Original Research Paper

Volume - 15 | Issue - 12 | December - 2025 | PRINT ISSN No. 2249 - 555X | DOI : 10.36106/ijar

Oncology

UNMASKING A SILENT THREAT – RARE CASE REPORT OF CARDIAC ANGIOSARCOMA

Dr. M. Vishwanath Senior Resident, Department of Medical Oncology, Madras Medical College/RGGGH, Chennai, Tamil Nadu, India.

ABSTRACT Primary cardiac angiosarcoma is an exceptionally rare yet highly aggressive malignancy, most commonly originating from the right atrium. We report the case of a 47-year-old male presenting with two weeks of chest pain and a paracardiac shadow on chest X-ray. Cardiac MRI demonstrated an 8 × 10.1 × 10.7 cm heterogeneously enhancing mass involving the right atrium and ventricle, encasing the right coronary artery and abutting major vessels. Histopathology revealed spindle-shaped cells with nuclear atypia and moderate eosinophilic cytoplasm, immunopositive for ERG and a Ki-67 index of 60%, confirming the diagnosis of cardiac angiosarcoma. Due to extensive local invasion, surgical resection was deemed unfeasible. The patient underwent concurrent chemoradiation with weekly paclitaxel (five cycles) and 59.4 Gy external beam radiotherapy, resulting in partial regression of the lesion (7.5 × 8.9 × 8.4 cm). The patient is currently under reevaluation for surgery. This case underscores the importance of multimodality management and highlights the potential role of paclitaxel-based chemoradiation in downstaging unresectable cardiac angiosarcoma.

KEYWORDS : Cardiac Angiosarcoma, Right Atrium, ERG, Paclitaxel, Concurrent Chemoradiation.

INTRODUCTION

Primary cardiac tumors are exceedingly uncommon, with an incidence of 0.001–0.03% in autopsy series. Among malignant types, cardiac angiosarcoma is the most frequent, typically arising from the right atrium and showing aggressive local invasion with early metastasis. Diagnosis is challenging due to nonspecific symptoms and limitations in imaging interpretation. Timely identification and multidisciplinary management are critical to improving outcomes in these highly vascular tumors. This report presents a case of locally advanced right atrial angiosarcoma managed with concurrent chemoradiotherapy and reviews relevant literature.

Case Report

A 47-year-old male from Chennai presented with chest pain for two weeks. He had no comorbidities, dyspnea, syncope, pedal edema, or B symptoms. Chest X-ray revealed a paracardiac shadow on the right side. Contrast-enhanced CT (CECT) of the chest showed a right atrial mass measuring 12 × 9.1 × 11.1 cm in the paracardiac mediastinal region with normal lung parenchyma and no mediastinal lymphadenopathy. Cardiac MRI demonstrated an 8 × 10.1 × 10.7 cm heterogeneously hyperintense lesion involving the right atrium and ventricle, extending to the right atrioventricular groove and protruding into the right atrial cavity. The mass encased the right coronary artery and showed loss of fat planes with the ascending aorta, superior vena cava, and suprahepatic IVC, with mild SVC compression. CT angiography confirmed infiltration of the pericardium and tricuspid annular region. Histopathological examination (HPE) of biopsy material revealed spindle-shaped tumor cells with moderate eosinophilic cytoplasm, nuclear hyperchromasia, karyorrhectic debris, and thin-walled vascular channels. Immunohistochemistry (IHC) showed strong nuclear ERG positivity (80%), Desmin negative, and Ki-67 index 60%, consistent with malignant vascular neoplasm – angiosarcoma. Routine blood investigations were within normal limits, and viral markers were negative. The case was discussed in a multidisciplinary tumor board involving Medical Oncology, Radiation Oncology, and Cardiothoracic Surgery (CTVS). Given the tumor's extensive invasion and inoperability, the consensus was to initiate concurrent chemoradiation therapy (CCRT). The patient received weekly paclitaxel (80 mg/m²) for five cycles concurrent with external beam radiotherapy (59.4 Gy/33 fractions, 1.8 Gy per fraction). Interim MRI after completion of treatment showed partial regression with the lesion reducing to 7.5 × 8.9 × 8.4 cm. He is currently on follow-up and under reassessment for surgical operability.

DISCUSSION

Primary cardiac angiosarcomas are the most frequent malignant cardiac tumors, with a male predominance (M:F ratio 3:1) and peak incidence in the third to fifth decades. Approximately 90% originate from the right atrium, often involving the atrial wall and pericardium. Symptoms are typically nonspecific—chest pain, dyspnea, or features of right heart failure. Because of their intracavitary growth and invasiveness, patients may develop pericardial effusion, tamponade, or arrhythmias. Echocardiography remains a useful initial tool, though MRI and CT angiography offer superior delineation of tumor extent and vascular invasion. Histopathology with IHC confirms diagnosis, showing positivity for CD31, CD34, ERG, and FLI-1 and negativity for epithelial markers. Due to the rarity of cardiac angiosarcoma, treatment protocols are based on case reports and retrospective series. Surgical resection offers the best chance of prolonged survival but is often infeasible due to extensive invasion. Chemotherapy regimens include taxanes, anthracyclines, gemcitabine, ifosfamide, platinum agents, and dacarbazine. Emerging data suggest that paclitaxel-based chemoradiation may provide tumor shrinkage and symptom control in unresectable cases.

CONCLUSION

Cardiac angiosarcoma, though rare, should be considered in patients presenting with unexplained right atrial masses. MRI and histopathology are critical for diagnosis. Multimodal therapy, including chemoradiation, offers potential benefit in downstaging inoperable disease and facilitating surgical intervention. This case demonstrates the promising role of weekly paclitaxel with radiotherapy in locally advanced cardiac angiosarcoma and underscores the need for collaborative management.

REFERENCES

1. Patel SD, Peterson A, Bartczak A, Lee S, Chojnowski S, Gajewski P, Loukas M. Primary cardiac angiosarcoma – a review. Med Sci Monit. 2014;20:103–109.
2. Look Hong NJ, Pandalai PK, Hornick JL, Shekar PS, Harmon DC, Chen YL, et al. Cardiac angiosarcoma management and outcomes: 20-year single-institution experience. Ann Surg Oncol. 2012;19(8):2707–2715.
3. Simpson L, Kumar SK, Okuno SH, Schaff HV, Porrata LF, Buckner JC, et al. Malignant primary cardiac tumors: review of a single institution experience. Cancer. 2008;112(11):2440–2446.
4. Isambert N, Ray-Coquard I, Italiano A, et al. Primary cardiac sarcomas: a retrospective study of the French Sarcoma Group. Eur J Cancer. 2014;50(1):128–136.
5. Lestuzzi C. Primary tumors of the heart. Curr Opin Cardiol. 2016;31(6):593–598.
6. Randhawa JS, Budd GT, Randhawa M, Ahluwalia M, Jia X, Daw H, et al. Primary cardiac sarcoma: 25-year Cleveland Clinic experience. Am J Clin Oncol. 2016;39(6):593–599.
7. Frezza AM, Benson C, Judson I, et al. Paclitaxel in angiosarcoma: a review of clinical activity and biological rationale. Crit Rev Oncol Hematol. 2017;119:67–74.
8. Wang JG, Li YJ, Liu H, Li NN, Zhao J, Xing XM. Primary cardiac angiosarcoma: a retrospective analysis of 6 cases and literature review. J Cardiothorac Surg. 2016;11(1):46.