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Primary Cardiac Tumor: A Rare Case Report of Right Atrial Angiosarcoma

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Abstract: ***Primary cardiac angiosarcoma is a relatively rare tumor with early metastasis and poor prognosis. Radical resection of the primary tumor remains the primary approach for the optimal survival of patients with early-stage cardiac angiosarcoma without evidence of metastasis. In addition, literature analysis showed that surgery remains an effective way of treating primary early angiosarcoma. This case involves a 20-year-old female who came to emergency department of Dr. D Y Patil Hospital, Navi Mumbai with complains of breathlessness, fever and cough without expectoration.***

Keywords: cardiac angiosarcoma, cardiac imaging, pericardial effusion, right atrium

1. Introduction

Primary cardiac angiosarcoma is an exceedingly rare, highly aggressive neoplasm arising from malignant endothelial cells within the cardiac vasculature, most commonly originating in the right atrium. As the predominant histologic subtype of primary malignant cardiac tumors in adults, it is characterized by rapid infiltrative growth, early local invasion, and a high propensity for distant metastasis

Approximately 20% to 30% of all primary cardiac tumors are malignant, including angiosarcoma, lymphoma, fibrosarcoma, myosarcoma, and myosarcoma. Of these, angiosarcoma is the most common type of primary cardiac malignancy in adults, accounting for 28.6% of these malignant cardiac tumors.

Clinical presentation is often insidious and non-specific, including symptoms such as exertional dyspnea, chest discomfort, pericardial effusion, and signs of right-sided heart failure, frequently leading to delayed diagnosis. Definitive diagnosis necessitates multimodal imaging—typically including echocardiography, cardiac MRI, or CT- followed by histopathologic and immunohistochemical confirmation. Despite advances in surgical resection, chemotherapeutic regimens, and radiation therapy, the overall prognosis remains dismal, with median survival often limited to a few months post-diagnosis due to the tumor's aggressive clinical course.

2. Case Report

Here, we describe the case of a 20-year-old female patient who came to emergency department of Dr. D.Y. Patil Hospital, Navi Mumbai with complains of breathlessness, fever insidious in onset, gradually progressive associated with chills and rigor. Patient also complains of cough without expectoration.

2D echo and X-RAY Chest was advised.

On 2D echo.
Two-dimensional transthoracic echocardiography was performed using the conventional 2D probe . It revealed normal left ventricular size, systolic and diastolic function. Subcostal imaging revealed a large cardiac mass measuring 8 × 4 cm adjacent to the right atrium (*Figure 1*) with pericardial effusion (*Figure 2*)

