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Cardiac Angiosarcoma: A Rare Cause for Syncope and Chest Pain

Geetha Narayanan[1], Anitha Mathews[2], Gayatri Gopan[3], Lali V Soman[4]
[1]Professor and Head of Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India
[2]Additional Professor of Pathology, Regional Cancer Centre, Trivandrum 695011, India
[3]Senior Resident, Regional Cancer Centre, Trivandrum 695011, India
[4]Medical officer in Medical Oncology, Regional Cancer Centre, Trivandrum 695011, India

***Corresponding author**
Geetha Narayanan
Email: geenarayanan@yahoo.com

Abstract: Primary cardiac tumors are rare and sarcomas account for 20% of all cardiac neoplasms. Cardiac angiosarcomas are rare rapidly progressive tumors with poor prognosis and often present a diagnostic challenge. We present the case of a 36 year old man who presented with syncope, chest pain and shortness of breath. ECHO and CT scan revealed a right atrial mass. He underwent debulking surgery, chemotherapy and radiation and was disease free for 1 year. Subsequently he recurred, received second line chemotherapy and achieved remission. However he recurred locally after another year and died at 2 years. It is important to diagnose these patients early since complete surgical resection offers the best result.

Keywords: Angiosarcoma; Cardiac, chemotherapy

INTRODUCTION

Primary cardiac tumors are rare, with an incidence of 0.0017% to 0.05% [1]. Most of them are benign, only 25% are malignant [2]. Sarcomas account for 20% of all cardiac neoplasms and angiosarcoma constitute 33% of cases[3]. The majority of cardiac sarcomas occur between the 3rd and 5th decade with a male predominance. Primary cardiac angiosarcoma is very rare. We present the case of a young man with primary cardiac angiosarcoma. This case is significant in view of the common symptom we encounter in clinical practice and the rare diagnosis.

CASE REPORT

A 36 year old man presented with syncope, chest pain and shortness of breath of 3 weeks duration. There was no history of coronary artery disease. Echo cardiogram showed a large intracardiac mass attached to the interatrial septum and projecting from right atrium (RA) to right ventricle compromising the inflow. Contrast enhanced computed tomogram (CT) scan of thorax showed a well defined soft tissue density mass 5.8x6.6x7.5cm with heterogenous enhancement and cystic areas in the RA and enlarging it. Debulking of the right atrial mass was done and subsequently he presented to us. Peroperatively, a mass was noted at the superior vena cava (SVC) RA junction which was filling the RA cavity and infiltrating the right atrial wall and adjacent pericardium.

On examination, he had a ECOG performance score of 3 and he was dyspnoeic. His hematology and serum chemistries were normal, and LDH was 466 U/L. ECHO showed a 2.5x2cm residual mass attached to right atrium. Left ventricular ejection fraction was 65%. CT scan of chest showed a residual lesion. Histopathology showed a neoplasm composed of cells lining well formed irregular anastamosing and sinusoidal vascular channels filled with RBCs, with intervening solid spindle cell areas, with brisk mitosis, extensive areas of necrosis and haemorrhage (Figure 1). Tumor cells were positive for CD31 and CD54 (Figure 2& 3).

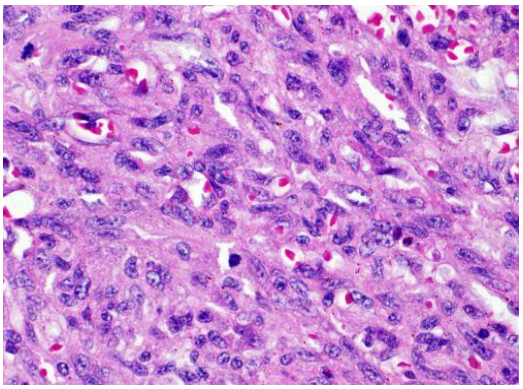

Fig-1: H&E x 400 - Spindle cells in sheets with formation of blood filled vascular channels. Mitotic figures are evident

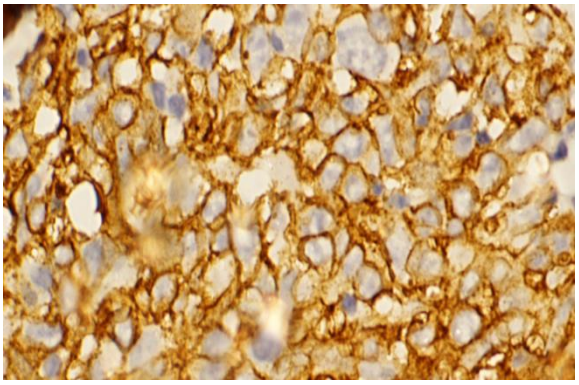

Fig-2: Immunohistochemistry showing the tumour cells strongly positive for CD31

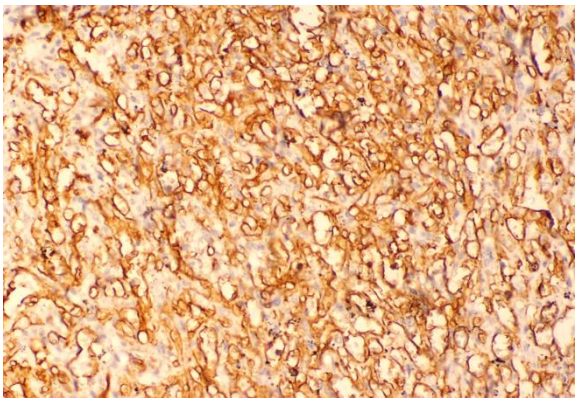

Fig-3: Immunohistochemistry showing the tumour cells strongly positive for CD34

This was diagnostic of high grade angiosarcoma of the right atrium. He received chemotherapy with ifosfamide 2gm D1 to D5 with mesna uroprotection and liposomal doxorubicin 60mg D1 for 6 cycles. The tumor resolved completely and he was consolidated with irradiation 36Gy/20# and 9Gy boost to primary site. He was disease free for 1 year after which his disease recurred locally. He was treated with paclitaxel 175mg/m^2 for 6 cycles and he achieved a second remission. However, his disease progressed and he died after 24 months.

