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Clinicopathological Study of Cardiac Myxomas: A case series of three years' experience with seven cases

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Abstract

Background: Primary tumors of the heart are extremely rare, with an estimated incidence ranging from 0.0017% to 0.33% at autopsy. (1) Cardiac myxomas represent the most frequent benign cardiac tumors. In most surgical series, they account for almost 80% of cases. (2)

Objectives: The aim of the present study was to analyze clinical and histopathological features of cardiac myxomas. The relationship between the pathological characteristics and clinical behaviors of myxomas was also studied.

Methodology: This is a retrospective hospital-based study. Histopathological samples of cardiac tumors received for a duration of three years were included in study. The clinical history including electrocardiography and echocardiographic findings were noted. All the samples were examined grossly and microscopically. Histopathological findings were noted and correlated with clinical findings. Data was analyzed by excel spread sheet; results were documented in proportions and percentages with appropriate statistical tests.

Results: The incidence of myxomas in the present series was low. Out of a total of seven cases, four were women and three were men. Average age at diagnosis was 40.2 years in men

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and 45 years in females. The most common complaint was dyspnea in 5 cases (71.4%) and the most common sign was systolic murmur in 4 cases (57.1%). During follow up (average 3 years), there was no tumor recurrence, hence surgery can be considered as a cure for myxomas.

Conclusion: Cardiac Myxomas are rare primary tumors of heart diagnosed by histopathology and are curable by surgery. The present study showed that the pathological features of cardiac myxomas were not related to the clinical presentation.

Keywords: Cardiac Myxomas, Left atrium, Histopathology, Surgery

Introduction

Primary tumors of the heart are extremely rare, with an estimated incidence ranging from 0.0017% to 0.33% at autopsy (1). Cardiac myxomas represent the most frequent benign cardiac tumors. In most surgical series, they account for almost 80% of cases (2). The cells giving rise to the tumor are multipotential mesenchymal cells that persist as embryonal residues during septation of the heart (3-4). They are also thought to arise from cardiomyocyte progenitor cells, subendothelial vasoformative reserve cells or primitive cells which reside in the fossa ovalis and surrounding endocardium or endocardial sensory nerve (5-8).

Two types of macroscopic appearance are observed: polypoid type and papillary type (11-12). The histopathological diagnosis of a cardiac myxoma depends on the identification of the myxoma cell, which has occasionally been called the lepidic cell (13). The cells are arranged singly or in small clusters, or formed capillary like channels (2). Some morphological and immunohistochemical features may be related to the clinical presentations. Burke found that embolic myxomas were less often fibrotic than non-embolic myxomas and were more likely thrombosed and extensively myxoid with an irregular frond-like surface. Fibrotic and non-thrombosed tumors had a longer mean duration of clinical symptoms and were found in older persons. Recurrent, multiple, and familial myxomas were more often found in younger women and, more likely irregular surfaced and histologically myxoid (4). Endo's group reported that tumors associated with constitutional signs were significantly more likely to be large, multiple, or recurrent than those unassociated with constitutional signs (14). Papillary surface myxomas are thought to be related to embolism, and large left atrial tumors are

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related to atrial fibrillation. Myxoma cells usually express IL-6, and some tumors have abnormal cellular DNA content (15). A C769T PRKAR1a mutation has been observed in "familial myxomas" (16). Therefore, we presented a retrospective review of our institution's experience to study the clinical and pathological features of cardiac myxomas.

Materials and Methods

Study design: This is a retrospective hospital-based study conducted at a tertiary healthcare centre. Histopathological samples of seven cases of cardiac tumors received for a duration of three years from June 2020 to July 2023 were included in study. Other cardiovascular surgery cases were excluded from study.

Data collection: The clinical information of cases including age, sex, history and electrocardiography and echocardiographic diagnosis were noted. All the samples were grossly examined, sections given for tissue processing. Sections were stained with H & E and PAS stains wherever necessary.

Statistical analysis: Data was entered in excel spreadsheet and SPSS software was used to analyzed the data. Mean and standard deviation was calculated for quantitative variables whereas number and percentage were calculated for qualitative variable.

Results

Out of a total of seven cases of cardiac myxomas, four were women and three were men. Average age at diagnosis was 40.2 years in men and 45 years in females. The most common complaint was dyspnea in 5 cases (71.4%) and the most common sign was systolic murmur in 4 cases (57.1%). All the seven myxomas involved the left atrial cavities. On gross examination, the tumors were soft, polypoid and lobulated masses. Areas of hemorrhage were seen in some of the masses (Figure 1). Microscopically, tumors were composed of round to polygonal cells surrounded by abundant loose stroma. There was no evidence of necrosis, pleomorphism or significant mitotic activity (Figure 2). During follow up (average 3 years), there was no tumor recurrence, hence surgery can be considered as a cure for myxomas.