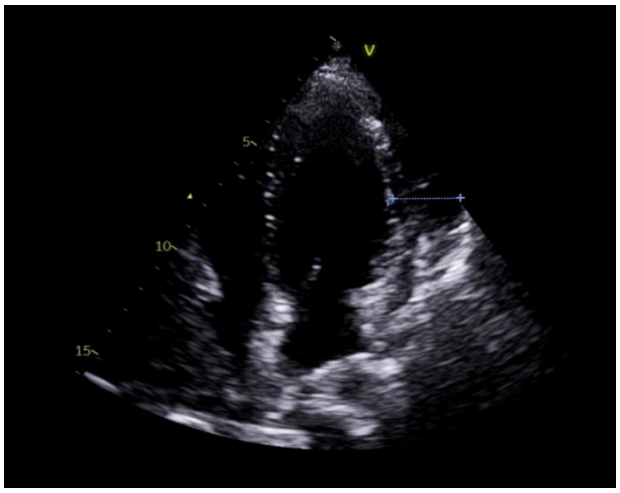


Figure 1

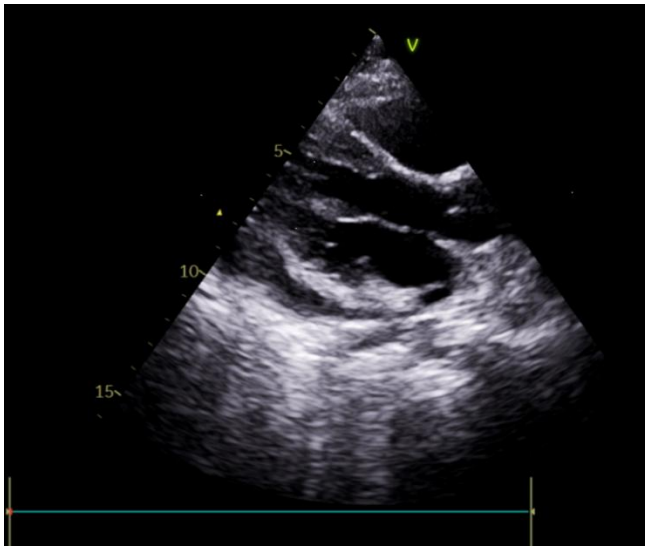


Figure 2

X-RAY Chest

Findings:
A well -defined opacity is seen in the right paracardial region. Further correlation with HRCT THORAX was advised.

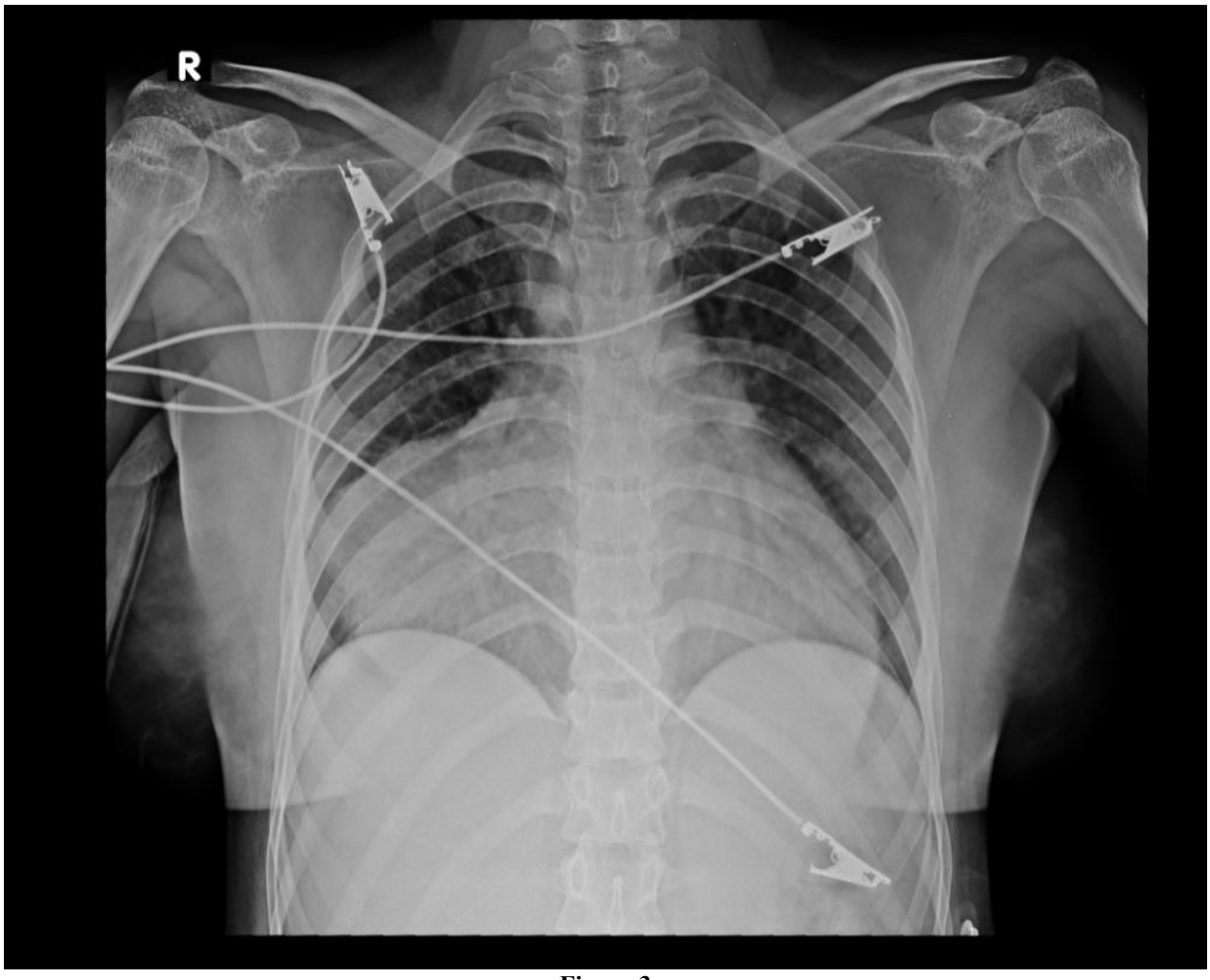


Figure 3

Chest X-ray shows well -defined opacity is seen in the right paracardiac region. There is also associated moderate cardiomegaly seen.

