

Cardiac Myxomas: A Histodemographic Analysis

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Abstract

Background: Cardiac myxoma is the most common benign cardiac tumour, accounting for more than half of all primary cardiac tumours. Most myxomas are sporadic and the cause is largely unknown. Familial variants with an autosomal dominant inheritance exist. It is localized generally in the left atrium and typically develops in females. Clinical manifestations can mimic cardiac conditions and depend on the natural behaviour of the tumour and its location within the heart, ranging from being completely asymptomatic to causing sudden death. Establishing an early diagnosis is essential, which is confirmed by histopathology. **Objective:** The aim was to find the relation between cardiac myxomas with age, sex and cardiac sites. **Materials and method:** This cross sectional study was done in the pathology department of Delta Hospital Limited, Dhaka, Bangladesh, during the period of January 2014 to July 2016. A total of 24 cases were studied irrespective of age, sex along with clinical diagnosis. **Results:** Among the study subjects females were predominant (70.83%) and highest frequency of cases occurred in between 41-50 years of age (41.67%). Commonest site was left atrium (83.33%). **Conclusion:** The present study revealed that cardiac myxoma occurs most commonly in the 5th decade with female predominance and the most common site is left atrium.

Keywords: Cardiac myxomas; demography; histopathology.

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Introduction

Myxoma of heart is the commonest intra-cavitary cardiac tumour.^{1,2} The clinical manifestations of these tumours are variable and despite the increase of clinical awareness and improved diagnostic techniques, still their diagnosis occasionally

appears as a surprise at surgery or autopsy.³⁻⁵ Also, despite several histochemical and structural studies of cardiac myxomas, the cell of origin and certain of the structural features of these tumours remain subject of controversy.⁶⁻¹²

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Most cases of myxomas are sporadic. Approximately 10% are familial and are transmitted in an autosomal dominant mode.^{13,14} At present the Carney complex is used to describe an autosomal dominant trait, that includes cardiac myxomas, cutaneous myxomas, spotty pigmentations on the skin, endocrinopathy and both endocrine and non-endocrine tumours.¹⁵ These patients are considerably younger at the time of diagnosis when compared to patients with sporadic myxomas.¹⁶

Myxomas are polypoid, round or oval. They are gelatinous with a smooth or lobulated surface and usually are white, yellowish or brown.

Approximately 75% are located in the left atrial cavity at the fossa ovalis, 23% in the right atrial cavity and about 2% in the ventricular cavity.^{16,17} The cells arise from multipotential mesenchymal cells and are characterized as lipidic cells embedded in a vascular myxoid stroma.¹⁶

The light microscopic study of resected tumours revealed abundant mucoid stroma, varied cellularity from area to area along with areas of fibrosis and haemorrhage. The myxoma cell, singly or in clumps, lay in pools of mucoid stroma and they vary in shape from round to spindle to stellate.^{18,19}

The histochemical studies showed that the tumour's ground substance is composed of only hyaluronic acid and chondroitin-4 or chondroitin-6 sulphate.²⁰

While small myxomas can be asymptomatic, the majority may present with one or more of the triad of intracardiac obstruction, cardioembolism and/or non specific constitutional manifestations. The clinical presentation varies depending upon the physical behaviour of the tumour and its location within the heart.²⁰ Obstruction of the circulation through the heart or heart valves

commonly gives rise to symptoms of left (dyspnea, recurrent pulmonary oedema, paroxysmal nocturnal dyspnea, orthopnea) or right sided (peripheral oedema, ascitis, fatigue, hepatomegaly) heart failure, often mimicking mitral or tricuspid stenosis.²¹ Embolisation which occurs in 30-40% of myxomas is usually systemic but may also be pulmonary.¹⁶ Constitutional or systemic symptoms such as fatigue, fever, rashes, joint pains and weight loss can also be seen.

Laboratory abnormalities are usually seen as elevated inflammatory markers such as ESR, serum C-reactive proteins and globulin levels as well as anaemia and high serum interleukin-6 (IL-6) levels.^{16,22} Sometimes low grade but long standing fever can be the only symptom.²³

Once the diagnosis of cardiac myxoma has been made, the patient should be operated upon as soon as possible before undesirable complication, embolization or sudden death occurs. An 8% mortality has been reported in patients awaiting operation following definitive diagnosis.²⁰

Different studies on age and sex distribution relating cardiac myxoma explored that cardiac myxomas are the most common benign primary cardiac tumours in adult.²⁴ They are three times more common in females and 90% diagnosed in the fourth to sixth decades of life.^{25,26} They are rarely seen in children, in whom they constitute 15% of cardiac tumours.^{25,27,28} They can be seen anywhere in the heart, but arise most commonly in the left atrium (60-80%).^{25,29}

Our aim was to find the relation of cardiac myxomas with age, gender and cardiac site in our population.

Materials and method

This cross-sectional study was done on 24 specimens of cardiac tumour biopsy from subjects, irrespective of age and sex from January 2014 to July 2016 in the pathology laboratory of

Delta Hospital Limited, Dhaka, Bangladesh. The specimens were received in 10% formalin as a routine procedure. Paraffin blocks were embedded from each of the specimen and processed in the automatic tissue processor and cut about the thickness of 4 micron and stained with Haematoxylin and Eosin stain for histopathological diagnosis.

