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Clinicopathological profile of cardiac Myxomas in western Maharashtra population

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Abstract

Introduction: Cardiac tumors constitute only 0.2% of all tumors. Primary cardiac tumors are extremely rare with an incidence of 0.0017 to 0.19. Cardiac myxomas are the most common benign neoplasm of the heart. The prevalence is rare and incidence is 0.001% to 0.03%.

Objective: To review our experiences with this rare entity and highlight the various aspects of myxoma presentation, histopathology and diagnosis in Western Maharashtra Population.

Methodology: A total of 18 cases were accrued from the histopathology files of MH Cardiothoracic centre from 2009-2019 and studied.

Results: A total 18 cases were included in the analysis. Mean age was 45 years, with equal male to female ratio (50%). Patients ranged from 01 months to 84 years old. Echocardiography was the diagnostic technique employed. The tumor was demonstrated by angiocardiology prior to surgery. Hemodynamically, left atrial myxomas revealed moderately severe pulmonary hypertension and right atrial myxomas, with right atrial hypertension. There were 12 myxomas in the left atrium, 3 in the right atrium, 1 in left ventricle, 1 in the Aortic valve and one case of papillary fibroelastoma in left atrium was reported. Eighteen patients underwent open-heart operation with removal of the myxoma. The size of myxoma varied from friable centimetric fragments to globular mass measuring 8.5cm in greatest dimension. The mean dimension of myxoma measured on echocardiography was 3.69 cm. 77.7% percent had left sided and 16.6% had right sided myxoma.

Conclusion: Our results showed that the prevalence of cardiac myxoma in Western Maharashtra is comparable with national and global estimates however the female to male ratio was same, no gender predominance was noted. The mean age of presentation was 45 years, slightly lower than global pattern. Mean greatest dimension was 3.69cm. Prevalence of right atrial myxoma was slightly higher in our sample compare to other national and international published literature.

Keywords: Clinicopathological, cardiac, Myxomas

Introduction

Cardiac tumors constitute only 0.2% of all tumors. Primary cardiac tumors are extremely rare with an incidence of 0.0017 to 0.19 [1]. Cardiac myxomas are the most common benign neoplasm of the heart. The prevalence is rare and incidence is 0.001% to 0.03% [2]. They are known to occur at any age with mean age of 50 years. Female predominance is noted with females getting twice as affected as men. Myxoma can arise in any of the chambers of heart but left atrial myxomas are the most prevalent than right atrial myxomas. Clinical profile of a myxoma is determined by its location, size and mobility. Symptoms are a result of mechanical obstruction caused by large pedunculated masses, embolism (friable papillary myxomas) or constitutional effects [3]. Clinical symptoms vary from being completely asymptomatic to sudden death caused by obstruction and tumour embolus [4]. Despite the wide spectrum of presentation cardiac myxomas present with one of the symptoms of classical triad of symptoms: 1) symptoms of mitral valve obstruction; 2) symptoms of embolism; 3) constitutional symptoms. Surgical excision yields an excellent prognosis and recurrence rate of only 3% with a very low morbidity and mortality [5].

Objective

We aimed to review our experiences with this rare entity and highlight the various aspects of myxoma presentation, histopathology and diagnosis in Western Maharashtra Population.

Materials and Methods

A 10 year retrospective review done on patients who underwent excision of myxoma with histo-pathological confirmation since 2009 to 2019 at a single Cardiothoracic Centre . Immunohistochemistry and special stains were carried out and the slides were reviewed by two pathologists.

Results

A total 18 cases were included in the analysis. Mean age was 45 years, with equal male to female ratio (50%). Patients ranged from 01 months to 84 years old. Echocardiography was the diagnostic technique employed. The tumor was demonstrated by angiocardiology prior to surgery. Hemodynamically, left atrial myxomas revealed moderately severe pulmonary hypertension and right atrial myxomas, with right atrial hypertension. There were 12 myxomas in the left atrium, 3 in the right atrium, 1 in left

ventricle, 1 in the Aortic valve and one case of papillary fibroelastoma in left atrium was reported. Eighteen patients underwent open-heart operation with removal of the myxoma. The size of myxoma varied from friablecentimetric fragments to globular mass measuring 8.5cm in greatest dimension. The mean dimension of myxoma measured on echocardiography was 3.69 cm. 77.7% percent had left sided and 16.6% had right sided myxoma. Most common symptom was dyspnoea, constitutional symptoms, palpitations and stroke. Echo being the diagnostic modality of choice. All patients had a solitary tumour on echocardiogram and each was completely excised. Seventeen revealed typical myxoma features while one was diagnosed as papillary fibroelastoma. Immunohistochemistry including Calretinin, CD68, SMA, EMA, CD34 Vimentin, CK and special stain (Alcian blue and PAS) were performed for confirmation of diagnosis.