HRCT THORAX:

Findings:
A 8.0 x 5.0 x 8.9 cm (AP x TR x CC) sized ill-defined heterogeneously enhancing mixed soft tissue mass involving the right para-cardiac region, not seen separately from the lateral aspect of the right atrial wall. It was seen splaying the pericardium away from the right atrium and right ventricle. It was also seen extending into the right atrial cavity. The lesion is seen extending from D5 to D10 vertebral body levels. Few areas of hyperdensities of blood attenuation was noted within suggestive of hemorrhagic components. Few intra lesional vascular channels are noted in venous phase. The lesion was

seen encasing proximal aspect of SVC and abutting the root of aorta

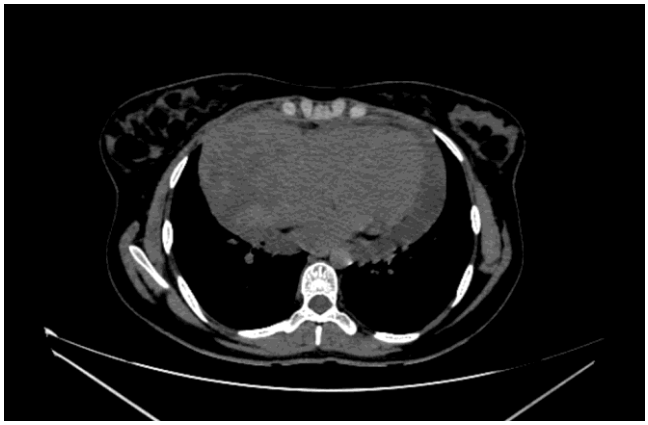


Figure 3 (a):

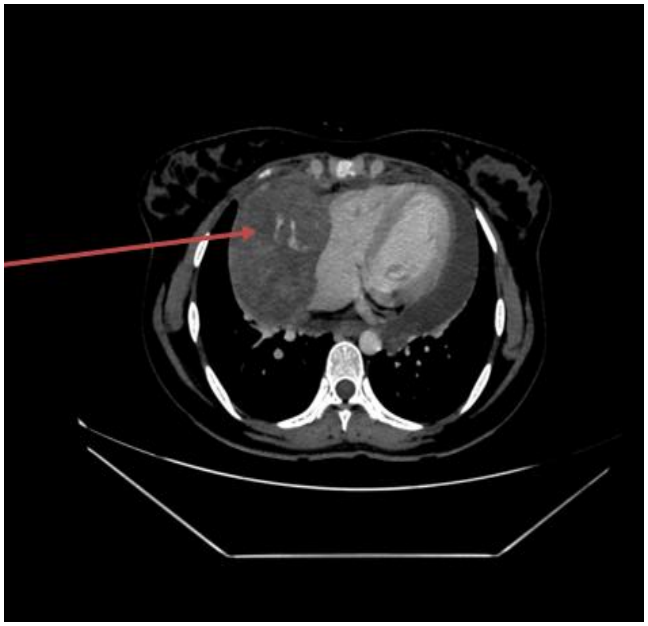


Figure 3 (b)

