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# Minimally invasive approach for the management of right atrial angiosarcoma; A case report

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## Abstract

Cardiac angiosarcoma is a rare primary cardiac tumor. Outcomes of minimally invasive resection of cardiac angiosarcoma are rarely reported in the literature. A male patient aged 28 years old presented with a right atrial mass compressing the superior vena cava and associated with pericardial effusion. Pericardiocentesis was done, and a preoperative workup revealed no distant metastasis. We planned excision of the mass through a right mini-thoracotomy approach. Intraoperatively, we found the mass invading the entire atrial wall thickness, and excision of the mass with a reconstruction of the right atrial wall was performed. Minimally invasive resection of atrial angiosarcoma could be feasible. Atrial angiosarcoma could present with vague signs and symptoms.

**KEYWORDS**

Cardiac angiosarcoma; Minimal invasive resection; Multidisciplinary approach



## Introduction

Primary cardiac tumors are a rare entity; one-fourth of them are malignant [1]. Cardiac sarcomas are the most common histopathological subtype, commonly arising from the right atrium (RA) [2]. Angiosarcomas are uncommon aggressive malignant neoplasms with a high rate of local recurrence and systemic metastases. This degree of malignancy renders the prognosis very poor, with high mortality within a few months of diagnosis [2]. The standard treatment is not well-established; however, surgical management is the preferred modality in fit patients [3]. Patients with primary cardiac angiosarcoma are usually asymptomatic; symptoms appear when the tumor reaches a certain size or when local or distant metastases occur [2]. Outcomes of minimally invasive resection of cardiac angiosarcoma are rarely reported in the literature. We present a case of right atrial angiosarcoma in a young male who had a minimally invasive tumor resection approach.

## Case Presentation:

A 28-years-old gentleman was presented with a two-month history of dyspnea on exertion (New York Heart Association (NYHA) class III), followed by chest pain upon lying flat and palpitations. The patient had no comorbidity and was healthy prior to this presentation. Echocardiography showed a pericardial effusion and RA mass. The patient had pericardiocentesis to relieve the dyspnea before further investigations. (Figure 1)

The chest computed tomography scan (CT) showed a large soft tissue mass from the RA with an adjacent pericardial thickening. The mass was compressing the superior vena cava. There was no

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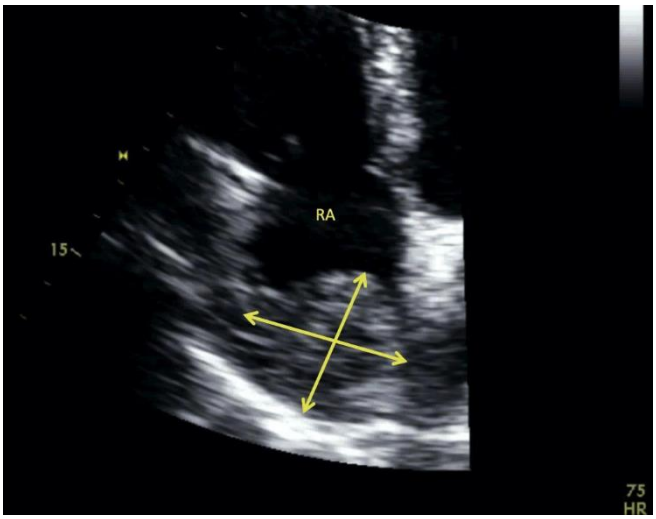


*Figure 1: ECHO view showing large homogeneous fixed echogenic mass with regular borders seen in the right atrium measuring 4.5 x 2.5 cm.*

mediastinal, axillary, or supraclavicular lymphadenopathy. Both lungs appeared normal with no intrapulmonary mass or nodule, and there was no destructive bony lesion.

