

Left Atrial Myxoma: A Rare Case

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Abstract: Primary cardiac tumours are rare, having an autopsy incidence as 0.001% to 0.03%. 75% of these tumours are benign and nearly half of benign tumours are myxomas which frequently cause systemic problems. We present a case of left atrial myxoma in a 40 year female patient complaining of breathlessness and non-radiating gradually progressive chest pain since 6 months. The 2D- ECHO revealed pedunculated mobile homogenous mass arising from fossa ovalis and protruding into left atrium. The tumour was excised; the gross examination revealed a gelatinous mass which was diagnosed histologically as left atrial myxoma. We present this case in view of its rarity and to highlight the clinico-pathological features of cardiac myxomas.

Keywords: Myxoma, Left atrium

INTRODUCTION

Primary cardiac tumours are rare and account for about 0.3% of cardiac surgeries. Amongst these, 50% tumours are cardiac myxomas and are commonly seen in women in the age group of 30-60 years [1]. Cardiac myxomas are commonly located in the left atrium and present clinically with intra-cardiac obstruction, embolism or non-specific constitutional symptoms [2].

CASE REPORT

A 40 year old female presented with breathlessness and non-radiating, gradually progressive chest pain since 6 months. The patient had no past history of diabetes mellitus, hypertension or any other major illness.

Investigations

Hb and ESR: within normal limits.

2D-ECHO: Intra-cavitary pedunculated mobile homogenous echogenic mass measuring 5.6 x 3.6 cm arising from fossa ovalis of inter-atrial septum protruding into left atrium without causing mitral stenosis.

The mass was excised with pedicle and sent for histopathological examination.

Histopathological Examination

Gross: Received a polypoidal friable gelatinous whitish soft tissue mass measuring 5.5 X 4 X 2 cm. Cut section was grey-white gelatinous with multiple areas of haemorrhages (Fig. 1).



Fig. 1: Gross examination: A gelatinous polypoid soft tissue mass

Microscopy revealed a polypoidal tumour covered by a single layer of flat endothelial cells enclosing stellate and polygonal cells with uniform nuclei set in a myxoid stroma. Stroma revealed areas of haemorrhages (Fig. 2).

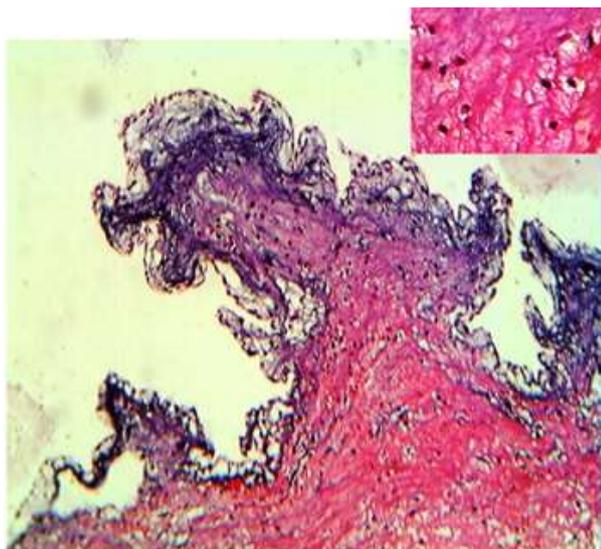


Fig. 2: Photomicrograph reveals a polypoid mass (H&E 100X) composed of stellate cells in myxoid stroma (inset-H&E 400X)

Considering these histological features, the tumour was diagnosed as 'Left Atrial Myxoma' (sporadic).

DISCUSSION

Cardiac tumours are rare, amongst these primary cardiac tumours are even rarer with an autopsy incidence of 0.001-0.03% [3]. The incidence amongst cardiac surgeries is 0.3%. Cardiac myxomas account for 50% of primary cardiac tumours and is commonly seen in women of 30-60 years age group [1, 4]. The typical location is left atrium attached to inter-atrial septum. Similar findings were noted in our case. Myxomas arising from other sites are referred as 'atypical myxomas' [5]. Cardiac myxoma was first described in 1845[4].

The myxomas can be familial (4.5-10%) or sporadic [1]. The familial tumours are seen in young males with multicentric tumour masses and can affect sites other than left atrium alongwith extra-cardiac abnormalities as a part of Carneys complex. Sporadic tumours are seen commonly in middle aged female patients affecting left atrium without extra-cardiac manifestation [6]. Presentation in our case was sporadic in nature.

Cardiac myxomas have uncertain histogenesis. Ultra-structural studies have postulated that cardiac myxomas develop from multipotent mesenchymal stem cells [2, 6].

Clinical Features are due to flow obstruction (palpitations, shortness of breath, syncope, pedal oedema); systemic symptoms (fever, fatigue, anaemia, raised ESR) and symptoms of arterial embolism [1].

The clinical features in our case were due to obstruction with no evidence of systemic symptoms or embolism.

Diagnosis

These tumours are diagnosed with echocardiography, MRI is required to rule out embolism.

Differential diagnosis of cardiac myxomas include organising thrombus, primary or metastatic sarcomas such as low-grade fibromyxoid sarcomas, inflammatory myofibroblastic tumour, myxoid leiomyosarcoma, angiosarcoma and rheumatic valve disease [2, 7].

Histopathological features

On gross examination, these tumours vary in diameter from 0.4-8 cm; are pale glistening gelatinous and can be polypoid or papillary. Polypoid lesions cause obstructive symptoms while papillary tumours present with embolic episodes. Microscopy reveals large stellate or spindle shaped cells separated by myxoid material [2, 7, 9]. Our case revealed a polypoid tumour and presented with obstructive symptoms. On immunohistochemistry, the tumour cells are positive for vimentin and CD 34 [1].

Management

The tumour should be excised promptly under direct vision with the aid of cardiopulmonary bypass [7].

Prognosis

The long term prognosis is excellent. Multiple lesions or familial myxomas are likely to relapse and require follow-up with serial echocardiography [1, 8].

CONCLUSION

- Primary cardiac tumours are rare, amongst which cardiac myxomas are most common and have uncertain histogenesis.
- Echocardiography is highly sensitive for pre-operative diagnosis.
- These tumours present with vague clinical symptoms, hence may be misdiagnosed clinically.
- Clinician should be aware of this rare tumour which is completely curable with surgical excision.

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