On Post Contrast CT Study - The Lesion is Seen Encompassing the RCA

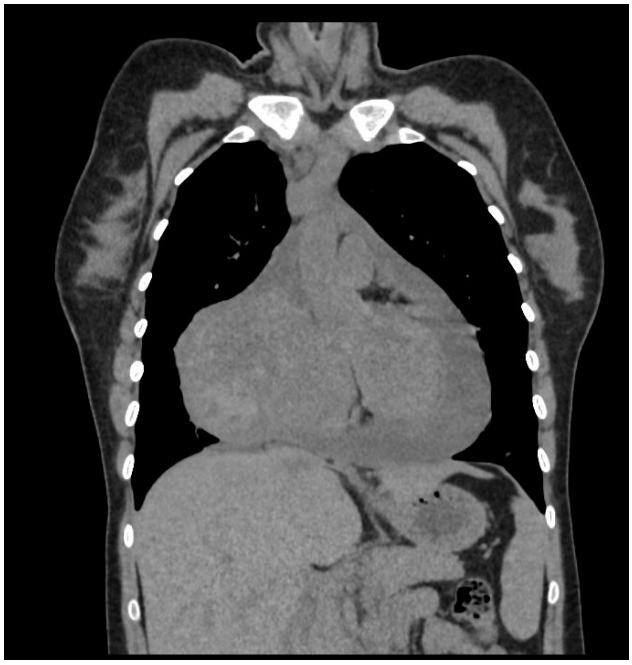


Figure 3 (c)
Figure 3

Chest computed tomography demonstrates a solid mass measuring 8.0 cm anteroposterior, 5.0 cm transverse, and 8.9 cm craniocaudal, involving the right para-cardiac region, not seen separately from the adjacent cardiac walls of the right atrio ventricular groove and proximal aspect of right ventricle with loss of intervening fat planes.

On Contrast Studies:

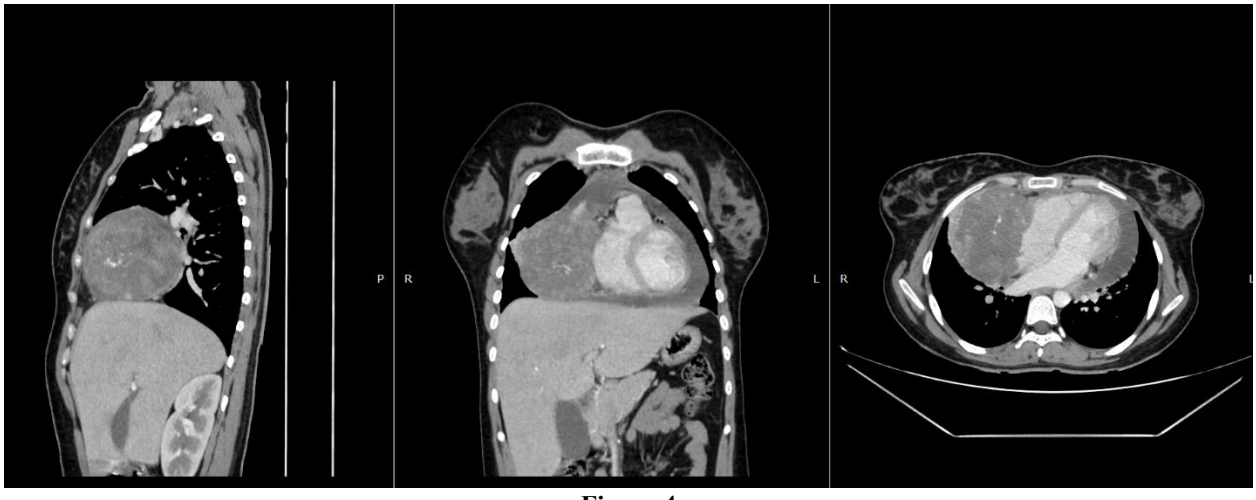


Figure 4

Ill-defined heterogeneously enhancing mixed soft tissue mass/ lesion is noted involving the right para-cardiac region. Few areas of hyperdensities of blood attenuation are noted within suggestive of hemorrhagic components.

