



Right atrial sarcoma masquerading as myxoma presenting with fatal cardiac tamponade: A rare case report

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ABSTRACT

Primary sarcoma of the heart is an exceptionally rare condition with grave prognosis. All cardiac tumours can cause life threatening complications. Diagnosis is much more challenging as cases present with non-specific symptoms. In our case, a series of investigations were made which ultimately led to our diagnosis of a right atrial sarcoma. Here, we recount a fatal case of a locally invasive primary cardiac sarcoma originating from the right atrium and presenting with a cardiac tamponade.

Keywords: Primary Cardiac Sarcoma, Cardiac Tamponade, Myxoma

1. INTRODUCTION

Primary heart tumors are rare. About 25% tumors of the heart are malignant, most of which are sarcomas (Kim et al., 2010). Sarcomas are right sided tumors as seen in most cases which grow rapidly and often invade the pericardial space and obstruct the

flow of blood from the venae cavae and into the ventricles. The principal diagnostic tool is a 2D-Echocardiogram which also aids pericardiocentesis. Cases are fatal mainly due to the tumors local spread, distant metastasis and hemodynamic compromise (Kong et al., 2009). This case study addresses a patient with a right atrial sarcoma with massive pericardial effusion resulting in cardiac tamponade that has been attempted to be surgically treated primarily to result in the tumor becoming inoperative which ended up in a fatal outcome.

2. REPORT

A 59 year old Indian male patient came to medicine department presenting with sudden onset breathlessness since 15 days with bilateral pedal edema. Dyspnea increased progressively from occurring on minimal exertion to occurring at rest. He denied any history of ischemic heart disease, chronic lung disease and hypertension or diabetes mellitus. His general physical examination revealed classical signs of right heart failure- a weak pulse, distended neck veins, hepatomegaly with pain in the right hypochondrium and bilateral pedal edema.

His ECG revealed low voltage complexes. On auscultation over the chest wall, there were muffled heart sounds with decreased air entry over the lower lung fields. Chest radiograph revealed pleural effusion on both sides with pericardial effusion. An echocardiogram (Figure 1) revealed a 5x5 cm mass affixed to the interatrial septum likely to be a tumor or thrombus, in the right atrium, seen prolapsing into the ventricle and obstructing the tricuspid valve. It also showed a massive pericardial effusion suggestive of a cardiac tamponade.



Figure 1 Echocardiogram showing right atrial mass

Clinically, the patient also showed the collection of three clinical signs constituting the Beck's triad- hypotension; muffled heart sounds and an elevated jugular venous pressure which indicated that the patient had cardiac tamponade. In view of these findings, an urgent pigtail catheterization of the pericardial cavity was done to drain fluid. Initially, about 250 ml of fluid was drained which was hemorrhagic. The fluid was sent for cytological analysis which revealed no acid fast bacilli and was a serous effusion. The pericardial effusion was slowly drained through the catheter with about 200 ml being drawn out per hour. His routine blood analysis revealed the following: Hemoglobin-12.9 gm%, WBCs-9600/mm³, Platelets-1,02,000/mm³, INR-1.26, urea-63, serum creatinine-2.1, ALT (alanine aminotransferase)-46 IU/l, AST (aspartate aminotransferase)-32 IU/l, total serum protein-6.6 g/dl, serum albumin-3.4 g /dl, total bilirubin-0.8 mg/dl. Analysis of pericardial effusion showed the following- Total leucocyte count (TLC)- 160 cells/hpf, RBCs-plenty/hpf, Differential leucocyte count- Polymorphs-80%, Lymphocytes-20%, glucose-104 mg/dl, LDH (lactate dehydrogenase)-323 IU/l and protein-3.6 g/dl. ADA levels for the same were normal. Culture of pericardial fluid had no growth and cytology showed plenty of RBCs with no malignant cells with findings pointing towards an exudative effusion. There were no malignant cells present in the fluid. The patients coagulation profile and a lower limb Doppler (both of which showed no abnormality) was done to rule out the presence of a thrombus. A contrast enhanced Cardiac CT angiogram (Figure 2) was done which confirmed a hypodense lesion in the right atrium with mild enhancement on post contrast study seen extending upto the tricuspid valve and projecting into the right ventricle and into the inferior vena cava. These findings were suggestive of a right atrial tumor.



Figure 2 Cardiac CT showing the tumor.

Coronary vessel angiogram was done which revealed normal epicardial coronaries. Patient was referred to cardiothoracic surgery for removal of the tumor and taken for surgery after stabilization. Intra-operative findings (Figure 3) showed the tumor was inside the atrium encroaching upto the IVC (inferior vena cava) and infiltrating the tricuspid valve annulus. It was seen projecting outside the right atrium and seen infiltrating the pericardial cavity and adherent to the pericardium. Grossly the tumor was stony hard in consistency and was completely adherent to the pericardium. The tumor was deemed to be inoperable and a cut section of the pericardium involved was taken and sent for histopathological analysis.

A section of the tumor (Figure 4) has shown below displays oval to elongated cells with modest amount of cytoplasm. The nuclei show anisocytosis with hyperchromasia. Also seen are extensive areas of necrosis with atypical mitotic figures. This suggested that the cardiac mass was a malignant mesenchymal tumor with high grade sarcoma – undifferentiated type. Unfortunately before any further intervention could be taken, the patient died three days after the surgery.