Table 1: shows the clinicopathological profile of the 18 cases

Case no.	Age	Sex	Tumour size	Tumour location	Opinion
1	57	F	3.5cm	Left atrium	Myxoma
2	42	F	10ml bits	Left atrial clot with mass	Papillary fibroelastoma
3	38	F	6.5cm	Left atrium	Myxoma
4	63	M	2.5	Left atrium	Myxoma
5	27	M	3cm	Right atrium	Myxoma
6	1mth	M	0.8cm	Left atrium	Myxoma
7	24	F	4cm	Left atrium	Myxoma
8	45	F	5cm	Left atrium	Myxoma
9	54	M	6cm	Left atrium	Myxoma
10	12	F	06ml bits	Left ventricle	Myxoma
11	57	F	10 ml bits	Left atrium	Myxoma
12	67	M	1.5	Aortic valve	Myxoma
13	60	F	8.5cm	Right atrium	Myxoma
14	65	M	5cm	Right ventricle	Myxoma
15	53	F	1.5cm	Left atrium	Myxoma
16	45	M	-	Left atrium	Myxoma
17	34	M	-	Left atrium	Myxoma
18	84	M	-	Left atrium	Myxoma

Discussion

Cardiac myxomas are the most common primary neoplasms of heart. [6] The cardiac myxoma are of primitive multipotent mesenchymal origin [7]. The cardiac myxomas are reported in all age groups with a mean age of 45 years. There is female preponderance with male to female ratio of 5:4. Majority of cardiac myxomas are sporadic, 7% are familial and are associated with clinical complex (Carneys complex). In these cases the tumour affects young males and is usually attached to right atrium or is multicentric [8]. Mean age in these cases is 24 years and 12-22% are known to be recurrent. Carneys complex is an autosomal dominant condition associated with NAME syndrome (Nevi, Atrial myxomas, Myxoid neurofibroma and Ephiledes), or LAMB syndrome (Lentigenes, , Atrial myxomas, Mucocutaneous myxomas and Blue nevi). Mutation of the PRKARIA gene are associated with familial cardiac myxoma in Carney complex [9].

Clinical presentation: Symptoms of mitral valve obstruction, in myxoma may lead to cardiac failure . Symptoms of embolism, is associated with 1 or more locations, most common being cerebral emboli with stroke. Males are statistically at greater risk than females of developing embolic complications. The third type of

manifestations consists of constitutional symptoms with fever, weight loss, or malaise are due to cytokine (interleukin-6) secretion overproduced in the myxoma tissue and secreted into the systemic circulatory system. This might contribute to the systemic inflammatory or autoimmune manifestations seen in cardiac myxoma [10]. Younger male have more neurologic symptoms, and female patients have more systemic symptoms. Cardiac auscultation abnormalities, occur due to pseudo-mitral valve disease (caused by larger pedunculated masses) and more rarely the suggestive tumor plop is known to occur. The tumor diameter ranged from 1 to 10 cm. The myxoma surface was friable in 50% of the cases, and smooth and globular in the other 50% cases. Friable myxomas presented with embolism symptoms; those in patients with cardiac symptoms, pseudo-mitral obstruction present with tumor plop, electrocardiogram or radiologic signs of left atrium hypertrophy and dilatation were noted in the larger tumors. The long-term prognosis is excellent post surgery. The recurrence rate is low (5%), but long-term follow-up and serial echocardiography are advisable especially for young patients [11].

Grossly : The tumour is spherical, soft grey white, gelatinous, lobulated tumour ranging from 1 to 10 cm in

maximum dimension, typically attached with a stalk, (can be sessile also) to the left atrium near fossa ovalis (most common site) [12]. Areas of hemorrhage and calcification are common. Extremely rare cases of heterologous bone formation are also reported.

Microscopically : According to WHO the histopathological diagnosis depends on identification of the Myxoma or Lepidic cell [13]. They can be seen singly scattered, in nests, rings, cords or form capillary like channels in abundant myxoid background. Myxoma cells are frequently stellate cells with eosinophilic cytoplasm and indistinct cytoplasmic borders, oval, pale nuclei with open chromatin and prominent nucleoli. Thin walled blood vessels are seen scattered throughout the lesion however the pedicle or stalk contains thick walled vessels. Long standing cases also show secondary changes in form of fibrosis, calcification, necrosis, hemosiderin deposition, thrombosis and Gamma Gandy body formation. In rare instances heterologous differentiation in form of gland formation, bone formation and extramedullary hematopoiesis are also noted.

Special stains and Immunohistochemistry: In cardiac myxomas the mucopolysaccharide-rich matrix (myxoid) background show positivity (appeared purplish red) in Alcian blue and Periodic acid schiff stains. The myxoma cells show positivity for Calretinin (75-100%), Vimentin (50%) and show variable immunoreactivity to CD34, SMA, EMA, CD68, CK and alpha 1 antichymotripsin.

