Introduction

Cardiac myxomas are the commonest benign tumours of the heart in adults.

The incidence of primary tumours of the heart is between 0.0017 and 0.19 percent in unselected patients at autopsy. About three quarters of these tumors are benign and of these, nearly half of them are myxomas in adults.

Left atrial myxoma was first described in 1845 and the first successful excision of a left atrial myxoma was reported in 1955 (1).

We report a case of left atrial myxoma excised successfully, showing a rare histological finding of Gamna Gandy bodies.

Case Report

A 45-year old woman presented with few months history of shortness of breath on exertion, unassociated with chest pain, sweating, syncopial attacks, weight loss and skin lesions.

Examination revealed a mid diastolic murmur over the apex of the heart.

Echocardiogram demonstrated a 7.4 x 4.4 cm pendunculated mobile mass in the left atrium attached to the inter-atrial septum at the fossa ovalis. The mass was seen prolapsing into the mitral valve reaching the middle of the left ventricle. Minimal posterior pericardial effusion was present.

Laboratory investigations revealed anemia (Haemoglobin of 9.9g/dl), leucocytosis (WBC Count 23.6 x 10^9/L) elevated ESR of 31mm/hr, raised C-reactive protein (33.4 mg/l) and hypergammaglobulinemia (42g/L).

The mass was excised with a 3 cm margin of the surrounding septum and sent for histopathological examination. The patient made an uneventful recovery.

Pathological Findings

The tumour was a firm well-circumscribed globular pale grey mass with a short pedicle and a smooth external surface that had hemorrhagic patches.

It measured 8 x 4 x 3 cm in dimensions. A small piece of the atrial wall was attached near the pedicle. The cut surface was gelatinous with conspicuous areas of hemorrhage (Fig.1).

Fig. 1: Gross Specimen of left atrial myxoma showing a globular mass with smooth surface and hemorrhagic discoloration.

Abstract

Primary benign tumours of the heart are rare, with myxomas being the commonest. They are neoplasms of endocardial origin and generally appear to grow rapidly. The presence of connective tissue fibers encrusted with iron and calcium known as Gamna Gandy bodies may indicate a longer duration of tumor.

We present a case of cardiac myxoma with Gamna Gandy bodies, which is an infrequent finding, and briefly review relevant literature.

Key words

Heart, myxoma, Gamna Gandy bodies.
Microscopic examination showed a vascularised myxoid matrix. Fusiform or stellate ‘myxoma cells’ with eosinophilic cytoplasm and indistinct nucleoli were scattered within the matrix. These cells were arranged singly and as complex interlacing networks around blood vessels and surrounded by a halo of myxoid matrix (Fig.2)

![Fig.2: Microscopic section showing the complex interlacing networks of myxoma cells around capillaries (Haematoxylin-eosin, original magnification x 40)](image)

Irregular dark brown fibrotic structures (Gamma Gandy bodies) were also observed. Special stains (Von Kossa and Perls stain) confirmed that these were encrusted with calcium and iron. These were especially seen around areas of haemorrhage (Fig. 3)

![Fig. 3: Microscopic section showing Gamma Gandy body within the stroma of the myxoma (Haematoxylin – eosin, original magnification x 20)](image)

Interspersed plasma cells, lymphocytes and macrophages were also noted.

Immunohistochemical stain for calretinin was strongly positive in tumor cells.

The resection margin was free of tumor.

The above described features were consistent with a cardiac myxoma with Gamma Gandy bodies.

**Discussion**

Primary tumours of the heart are rare, 75% of them are benign.

Myxomas comprise nearly 50 percent of the cardiac benign neoplasms, followed in frequency by rhabdomyomas, lipomas and fibroelastomas. Less frequently seen are fibromas, haemangiomas, teratomas and mesotheliomas of the AV node.

A search of the records of the Pathology department at Royal Hospital, Muscat yielded 10 benign cardiac tumours in a 10-year period from January 1997 to April 2007, of these 8 were atrial myxomas and 2 were rhabdomyomas.

The age of our patients with myxoma ranged from 22 years to 57 years, while the age of both the patients with rhabdomyomas was 3 years.

