location. Commonest clinical feature is dyspnea, chest pain, syncope as well as tumor embolization in 30-50% cases. Pedunculated tumor can go through atrio-ventricular valve during systole and cause valvular obstruction, syncope and sudden death. Specific constitutional symptoms are fever, malaise and mediating acute phase response.

Histopathological examination: On gross examination, these tumors present with variable size and are pale, glistening and gelatinous. The tumor can be polypoid or papillary.

Microscopy reveals large stellate and spindle shaped cells separated by myxoid material.[6] Rare microscopic features are calcification in 20%, Gamma Gandy bodies in 17%, ossification in 8%, extra medullary hematopoiesis in 7% cases, chondroid differentiation is very rare.[1,7,8]

Treatment of left atrial myxoma is radical excision. Recurrence has been observed with incomplete excision or in Carney complex.

Conclusion

- Primary cardiac tumours are rare, amongst which cardiac myxomas are seen commonly.
- Chondroid differentiation in a cardiac myxoma is a rare occurrence which can be misdiagnosed as chondrosarcoma.
- Histopathological examination gives definite diagnosis of this tumour.

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