Case Report

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Pathology Section

Right Ventricular Myxoma in an Asymptomatic Patient: A Rare Case Report

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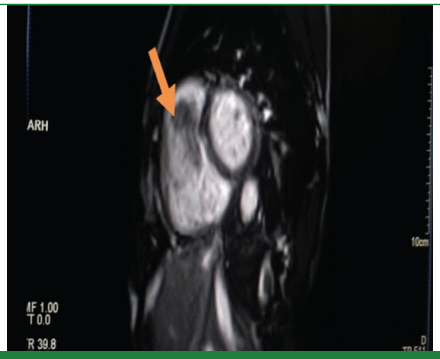
ABSTRACT

Myxomas are the most common primary tumours of the heart (40%). It usually occurs sporadically and are most commonly located in the left atrium (86%). Here, we report a 40-year-old male patient who was incidentally diagnosed of right ventricular myxoma. Myxomas of the right ventricle are very rare. Associated syndromes including carneys complex and family history should be sought when encountering a myxoma in a young adult and in an uncommon location. However, these were not demonstrated in the present patient.

Keywords: Benign, Sporadic , Ventricular Mass

CASE HISTORY

Here, we present a rare case of right ventricular myxoma detected incidentally in an asymptomatic patient. A 40-year-old male, presented to a private practitioner with vague complains of fatigue and weakness; patient was absolutely fine otherwise. During examination, the clinician detected abnormal heart sounds on auscultation. Patient was subsequently referred to Goa Medical College for further investigations and follow-up. Here, the patient underwent a thorough cardiac workup in view of abnormal heart sounds. A subsequent ECHO done on the patient revealed a large right ventricular mass measuring 3.8x3.4 cm attached to the lateral wall. The mass was seen impinging on the pulmonary valve; however causing no stenosis/regurgitation of pulmonary valve. Cardiac MRI of the patient also revealed a mass in the right ventricle with extension into the right ventricular outflow tract and main pulmonary artery [Table/Fig-1]. There was subtle minimal enhancement on post contrast sequences along right ventricular free wall just below tricuspid valve. Based on these imaging findings, two possibilities: 1) Benign neoplastic mass (myxoma) and 2) thrombus were suggested. Patient's haematological examination revealed Hb of 8.7 gm%. Other haematological parameters were normal. Biochemical and immunological investigations were within normal limits. Serologic tests for HIV, HbSAg, HCV, TPHA were negative. Patients ECG, coronary angiography, plain CT of abdomen and pelvis were normal.



[Table/Fig-1]: Cardiac MRI showing Right ventricularmyxoma.

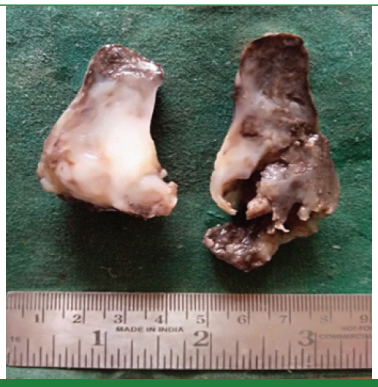
Patient was operated by open heart surgery, through mid- sternotomy incision. OT findings revealed marked cardiomegaly with dilatation of right atria, right ventricle and pulmonary artery. Large pedunculated mass was seen in the right ventricle just below the annulus of anterior tricuspid leaflet, attached to the free wall of right ventricle. Mass was enchroaching into right ventricular outflow tract. No other intracardiac anomaly was noted. The mass was surgically excised and sent for histopathological examination.

Patient is being followed up in cardiac OPD since 6 months. Patient's follow-up course is uneventful.