The patient was scheduled to excise the mass through a minimally invasive right mini-thoracotomy with femoral cannulation. Intraoperative inspection revealed a large mass with a pericardial invasion involving the entire RA free wall and associated with extensive pericardial adhesion. The RA wall was resected entirely and sent for pathology. (Figure 2)

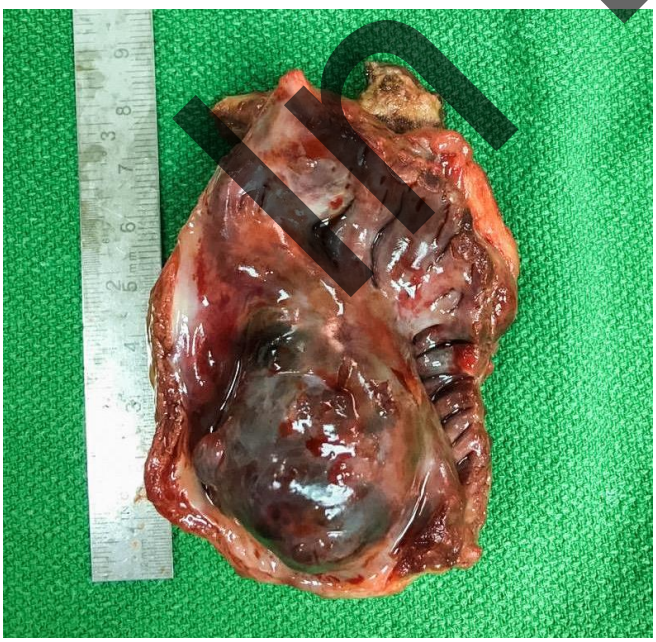

*Figure 2: Surgical specimen with the right atrial free wall with invasive mass and signs of necrosis*

The atrial wall was constructed using an autologous pericardial patch. Histopathology revealed an intracardiac mass that invaded the entire thickness of the atrial wall. There was no lymphovascular invasion; however, the tumor extended to the resection margin. This pathological finding was consistent with angiosarcoma (Figure 3). Staging workup showed no evidence of metastatic disease. The patient overall Eastern Cooperative Oncology Group (ECOG) performance scale was one.

The patient refused postoperative chemotherapy and discharged home after an uneventful postoperative course. A positron emission tomography (PET) scan was done two months after surgery and showed bone metastasis. Chemotherapy with adjuvant paclitaxel 80mg/m2 weekly and denosumab 120mg/monthly was started. After 12 cycles, PET CT showed partial response. A repeat metastatic workup six months later showed vertebral spine and liver metastasis. Palliative radiotherapy was applied, and the patient was started on the second line of chemotherapy with ifosfamide and doxorubicin.

## Discussion

Primary cardiac angiosarcoma has a poor prognosis, with a mean survival of 3.8 months without surgical resection [2]. Survival is affected by tumor size and tumor stage at the time of diagnosis and the presence of local or distant metastases. [2]. Surgical resection is widely recommended for localized tumors; it can improve survival [3]. Heart transplant has been utilized as a surgical option for primary localized cardiac sarcoma; however, it did not change long-term outcomes [4]. Although chemotherapy and radiation therapy benefits are still under investigation, an aggressive approach using a multimodality regimen with various combinations of surgery, chemotherapy, and radiation therapy has been proposed with some potential improvement of survival [2]. Some authors suggested neoadjuvant chemotherapy to decrease the tumor size aiming at total surgical resection. They have reported a median survival time of 27 months, and one patient lived for 9.5 years after surgery [4].