A female preponderance is noted in myxomas (2:1) which was also seen in our cases with 5 females and 3 male patients.

The majority of the cardiac myxomas are sporadic. Approximately 7 percent are familial and are a component of a complex hereditary syndrome affecting multiple organs, the best described of which is the Carney Complex. It is an autosomal dominant disease with a mutation of the PRKAR 1 alpha gene located on the long arm of chromosome 17. It is characterized by cutaneous spotty pigmentation, cutaneous and cardiac myxomas, non-myxomatous intra-cardiac tumours and endocrinopathies. Unlike the non-syndromic myxomas, it occurs equally in both sexes, is often bilateral, multi-centric and recurs frequently.

The clinical presentation of atrial myxoma often comprises a diagnostic triad of embolism, intra-cardiac obstruction and constitutional symptoms.
Our patient did not have any signs of embolism or any history suggestive of embolic episodes.

Intra-cardiac obstruction depends on the size and mobility of the tumour with obstruction of ventricular filling and subsequent dyspnea, pulmonary edema and right heart failure. Dyspnea was the presenting symptom of our patient.

Large tumours may damage the valves resulting in mitral or tricuspid insufficiency.

Constitutional symptoms such as fatigue, fever, erythematous rash, arthralgia, myalgia and weight loss have been described which may be manifested by interleukin-6 produced by the myxoma itself.

Laboratory abnormalities observed include anemia, elevated ESR, elevated C-reactive protein and hypergamma-globulinemia \(^6\). The anemia is usually normocytic normochromic but hemolytic anemia due to mechanical causes may be seen. In our patient anemia, leucocytosis, raised ESR and C-reactive protein as well as hyper gammaglobulinemia was noted.

Histopathologically, the overall picture of a cardiac myxoma is dominated by myxomatous matrix and a dispersed cellular component. Various types of cells are seen, the principal type is the ‘myxoma cell’, which is the true neoplastic cell. It may be elongated, fusiform, stellate or polyhedral (the latter is also referred to as ‘lepedic cell’ \(^7\) derived from the Greek word ‘lepis’ meaning scales, based on the fanciful resemblance to the scales on butterfly wings).

The cells are derived from the primitive subendocardial cells, which may show capacity for CD-31 positive endothelial differentiation \(^8\). Initially when described by Orr \(^9\) in 1942 these cells were considered to be of endocardial origin.

At the ultrastructural level, the typical myxoma cell is characterized by sparse intracytoplasmic organelles.

The myxoma cells may be arranged in various patterns, a common variety exhibiting perivascular arrangement as in our case. Five percent of the cases may show glandular patterns \(^10\). Areas of hemorrhages with hemosiderophages are common.

However, the formation of nodules of Gammagandy bodies, which are connective tissue fibres, encrusted with iron and calcium is rare. Trotter et al \(^11\) reported in 1990 4 such cases. Our case also showed Gammagandy bodies.

Hemorrhage and long duration of tumour are factors implicated in the development of Gammagandy nodules \(^11, 12\).

Inflammatory cells, cyst like areas subsequent to liquefaction of the matrix, calcification and fibrinoid degeneration following hemorrhage are other features that may be seen in the myxomas.

Immunohistochemical study reveals strong calretinin positivity in 73-100 percent of cases \(^13\), weak positivity for CD-15 and negative reaction with CEA. However, glandular element if present may express CEA antigens as well as cytokeratin and EMA positive reaction \(^14\).

The Calretinin stain was strongly positive in our case.

**Conclusion**

We have presented a case of left atrial myxoma with typical clinical presentation but with an infrequent histological finding of Gammagandy bodies in an otherwise typical picture of cardiac myxoma.

Hemorrhage and a long duration of the tumour is speculated to result in the formation of Gammagandy bodies, but unfortunately it is usually not possible to ascertain the likely age of the tumour from the symptomatology.
References

6. Mendoza CE, Rosado MF, Bernal L. The role of Interleukine-6 in cases of Cardiac myxoma. Clinical features, immunological abnormalities and a possible role in recurrence. *Tex Heart Inst J* 2001; 28(1); 3-7.