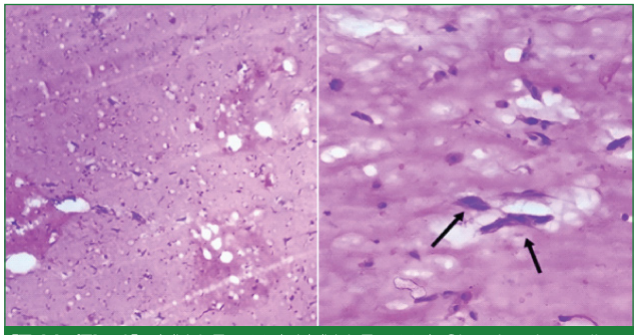
Grossly, it was a brownish white friable, soft polypoidal (globular) specimen measuring 6x4x2.5 cm. Surface was smooth. Cut section was whitish gelatinous with few areas of haemorrhage [Table/Fig-2].

On microscopic examination, multiple sections revealed a well-circumscribed tumoural mass comprising of myxoma cells dispersed in a myxoid matrix [Table/Fig-3a,b]. The myxoma cells were polyhedral to stellate shaped, with moderate amount of homogenous eosinophilic cytoplasm and hyperchromatic elongated to oval nuclei. Scattered aggregates of lymphocytes and histiocytes were observed, along with areas of haemorrhage. Considering the architecture, the lesion had a normal differentiation (well developed vessels surrounded by cells and then by matrix). Myxoma showed areas of variable cellularity and hence, was typed as mildly active. Mitosis was Nil. Overlying epithelium showed ulceration. A bit of underlying cardiac muscle was also observed [Table/

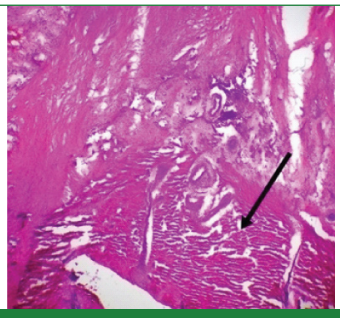
Fig-4]. Based on this histopathological picture, it was reported as: Myxoma arising in the Right Ventricle.



[Table/Fig-2]: Cut section- showing whitish myxoid area with areas of haemorrhage.



[Table/Fig-3]: a) (H & E, 100x); b) (H & E, 400x): Showing the stellate and polygonal cells in a myxoid background.



[Table/Fig-4]: Figure showing underlying cardiac muscle (H & E, 100x).

Special stains/IHC was not possible in this case. Diagnosis was made solely on H&E examination.

DISCUSSION

The prevalence of primary tumours of the heart has been reported between 0.007%, 0.35% to 0.3%. Three quarter of the tumours are benign and half of these benign tumours are myxoma [1].

Myxomas are the most common primary tumours of the heart, accounting for 50% of the primary cardiac tumours. Upto 75% of the myxomas are located in the left atrium, whereas right sided myxomas are rare (15-20%), with right ventricular myxomas being extremely rare (3-4%) [2]. Most myxomas occur sporadically while 10% cases occur as familial syndromes and are transmitted in an autosomal dominant mode [3]. Patients with familial syndrome (aka Carneys syndrome) usually have multicentric myxomas (which mostly occur in location other than left atrium) with associated extracardiac abnormalities, such as cutaneous and labial lentiginosis, eyelid and cutaneous myxomas, myxoid mammary fibroadenoma (often multiple and bilateral), adrenocortical nodular dysplasia associated with cushing syndrome and a large calcifying sertoli cell tumour of the testis [4]. Myxoma occurs mostly in females aged 20-60 years, whereas myxoma syndrome are most common in males (M:F-2:1) and presents at a younger age [5]. Now, as regards to this case, patient was a male presenting with myxoma at unusual site, it was a single intracardiac lesion (not multicentric), with no other cardiac anomaly and no other associated extracardiac abnormalities. Patient was neither a known case of any syndrome nor there was any family history suggesting the same. Considering this, the myxoma syndrome could be ruled out.