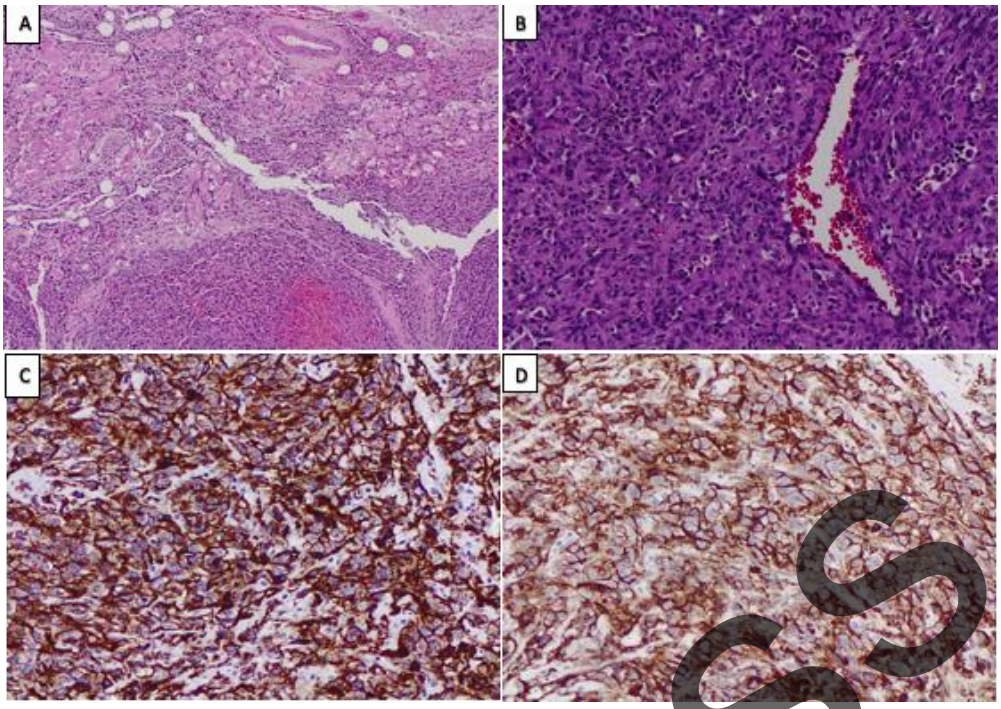


*Figure 3: Hematoxylin and eosin (H&E) histopathology and immunohistochemistry examination. (A): low power examination of large undifferentiated malignant neoplasm invading the adjacent cardiac wall (H&E: 4x). (B): Cardiac neoplasm composed of highly pleomorphic epithelioid cells, with a high nuclear to cytoplasmic ratio and prominent mitosis. Extravasated red blood cells are seen between tumor cells (H&E: 10x). (C): Neoplastic cells are diffusely and strongly positive for CD31 (10x). (D): Neoplastic cells are diffusely and strongly positive for CD34 (10x).*

Minimally invasive resections of cardiac angiosarcoma are rarely reported in the literature. In this case, we performed a minimally invasive left thoracotomy, and intraoperative inspection revealed a large tumor invading the pericardium. We think that this approach is feasible and does not increase surgical risk. Tumor invasion of the pericardium warranted postoperative chemotherapy. However, the patient refused further treatment and presented after a few months with metastatic disease. We believe that early diagnosis and intervention can change the fate of the disease. The feasibility of minimally invasive resection was demonstrated in another series of 225 patients; most of them were cardiac myxoma. They performed mini-thoracotomy in 12 patients and robotic resection in 60 patients. One patient had cardiac sarcoma and had a conventional approach [5].

## Conclusion

In conclusion, right atrial angiosarcoma could present with vague signs and symptoms, and minimally invasive resection is feasible.



**Conflict of interest:** Authors declare no conflict of interest.

## References

1. Look-Hong NP, Pandalai PK, Hornick JL, et al. Cardiac Angiosarcoma Management and Outcomes: 20-year single-institution experience. Ann Surg Oncol. 2012; 19: 2707–15
2. Naruse G, Kawasaki M, Yanase K, Tanaka T. Primary Angiosarcoma in the Right Atrium Diagnosed by a Cardiac Tumor Biopsy Using Intracardiac Echocardiography. J Med Ultrasound. 2020; 28(2): 120-122.
3. Bakaeen FG, Reardon MJ, Coselli JS, et al. Surgical outcome in 85 patients with primary cardiac tumors. Am J Surg. 2003; 186: 641–47
4. Uberfuhr PM, Meiser B, Fuchs A, et al. Heart Transplantation: An Approach to Treating Primary Cardiac Sarcoma? J Heart Lung Transplant. 2002; 21: 1135–39
5. Li S, Gao Ch. Surgical Experience of Primary Cardiac Tumor: Single-Institution 23-Year Report. Med Sci Monit. 2017; 23: 2111-2117.