DISCUSSION

Primary malignant tumors of the heart are rare, the most common are angiosarcoma followed by leiomyosarcoma, rhabdomyosarcoma, and osteosarcoma. Secondary cardiac neoplasms such as lymphomas, leukemias, melanomas, breast and lung cancers are 20-40 times more frequent than primary tumors.The low incidence of primary cardiac sarcoma is due to the low incidence of sarcoma in the general population and small weight of heart compared to muscles. Primary cardiac angiosarcoma is characterized by rapid growth, local invasion and distant metastasis and carries a very poor prognosis.

The common symptoms of cardiac sarcoma are dyspnoea on exertion, chest pain, cough, paroxysmal nocturnal dyspnoea, hemoptysis, embolic events, fever etc [4]. Malignant tumors are almost exclusively found in the right heart especially right atrium, in contrast to benign tumors which are located on the left side [5].

A 41 year old man with angiosarcoma of the right atrium underwent excision of the mass with reconstruction of right atrium and chemotherapy [6]. A 66-year old woman with a high-grade angiosarcoma of the right atrium and ventricle extending into the anterior superior mediastinum with lung metastasis was reported. She received chemotherapy and died after 16 months [3]. Another 63 year old man with a primary cardiac angiosarcoma presented with cardiac tamponade and was successfully resected, however he survived only for 4 months after diagnosis [7]. A 25-year-old man presented with pericardial effusion and echocardiographic evidence of an intracavitary right atrial mass which was confirmed to be angiosarcoma after complete surgical excision. He received chemotherapy and c-kit inhibitors and recurred at 18 months [8]. A review of 18 patients with cardiac angiosarcoma showed that 78% were male, 89% had disease originating from right atrium, 44% had localized or locally advanced disease. Treatment included resection in 44%, chemotherapy in 39% and radiation in 11%, median overall survival was 13 months [9].

Cardiac magnetic resonance imaging (MRI) is superior to CT for soft tissue characterization and for evaluating abnormalities intrinsic to the myocardium. On MRI, primary cardiac tumours are characterized by heterogeneity or isointensity on T1-weighted images and hyperintensity on T2-weighted images. Areas of increased signal intensity on T1 images denote the presence of blood products [10].

Cardiac sarcomas generally have a dismal prognosis with a median survival of only 6 months [5]. Given its low incidence, standard treatment guidelines do not exist, however current treatment of cardiac sarcomas is surgical resection. It is indicated in cases without evidence of metastasis and when the tumour is curatively resectable. Complete resection is rarely possible due to the aggressive nature of the tumour and the high prevalence of metastasis at diagnosis. Patients with complete resection have a survival of 24 months compared with 10 months in those with incomplete resection [11]. In cases where complete resection is achieved, local recurrence is common [2]. Complete resection of cardiac sarcoma is difficult, in view of the location and extent of involvement. Moreover, up to 80% of patients present with distant metastasis at diagnosis [12]. Early diagnosis is crucial for rapid, complete removal of the tumor. However, diagnosis can be a challenge, because cardiac angiosarcomas often remain clinically silent until the disease is advanced.

In general, recommendations for the treatment of nonmetastatic cardiac sarcoma include exploration

for local control of the primary tumor, to relieve obstructive symptoms and to prolong disease-free survival. In many cases, surgical resection has been used in combination with chemotherapy, with or without radiation therapy. Combined modality approach has been reported to be successful in few cases [13]. The dose of radiation for sarcomas is 60 to 65 Gy following complete resection. The role of adjuvant chemotherapy after surgically resected cardiac sarcoma remains controversial. There is some evidence to support adjuvant chemotherapy to relieve symptoms and prolong survival as part of the combined modality approach[11]. The common chemotherapeutic drugs are doxorubicin, ifosfamide, cyclophosphamide, dacarbazine, and paclitaxel. Our patient also had a disease free survival of 1 year with adjuvant chemotherapy and radiation. He achieved a second remission with second line chemotherapy and lived for another year before his disease progressed and he died. The response in this patient could be because of the debulking, radiation to residual disease and use of the most active drugs for angiosarcoma viz. paclitaxel, doxorubicin and ifosfamide.

The prognosis of cardiac angiosarcomas is poor due to the rapid local relapse and high incidence of systemic metastasis. The important prognostic factors are complete surgical resection, left-sided lesion, absence of necroses or metastases, and low mitotic count[4]. Alvarez et al.reported that the mean overall survival in patients with primary cardiac angiosarcomas was 5 months and on subgroup analysis, survival was 3-12 months in patients who underwent surgery plus adjuvant treatment, whereas 5 days to 7 months in those who underwent surgery only [14]. Heart and lung transplantation has been attempted, but limited experience resulted in poor outcome due to local recurrence and metastasis post-operatively [15].

CONCLUSION

Cardiac angiosarcomas are rare rapidly progressive tumours with poor prognosis and it often present a diagnostic challenge. It should also be considered in patients presenting with tamponade-type symptoms or recurrent pericardial effusions.It is an important to diagnose these patients early since complete surgical resection offers the best result.

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