Right ventricular myxomas are rare. Sporadic cases of right ventricular myxoma have been reported. Czapek in 1891, was among the first to provide a pathological description of right ventricular myxoma [6]. Myxomas in the right ventricle usually arise from the tricuspid valve, pulmonary valve, anterior papillary muscle, free wall and right ventricular apex [6]. Patients with right sided tumours, usually presents with signs of right outflow tract obstruction like dyspnoea, syncope, neck vein distention. However, some cases are asymptomatic and are detected unexpectedly [7]. In this case also, patient was asymptomatic and was detected incidentally, though the cardiac MRI showed extension of mass into the right ventricular outflow tract and main pulmonary artery. Similar cases of myxomas presenting asymptomatically has been reported in the literature [Table/Fig-5] [7-10].

Myxomas are often mistaken for various intracavitary cardiac lesions like thrombus, lipoma and non-myxomatous. neoplasms, most of which are malignant [11], hence histopathological data is always required for a definitive diagnosis [12].

It can give rise to complications like congestive heart failure, sudden death, cardiac arrhythmias, infection, embolisation, rupture and myocardial infarction [3]. Multiple embolisation can occur in systemic and pulmonary circulation, depending on their location and can lead to myocardial, pulmonary and cerebral infarcts. The production of emboli has been attributed to the overexpression of matrix metalloproteinases by the tumour [4]. Myxomas with multiple papillary fronds on the surface have the highest propensity to embolise [5]. Since in this case, it was a smooth surfaced globular mass, the risk of this complication was much less.

Surgery is indicated in all patients diagnosed of myxoma, and is considered urgent due to the risk of haemodynamic decompensation and embolisation present throughout the course of disease [1]. Surgery is often curative and prognosis is often excellent after total surgical resection of the tumour.

Year	Author	Case report	Diagnosis	Follow-up course
1993	Yamada T et al., [8]	Reported a case of 13-year-old asymptomatic girl with no history of cardiac disease, was found to have cardiac murmur during routine medical check up	Diagnosis: Diagnosis of Right Ventricular Myxoma was made on MRI, CT, angiocardiography, and ECHO	Patient recovered uneventfully
1993	Ono M et al., [9]	Reported a case of 21 year old male, otherwise healthy, detected incidentally of cardiac murmur	ECHO showed a tumour occupying the right ventricular outflow tract and protruding into main pulmonary artery in systole	Patient was followed up for 32 months; there was no sign of recurrence
1998	Kumamoto T et al., [10]	Reported a 71-year-old female with no symptoms or constitutional signs, except heart murmur.	CT, MRI, ECHO showed a mass in the right ventricular outflow tract, attached to the right ventricular free wall.	Follow-up course was uneventful
2013	Kim D–H et al., [7]	Reported a 42-year-old female, who presented with generalized weakness. Patient had previous history of parotid gland tumour, breast nodule, and recently diagnosed of uterine leiomyoma	Patient was incidentally detected of large mass in pulmonary outflow tract while being evaluated for uterine leiomyoma on CT	Though patient presented with multiple tumours, it was not associated with craneys complex. follow-up studies of this patient is not available

[Table/Fig-5]: Similar cases of myxomas presenting asymptomatically has been reported in the literature [7-10].

Recurrence of a sporadic myxoma is unusual, occuring in 1-3% of cases [13]. Recurrence is usually correlated with young age, myxoma syndrome, inadequate resection, intraoperative implantation or multiple growths [13]. Hence, the resection of entire area of attachment of myxoma is recommended. Recurrence is highest in patients with myxoma syndrome, and the development of a myxoma recurrence, should prompt surgeon to re-evaluate the case to rule out the possibility of myxoma syndrome [5]. The patient in this case will require a long term follow-up, to check for any recurrence and subsequent reinvestigation in case the myxoma recurs.

CONCLUSION

Right ventricular myxoma in an adult is extremely rare, and there is always a high possibility that these patients might be associated with myxoma syndrome. A thorough investigation of the case is always required to rule out the possibility of myxoma syndrome. Rarely, the myxomas occur sporadically in unusual sites and close follow-up of such patients are required to check for any recurrence after total surgical excision.

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