Data were analyzed by SPSS for Windows Version 19.0 and the results were expressed as proportion.

Gross picture of myxoma

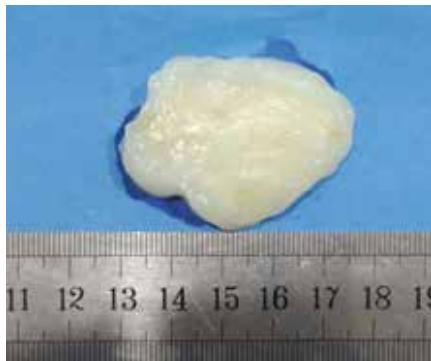


Fig 1: Typical gross appearance of myxoma showing myxoid cut surface

Microscopic pictures of myxoma

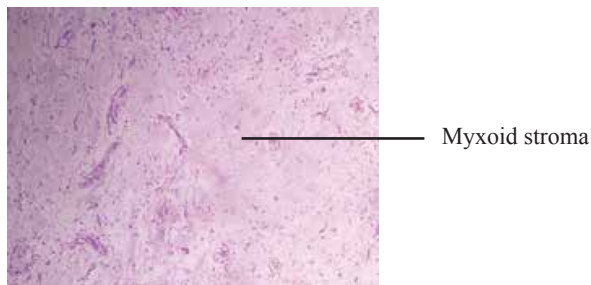


Fig 2: Microscopic picture of myxoma (Low power view)

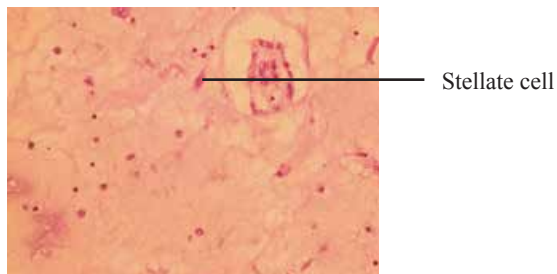


Fig 3: Microscopic picture of myxoma (High power view)

Results

A total 24 cases of both sex were enrolled in the study. The study was aimed at assessing the relation between cardiac myxoma with two major demographic variables, age and sex and also with anatomic site of heart.

Table I shows that 29.16% male and 70.84% female has Cardiac myxomas.

Table I: Sex distribution (N=24)

Sex	Frequency	Percentage
Male	7	29.16
Female	17	70.84

Table II shows majority of the myxomas occur in the left atrium (83.33%).

Table II: Site of distribution (N=24)

Site	Frequency	Percentage
Left atrium	20	83.33
Right atrium	3	12.5
Left ventricle	1	4.17

Table III shows majority of the myxomas occur in 5th decade (41.67%).

Table III: Age distribution (N=24)

Age in years	Frequency	Percentage
<20	1	4.17
20 – 30	2	8.33
31 – 40	5	20.83
41 – 50	10	41.67
51 – 60	5	20.83
>60	1	4.17

Table IV shows most female patients in the 5th decade and male patients in the 6th decade.

Table IV: Mean age distribution in both sex

Sex	Mean	Std. Deviation
Male (n=7)	53.1667	7.35980
Female (n=17)	41.2941	12.87325

Discussion

Myxoma of heart is the commonest primary cardiac tumour. They are three times more common in females and 90% diagnosed in the fourth to sixth decades of life.^{25,26} They can be seen anywhere in the heart, but arise most commonly in the left atrium (60-80%).^{25,29}

The present study shows 70.83% cardiac myxomas occur in female and male to female ratio is 1:2.43. Similar studies had been done previously. A study in China with an observation period of 2007-2012, showed male to female ratio was 1:1.54, which represented female predominance.³⁰ Another study by Zheng et al.³¹ among a series of 66 cardiac myxomas also reported similar results regarding gender distribution of myxoma, where male to female ratio was 1:2.7. A study by King showed female predominance in cardiac myxomas.¹³ Two other studies showed that cardiac myxomas are three times more common in females.^{25,26} All the results are very close to the present study but do not reflect the exact scenario of sex distribution. The possible explanation may be the very low number of myxoma patient was included in this study.

The present study demonstrated that the highest incidence of myxoma is in the 5th decade with most female patients also in the 5th decade and most male patients are in the 6th decade. Panagiotis et al. with an observation period of 1963-1975 reported the highest incidence was in between 30-60 years which was a wide range in respect to the present study.²⁰ A similar study done in China reported the highest incidence to be in the 48.8 years and 51.9 years, that is in the 5th and 6th decades in female and male respectively, which was very close to the present study.³⁰ Another study by Aggarwal et al. which showed the mean age for cardiac myxoma is 56 years.³² The possible explanation for this discordance is that the previous studies, where sample size was larger than the present one, which is a hospital based study.

In the present study 83.33% of myxomas occurred in left atrium and 12.5% occurred in right atrium and remaining were in the left ventricle. Many similar results were found previously through many studies. Larsson et al. showed that 75-85% of cardiac myxomas occur in left atrial cavity and upto 25% occur in right atrium.³³ Another study by Wang et al. shows 88.5% cardiac myxomas occur in left atrial cavity, 9.8% in right atrium and 1.7% in right ventricle.³⁰

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