Differential diagnosis: include lipomas, Thrombus, papillary fibroelastomas and myxoid sarcomas.

1. Lipomas are well circumscribed lesions which lack the myxoma cells and shows only mature fat cells.
2. Thrombus will show zonation pattern with fibrin and lacks myxoma cells.
3. Papillary fibroelastomas occur on valvular cusps and show papillary fronds lined by endothelial cells and lack the myxoma cells [14].
4. Myxoid sarcomas will show pleomorphic spindle cells with increased cellularity and mitotic activity and often associated with areas of osteosarcoma and chondrosarcoma [15].

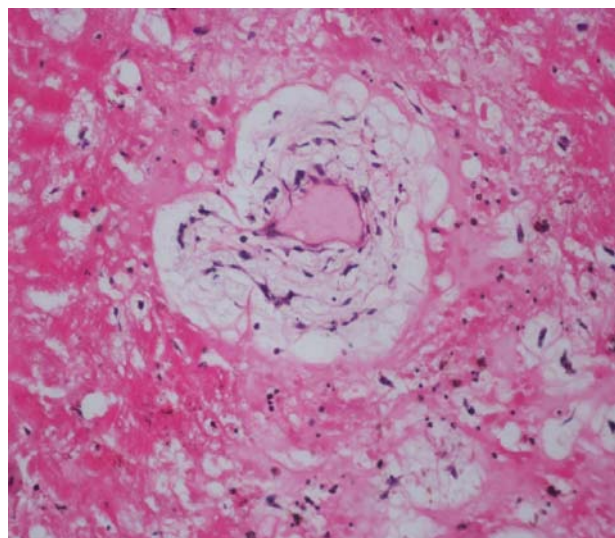


Fig 1: Cardiac Myxoma showing Lepidic cells in nests, HE x 200

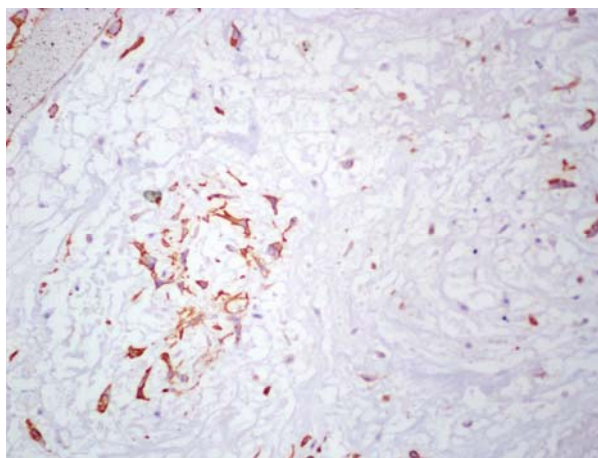


Fig 3: Myxoma, positive staining of vessels by IHC for CD 34 (x 200)

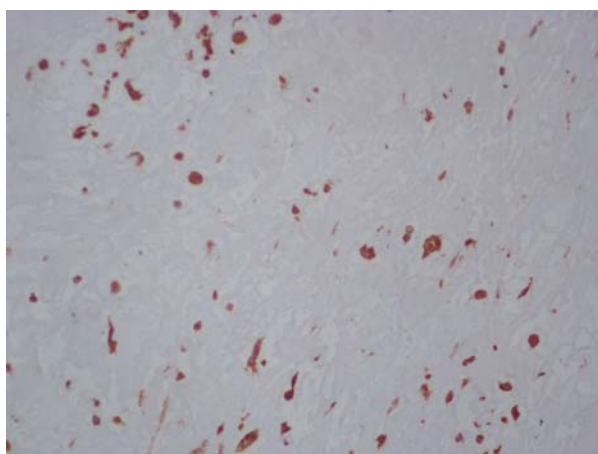


Fig 3: Myxoma, Positive staining by IHC for vimentin (x 200)

Conclusion

Our results showed that the prevalence of cardiac myxoma in Western Maharashtra is comparable with national and global estimates however the female to male ratio was same, no gender predominance was noted. The mean age of presentation was 45 years, slightly lower than global pattern. Mean greatest dimension was 3.69cm. Prevalence of right atrial myxoma was slightly higher in our sample compared to other national and international published literature. Dyspnoea on exertion, palpitation and chest pain were the main presenting symptoms. Echocardiography was the diagnostic modality of choice prior to surgery. None of the patients were reported to have recurrence following surgical excision.

Postoperative hemodynamic and clinical improvement was more striking in patients with a left atrial myxomas reported in previous studies [16]. Immunohistochemistry and special stains were performed on the cases which revealed diffuse and strong positivity for Calretinin and Vimentin whereas variably positivity with SMA, EMA, CD34, CD68 and CK. Myxoid stroma was positive for Alcian blue and PAS stain.

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