<u>Cardiac MRI</u>:

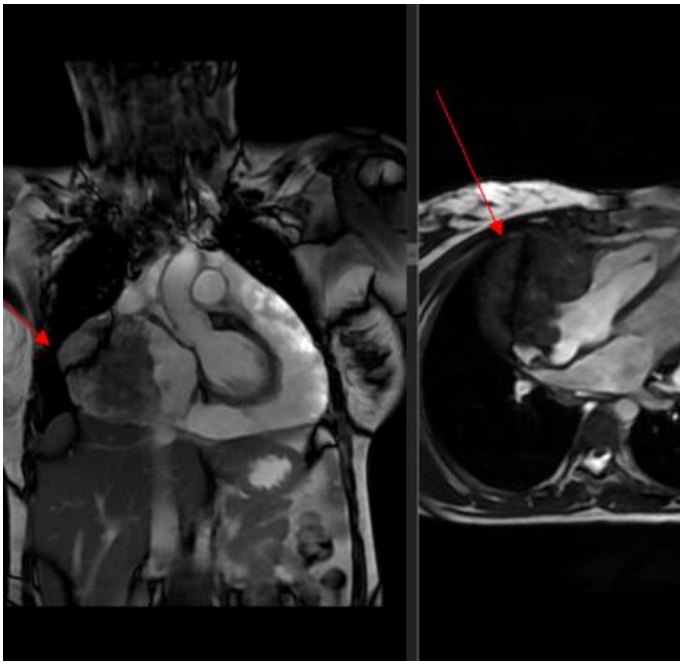
Findings:

A 8.9 x 7.2 x 8.0 cm (AP x TR x CC) sized ill-defined lobulated, non-mobile, heterogeneously enhancing mixed soft

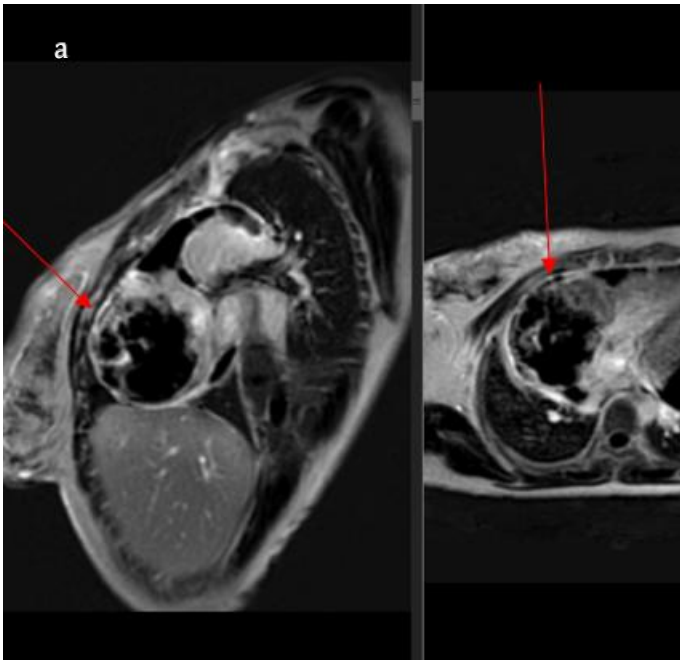
tissue mass is noted involving the anterior and lateral wall of the right atrium and the right atrial appendage It is seen splaying the parietal pericardium away from the right atrium and right ventricle. It is also seen extending into the right ventricle cavity involving the basal RV free wall and intraventricular groove also. The lesion is seen extending from D5 to D10 vertebral body levels. The mass lesion is seen abutting the superior vena cava in its anterior most aspect just before its entry into the right atrium

Rest perfusion and delayed images reveal: Heterogeneous echogenic enhancement of mass/lesion involving the right atrium showing a central non enhancing area which may represent necrotic/ cystic component.

