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Original Research Article

Cardiac myxoma - 2 years experience at a tertiary care center

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ABSTRACT

Background: Cardiac myxomas are the most common primary neoplasms of the heart. The current study aims to ascertain the clinical relevance of histomorphological classification and to postulate the possible cell of origin of cardiac myxomas.
Methods: A total of 7 cases were analysed and reviewed with the assessment of the clinical presentations and gross and microscopic pathology along with immunohistochemical findings. All these patients were followed up to look for recurrence.
Results: Of the total cases there were 5 women and 2 men. The mean age at presentation was 49 years. The most common presenting complaint was dyspnoea. On microscopy all the cases had a myxoid stroma with cells arranged in a predominantly single stellate cell pattern subtype. The tumor cells in all the cases were diffusely and strongly positive for vimentin with variable expression of S 100, CD 34, CD 68 and desmin.
Conclusions: The identification of histological variability of cardiac myxoma has no significant clinical impact. Our study confirms the postulation that myxomas develop from multipotent mesenchymal stem cells.

Keywords: Cardiac tumor, Immunohistochemistry, Myxoma

INTRODUCTION

Myxomas are intracavitary benign tumors occuring within any of the cardiac chambers, with a predilection for the atria, particularly the left atrium (75%).[1] The majority of the cardiac myxomas arise from the atrial septum (90%), usually from limbus of the fossa ovalis. All the evidence reveal that the cardiac myxomas are benign neoplasms and slowly proliferating lesions.[1,2]

The previous studies have reported a large series of myxomas, however little have been focused on the histopathological classification and their relevance. Cardiac myxomas show a lot of histomorphological variability. Another contensious issue is the origin of myxoma cell.

We present a retrospective review of our institution's experience in the clinical and pathological evaluation of cardiac myxomas. The focus of the study has been to ascertain the clinical relevance of histomorphological classification and also to postulate the possible origin of cardiac myxomas.

METHODS

For our study we identified 7 consecutive cases of cardiac myxomas surgically resected at our cardiac center during 2012 -2014. The medical records were reviewed for clinical presentations, methods of diagnosis and management. 2D echocardiography was performed in all patients which could well determine the tumor location, size, shape, attachment and mobility.

All patients underwent complete myxoma excision in the same institution with cardiopulmonary bypass (CPB). For the one case which presented as aortoiliac thromboembolism a second surgery was done later for left atrial myxoma. Follow up was done in all cases which ranged from a minimum of 1 year 4 months to a maximum of 3 years 7 months (mean- 2 years 6 months).

RESULTS

All the cases in our study were sporadic myxomas. The age ranged from 36 to 65years (mean age was 49 years).

The most common site of involvement was left atrial cavity (86%) and one case was in the right ventricle (14%). The most common presenting symptom was dyspnoea in 71% followed by palpitation, cough, fatigue and fever. Course of the disease ranged from 2days to 6 years. The most common sign was trivial aortic regurgitation or tricuspid regurgitation.

All the seven cases had a predominant myxoid stroma with intervening cells. These cells were arranged in a predominantly single stellate cell pattern subtype.

Table 1: Clinical and histological findings with follow up in 7cases.

Age (years) /Sex	Location	Size (cms)	Microscopy features	Follow up
51/F	L atrium	3.5	Stellate cells, single file, cord like pattern	No recurrence 3yrs 7 months
43/F	L atrium Interatrial septum	6.5	Stellate cells, single file, foci of hemorrhage	No recurrence 3 yr 4 months
52/F	Aortoiliac thromboembolism Followed by left atrium	3	Stellate cells single file, foci of cartilage differentiation	No recurrence 2yrs 11 months
52/F	L atrium	3.5	Stellate single cells, vasoformative area	No recurrence 2yrs 5months
46/F	R ventricle	7	Stellate cells single file, cord like pattern	No recurrence 2 yrs 1month
65/M	L atrium	6	Stellate single cells, vasoformative area	No recurrence 1 yr 9 months
34/M	L atrium	5	Stellate cells, single file, foci of ossification	No recurrence 1 yr 4 mnths

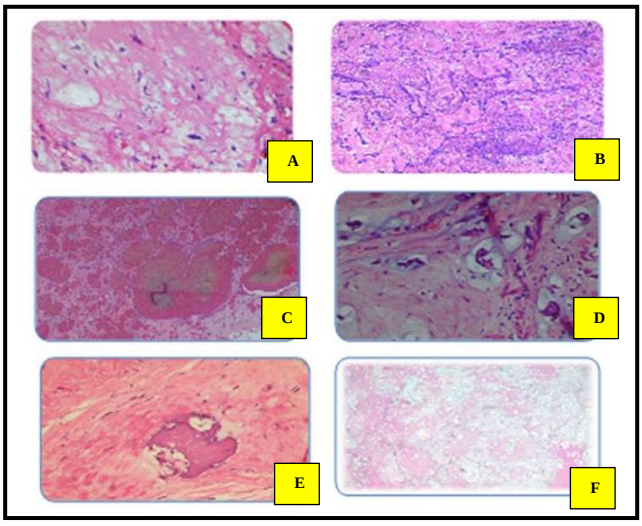

Figure 1: Varying histomorphological patterns (A to E: H and E X 200); (A) single stelate cell in myxoid background; (B) Cell cord pattern; (C) Vasoformative pattern; (D) Area of cartilageneous differentiation; (E) area of osseous differentiation; (F) AB PAS stain showing blue myxoid areas (X200).