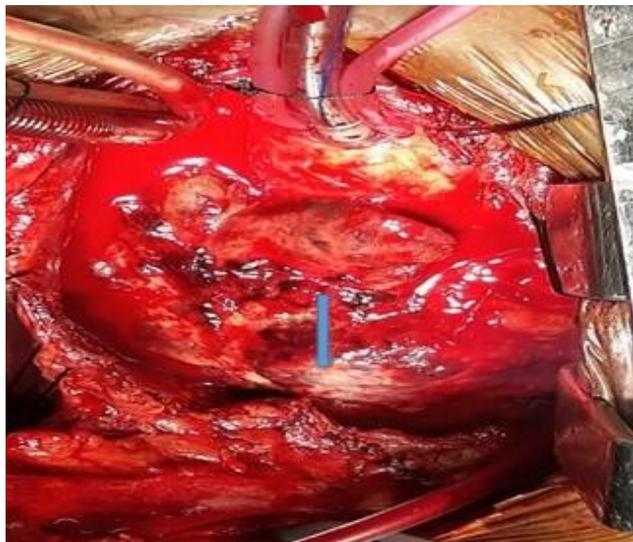


Figure 3 Intraoperative view of the tumor (marked with blue arrow) as described above

3. DISCUSSION

Primary tumours of the heart are extremely rare. Metastasis of secondaries to the heart is relatively more common, occurring more often than primary tumours (Kim et al., 2010; Kong et al., 2009). Tumours metastatic to the heart mainly include malignant melanomas, leukaemias and lymphomas. Only twenty five percent of all 1^o cardiac tumours are malignancies with sarcomas being the ones which are predominant. Sarcomas mostly involve the right side of heart as compared to myxomas which commonly involve the left side. Breathlessness on exertion is the most common symptom on admission, accompanied by chest pain which is non-specific, cough, PND (paroxysmal nocturnal dyspnoea), while some cases also report with embolic incidents like stroke, TIA (transient ischemic attack), haemoptysis and constitutional symptoms like pyrexia of unknown origin, weight loss, fatigue and malaise (Kong et

al., 2009; Vander et al., 2000). Life threatening complications may ensue such as arrhythmias, tumour embolization, obstruction of intra-cardiac blood flow and recurrent pericardial effusion eventually presenting as cardiac tamponade.

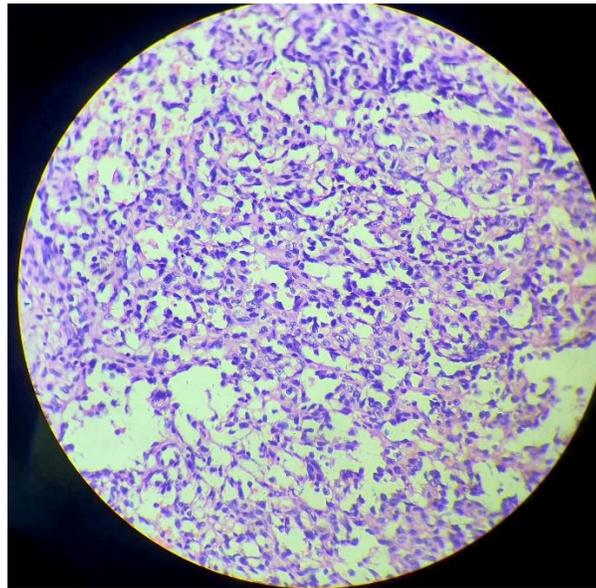


Figure 4 Histopathological section displaying oval to elongated cells with modest amount of cytoplasm. The nuclei show anisocytosis with hyperchromasia.

Echocardiography is the excellent as it aids diagnosis as well as pericardiocentesis in cases with pericardial effusions. Cardiac CT scan as well as a CT angiogram can outline the extent of growth and extracardiac infiltration of the tumour. Pericardial biopsy, similar to pleural biopsy in lung cancers may help in diagnosis. Mean survival in cardiac sarcomas is about 6 months, due to advancing symptoms and insufficient clinical awareness. Non-metastatic disease can be managed with extensive surgical excision but there remain persistent risks of metastasis and recurrence. Tumours which are nonresectable and sarcomas with widespread metastasis can be managed with a combination of chemo- and radiotherapy but desired response is not always (Kumar et al., 2020; Shanmugam, 2006).

4. CONCLUSION

Once a presumptive detection of sarcoma has been made on imaging, the ideal step is complete resection followed by neoadjuvant and post-surgical chemotherapy to minimize the risk of fatal complications (embolization and cardiovascular) including early death as was the case in our patient. In general, prognosis in cases of primary cardiac sarcomas is poor and response to treatment may not be optimal, symptomatic relief is important and attainable.

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Author contributions

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Conflict of interest

The authors declare that they have no conflict of interest.

Informed consent

Informed consent was taken from the participant included in the study.

REFERENCES AND NOTES

1. Anurag Kumar, Sonaakshi Kushwaha, Shubham Jaiswal, Pratyush Singh, Rashmi Deshpande, Sourya Acharya. Unusual presentation of Cardiac Myxoma. Medical Science, 2020, 24(102), 521-525
2. Kim DM, Hong JH, Kim SY, Yoo KD, Seo JY, Rhee KJ, et al. Primary Cardiac Angiosarcoma Presenting With Cardiac Tamponade. Korean Circ J. 2010 Feb;40(2):86–9.
3. Kong PK, Theodorou S, Beran E, Singh H, Dimitri W. Primary cardiac undifferentiated sarcoma of the right atrium presenting with cardiac tamponade. Cardiovasc Pathol. 2009 Mar 1;18(2):110–3.
4. Shanmugam G. Primary cardiac sarcoma. Eur J Cardiothorac Surg 2006; 29:925–32.
5. Vander T, Salm J. Unusual primary tumors of the heart. Semin Thorac Cardiovasc Surg 2000;12:89–100.

Data and materials Availability

All data associated with this study are present in the paper.

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