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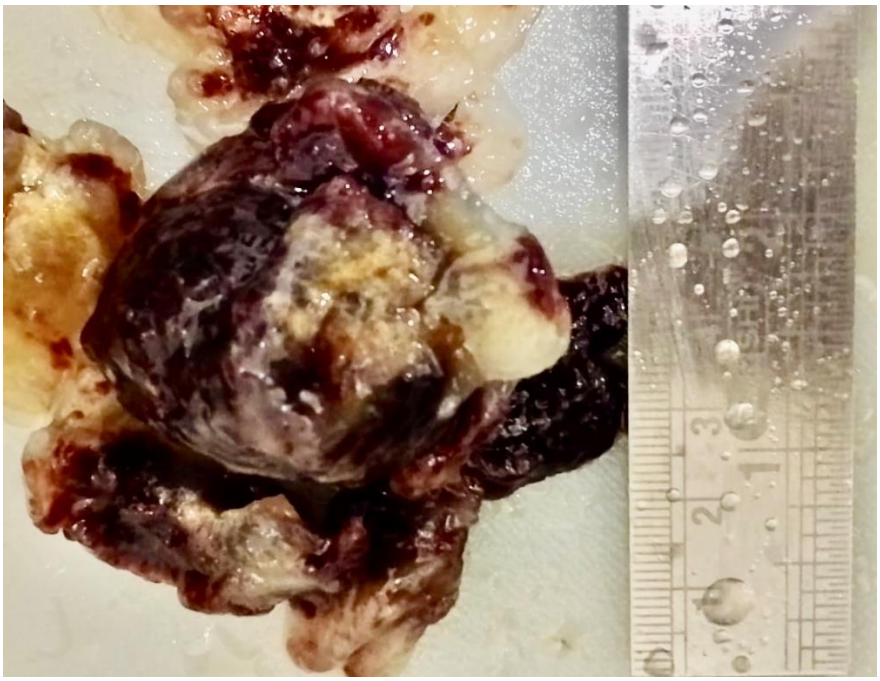

Figure 1: Tumor mass is soft, polypoid and lobulated with areas of hemorrhage (Gross morphology).

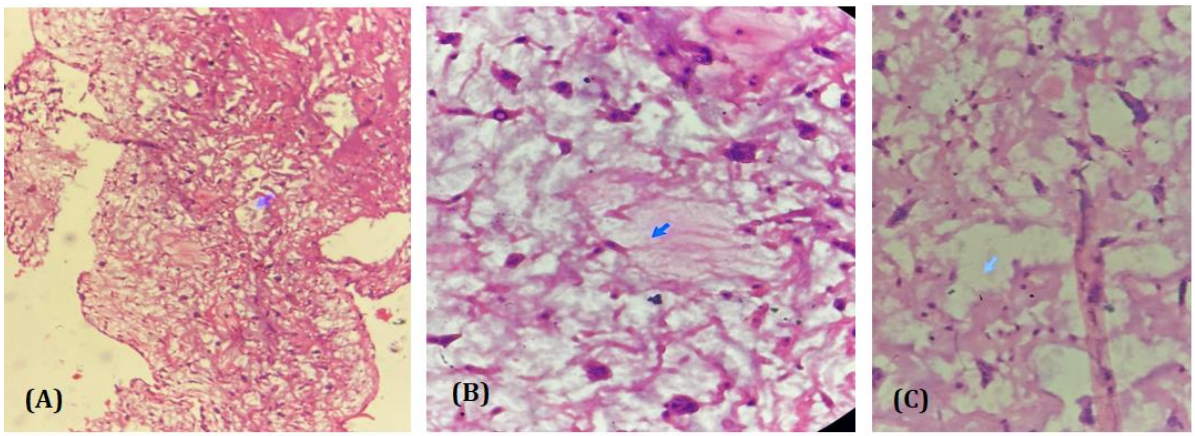

Figure 2: Microscopically, tumors was composed of round to polygonal cells surrounded by abundant loose stroma. There was no evidence of necrosis, pleomorphism or significant mitotic activity. Tumor mass is soft, polypoid and lobulated with areas of hemorrhage. (A) x100 (B) x400 and (C) x400.

Discussion

Study by Ji-Gang Wang et al reported that cardiac myxomas are the most common primary tumor, 90% of the tumors are located in the atria (9). Only 3-4% of myxomas are detected in the left ventricle, and only 3-4% in the right (11). Multilocular myxomas are extremely rare,

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which often result in local recurrences (17). Familial myxomas are estimated to account for 7% of atrial myxomas are more often multiple, recurrent and right sided, as compared to sporadic myxomas (18). The affected patients are also younger, most presenting at 20-30 years of age (19-21). Study by Ji-Gang Wang et al reported surface appearance is associated with the tumor cell arrangement structure (9). Clinical presentations have no relations with the pathological features, which may be different from the previous study (15). Constitutional symptoms, dyspnea, and tumor embolism were the most frequent clinical manifestations (10).

The clinical presentation of myxomas is diverse and dependent upon tumor location, size and mobility (22-25). According to a previous study, the most common symptom is dyspnea (54%), and then followed by palpitation (35%) (15). Dyspnea and edema of lower limbs are thought to a consequence of atrioventricular valve obstruction. Nevertheless, the intracardiac obstruction may also lead to narrowing outflow tract and atrial fibrillation, which could contribute to dyspnea and palpitation. Cough is thought to result in pulmonary venous hypertension and frank pulmonary edema. Angina may be caused by insufficient blood supply. Embolism is also a classic symptom of myxomas, which have been reported to associate with the papillary surface (15). The cause of some constitutional disturbances is still unclear. Some findings suggest the cytokine interleukin-6 (IL-6) may be responsible for that. The relationship between IL-6 and constitutional syndromes is still controversial. In some cases, the right atrial myxomas may induce pulmonary hypertension because of embolism of tumor fragments. Right ventricular myxomas may mimic stenosis of pulmonary valve and cause syncope.

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