The other subtypes were cell cord predominant pattern and vasoformative ring subtype seen in two cases each. The tumor cells were mainly stellate to spindle and polygonal cells in a few with round to oval nuclei and pink cytoplasm. The myxoid stroma surrounding the tumor cells showed pink to blue areas on PAS stain.

None of the cases showed any mitotic activity. The case with vasoformative ring predominant pattern showed dilated and distended blood sinus like areas. One case showed areas of cartilaginous differentiation while another case showed focus of osseous differentiation (Figure 1). No gland formation was noted in any of the cases (Table 1).

Immunohistochemical staining was carried out in all the 7 cases. The tumor cells were diffusely and strongly positive for vimentin in all the cases. There was variable staining with S 100, CD34, CD68 and desmin (Figure 2). The diagnosis was in concordance with the echocardiography findings.

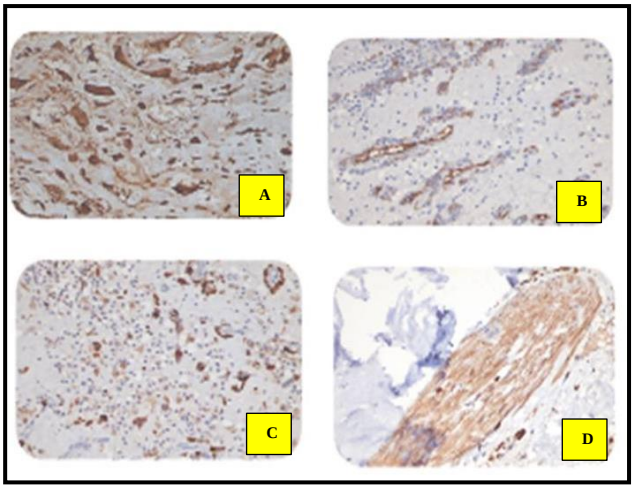

Figure 2: Immunohistochemical staining showing positivity in tumor cells for vimentin (A), CD34(B), CD68 (C) and S100 protein (D).

DISCUSSION

Cardiac myxomas are very rare tumors. We came across 7 cases in a 2 year period with 6 left atrial tumors and 1 right ventricular tumor. One unique case presented with aorto-iliac embolism and only later was found to have a myxoma in left atrium. However cases have been reported with cerebral embolic presentation with as many as 30% having such presentation in one study.[3]

Myxomas may have varying morphology but the most commonly identifiable in our study has been a neoplasm composed of stellate cytologically bland cells in a prominent myxoid stroma. Three different histological patterns were appreciable in our study.[4] But this classification has no clinical or prognostic significance as has also been deduced in other studies.[5] None of our cases have recurred .The cellularity was variable in our cases but no mitosis was noted. But cellularity and mitotic index do not correlate consistently with recurrence.[6] Our cases showed different mesenchymal differentiation along vascular, cartilage, neural and osseous lines, but no glandular areas. Glandular differentiation in myxoma is rare and has been reported mainly in association with Carney's complex.[7] All our cases were sporadic cases of cardiac myxomas.

Our cases on immunohistochemical (IHC) analysis showed variable expression of S100 protein, desmin, CD68 and CD34 with none positive for cytokeratin. These results support the hypothesis of myxoma originating from multipotent mesenchymal stem cell which is capable of differentiation along smooth muscle, vascular, neural and histiocytic lines.[4,8] Although we had vimentin expression in all our cases, calretinin has been shown to be expressed in75 to 100 percentage cases in other studies.[9] Some earlier studies have shown significant schwann cell and neuroendocrine differentiation and proposed that myxomas originate from endocardial sensory nerve tissue.[10,11]

Familial cardiac myxomas seen in Carney complex on cytogenetic analysis are principally involved with mutation in CNC1 and CNC2 genes.[12] No single gene mutation has been identified with sporadic cardiac myxomas although structural reaarangement in PRKAR1A has been identified in some cases.[13] Molecular analysis in cardiac myxomas have shown overexpression of IL6, VEGF,FGF and PCNA in highly proliferative angiogenic and malignant myxomas.[14] A further understanding of upstream and downstream regulators will in future help define potential drug targets in cardiac myxomas.

CONCLUSION

Cardiac myxomas are among the most common primary benign cardiac tumor. The identification of histological variability has no significant clinical impact. Our study confirms the postulation that myxomas develop from multipotent mesenchymal stem cells.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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