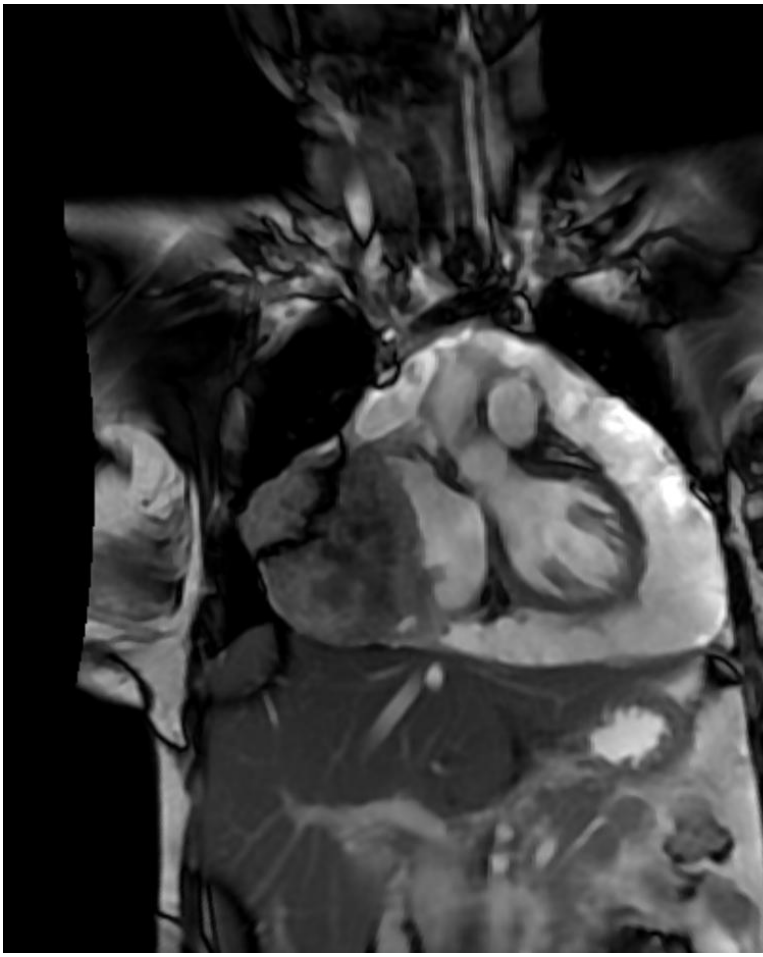
Further evaluation with Histopathological and PET-CT correlation was advised.



Pre-contrast MRI Study depicting Fig.5(a) – COR View; Figure 5(b) – 4 CH view; Figure 5(c) – 2 CH view



Post-contrast MRI Study depicting Fig.5(d) – 2CH View; Figure (e) – 4 CH View; Figure 5(f) – COR view



CMR: COR VIEW DEPICTING THE LESION AND PERICARDIAL EFFUSION

PET-CT

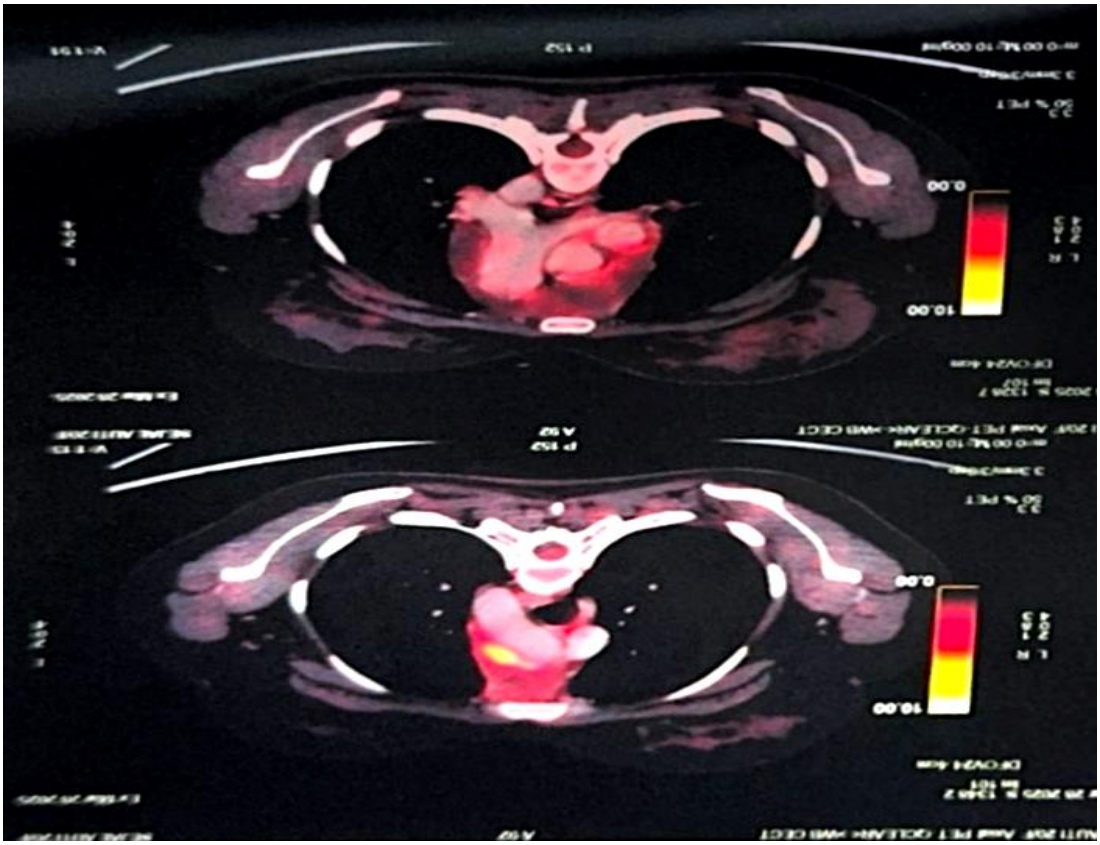


Figure 6

Findings:
FDG uptake is noted in heterogeneous mass lesion in the right para-cardiac region abutting the right atrium. The lesion is not seen separately from the right atrial wall. The lesion is seen extending from D5-D10 vertebral levels. The lesion is encasing the proximal aspect of SVC and abutting the root of aorta.

CT -GUIDED BIOPSY was performed and sent for HPE.

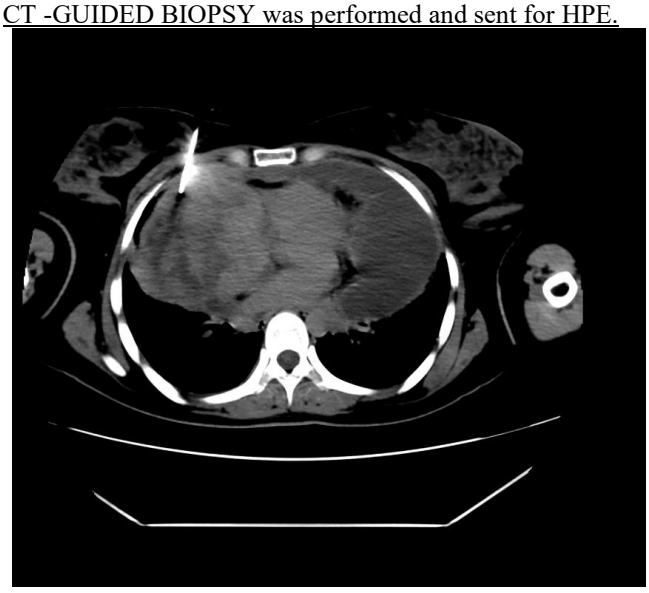


Figure 7a

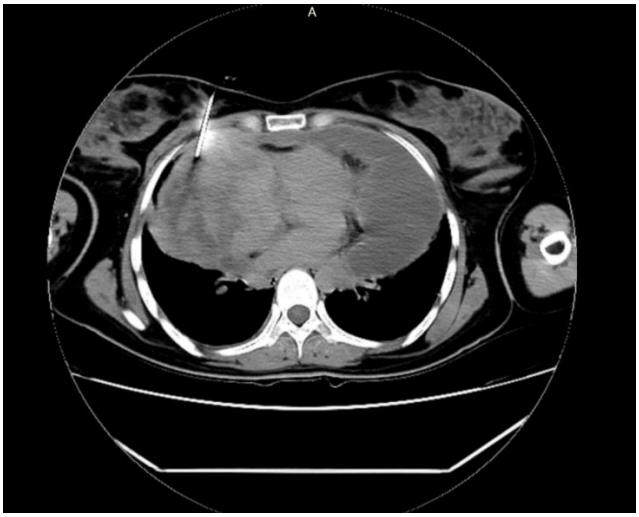


Figure 7b

Histopathological Findings:

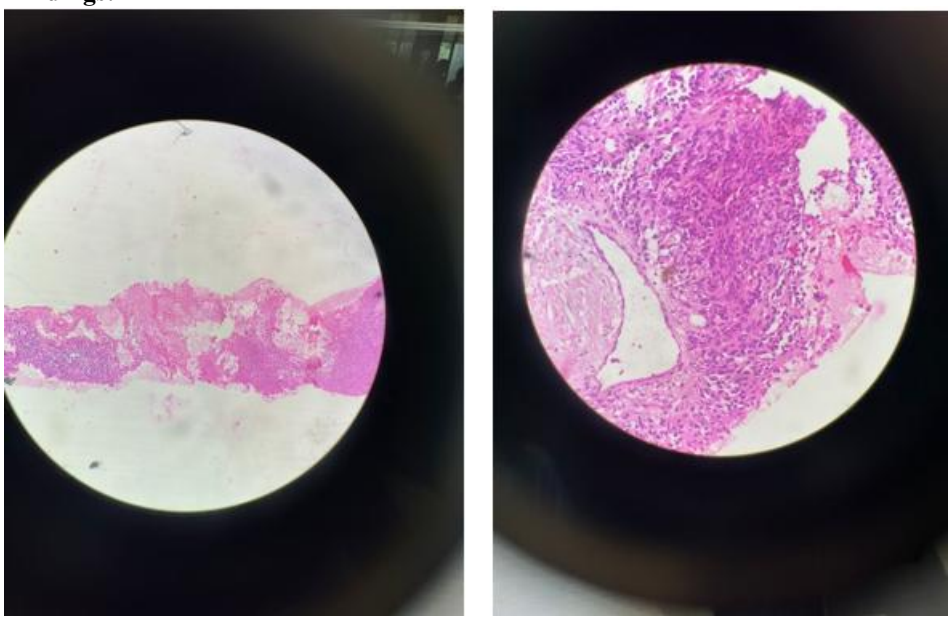


Figure 8

The histo-pathological microscopic finding showed malignant tumor composed of many anastomosing vascular channels and lined by pleomorphic endothelial cells, surrounded by areas of fibrosis, necrosis and hemorrhage. Individual tumor cells are round to oval with hyperchromatic oval to spindle nucleus and moderate cytoplasm.

Atypical mitosis was seen.

Immunohistochemistry:
CD34 -strong membrane positivity in the cells.

3. Conclusion

Here, we describe the case of a 20-year-old female patient with a heart tumor in the right atrium wall, which caused a large amount of pericardial effusion that invaded the surrounding area and is visible on transthoracic echocardiography, computed tomography, and magnetic resonance imaging. CT-guided biopsy of the lesion was performed and histopathological results confirmed this case as a right atrial angiosarcoma.

Primary cardiac angiosarcoma is rare, and its prognosis is extremely poor. The right atrium is the most common involved site, whereas peripheral invasion, including the pericardium, right coronary artery, tricuspid valve, and right ventricle, is universal.

Other differentials include:
1) Rhabdomyosarcoma
2) Extracardiac anterior mediastinal neoplastic etiology (less likely)

4. Discussion

Primary tumors of the heart are rare, with incidences ranging from 0.001% to 0.030% at autopsy. Among them only one-quarter are malignant. Most of the malignant tumors are sarcomas and around 40% are angiosarcomas. The main complaints in patients with angiosarcomas arise from: tumor mass obstruction, myocardial or atrial wall invasion and penetration which can cause arrhythmias and pericardial effusions, emboli, systemic symptoms like fever or weakness, metastases.

The diagnosis approach of angiosarcoma is based on TTE, CT, magnetic resonance imaging, histopathology and immunohistochemistry.

Cardiac magnetic resonance (CMR) and CT can provide more information about tissue characterization, precise location, and myocardial invasion.

In addition, CMR is helpful in differentiation of benign and malignant tumors. Before planning the treatment strategy, the malignancy and metastasis should be evaluated. (18)F-FDG positron emission tomography–computed tomography is a noninvasive preoperative test to reveal any potential metastases.

However, the definitive diagnosis and gold standard is cytology and immunohistochemistry. In the HE slide, angiosarcoma often shows abnormal mitosis with epithelioid, spindle, plasmacytic shaped cells and multiple prominent or bar-shaped nucleoli and chromatin strands.

Treatment guidelines for cardiac angiosarcoma are not yet established due to the rarity, but multidiscipline therapies including surgery, radiotherapy and chemotherapy are most commonly employed.

Surgery is the mainstay of angiosarcoma treatment.

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