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Low Grade Primary Leiomyosarcoma of the Right Atrium: Promising Survival with Complete Surgical Resection

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Abstract

Primary cardiac leiomyosarcoma has an extremely low incidence with overall median survival of approximately 6 months. Here, We report the case of a 60-year-old man who underwent complete surgical excision of right atrial mass. Histologic examination revealed leiomyosarcoma. The patient made a full recovery with no evidence of recurrence at 24 months. To the best of our knowledge, this is the first reported case of primary cardiac leiomyosarcoma in the Middle East.

Keywords: Primary leiomyosarcoma, Atrial mass, Cardiac surgery

1. Background

Differential diagnosis of cardiac tumors presents a challenge, and biopsy is the gold standard for histological confirmation. Published studies on primary cardiac tumors are still relatively limited due to their rarity. Most reported cases are benign, with the majority being myxoma [1]. Primary cardiac neoplasms are rare, and most confirmed cases are soft tissue sarcomas, with angiosarcoma being the most frequent histopathological type in adults. Leiomyosarcoma has an extremely low incidence, even among cardiac sarcomas [2].

2. Case Presentation

A 60-year-old man was admitted due to palpitation and chest discomfort. Initial examination revealed a regular pulse of 78 beats/min and blood

pressure of 121/74 mmHg. Nevertheless, the patient appeared cachectic (body mass index of 16.5 kg/m²). Electrocardiogram demonstrated a normal sinus rhythm with right bundle branch block. Chest radiography was unremarkable. Transesophageal echocardiography (TEE) showed a large (3.6 cm × 3.0 cm), heterogeneous, spherical, nonmobile mass in the right atrium (RA) attached to the interatrial septum (Fig. 1). There was no obstruction to blood flow, and cardiac chamber dimensions, pulmonary pressure and left ventricular function were normal. With these radiological features, a presumptive diagnosis of right atrial myxoma was made. Urgent surgery was performed with cardiopulmonary bypass using aortic and bicaval cannulation and single aortic cross-clamping. An incision was made in the RA, exposing the mass, which was occupying a large part of the RA. It was found to be attached to the interatrial septum at the base of the septal leaflet of the tricuspid valve between the anteroseptal commissure and coronary

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sinus (Fig. 2). However, the septal leaflet was freely mobile. Using sharp dissection, the mass was completely excised from the base. It appeared solid, unlike myxoma, and was sent for histopathological examination. The tricuspid valve was tested and appeared competent, and the RA was closed in double layers. The procedure was completed in the usual fashion. Subsequently, the patient was moved to the cardiovascular intensive care unit in a stable condition. A complete imaging workup was performed for a primary cause of the tumor and was negative with no evidence of metastasis. The case was discussed in the tumor board, and chemoradiotherapy was not recommended at that time. He was discharged from the hospital with regular follow-up at the outpatient clinic. After a follow-up of 24 months, the patient still has no clinical or radiological evidence of recurrence.

3. Pathological Findings

The resected mass was well circumscribed and lobulated. It measured $3.8 \times 3.5 \times 2$ cm. Upon

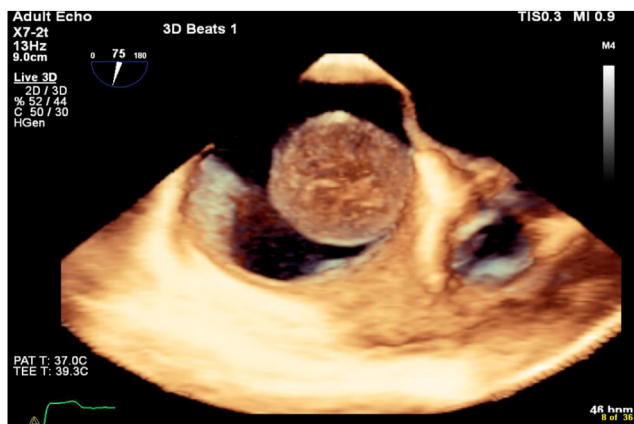


Fig. 1. TEE demonstrating right atrial mass.

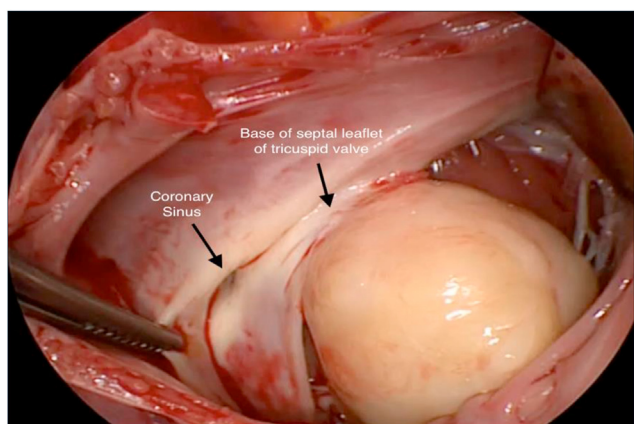


Fig. 2. Location of the tumor in the right atrium.

Abbreviations

TEE	Transesophageal echocardiography
RA	Right atrium

slicing, a gray-white homogeneous firm surface was revealed. Microscopically, it was composed of cigar-shaped spindle cells with eosinophilic and fibrillary cytoplasm, arranged in a fascicular growth pattern in a myxoid background. Scattered atypical cells were noted in multifocal areas (Fig. 3). There were no mitotic figures or necrosis. Immunohistochemical staining was positive for desmin, smooth muscle actin, and h-caldesmon. Considering the deep location and cellular atypia with myxoid stroma, a diagnosis of low-grade leiomyosarcoma was made.

4. Discussion

Few studies have clearly defined prognostic factors and outcomes of cardiac leiomyosarcoma, and there is limited knowledge on the diagnostic criteria due to its exceeding rarity [3]. Therefore, we depended only on morphological atypia in its diagnosis as leiomyosarcoma. Moreover, as observed in our case, myxoid changes can obscure the morphology. Wang et al. [4] reviewed 79 confirmed cases of primary cardiac leiomyosarcoma, of which most were diagnosed between the 4th and 6th decades of life. Further, no sex predilection was observed. The most common presenting symptoms were dyspnea, chest discomfort, and edema. However, arrhythmia-related symptoms, as observed in our patient, were reported in this review, which developed in <13% of cases. The left atrium was the most frequent location, observed in approximately 60% of cases. Tumors in the RA, as observed in our patient, developed in <16% of cases. Younger age at diagnosis and complete surgical resection were found to significantly impact the overall survival rate in the univariate analysis [4]. In the review by Nakashima et al., survival >24 months was observed in 17 cases after radical surgical treatment with or without adjuvant chemotherapy and radiotherapy [5]. Despite aggressiveness in the surgical approach, the prognosis of leiomyosarcoma remains controversial. The role of adjuvant chemotherapy and radiotherapy in prolonging survival is still controversial, and there are no statistically significant studies that examine their efficacy in addition to surgery due to the limited evidence on the treatment of cardiac leiomyosarcoma. However, Bakaeen et al. recommended that surgery with multimodal therapy could be helpful in enhancing

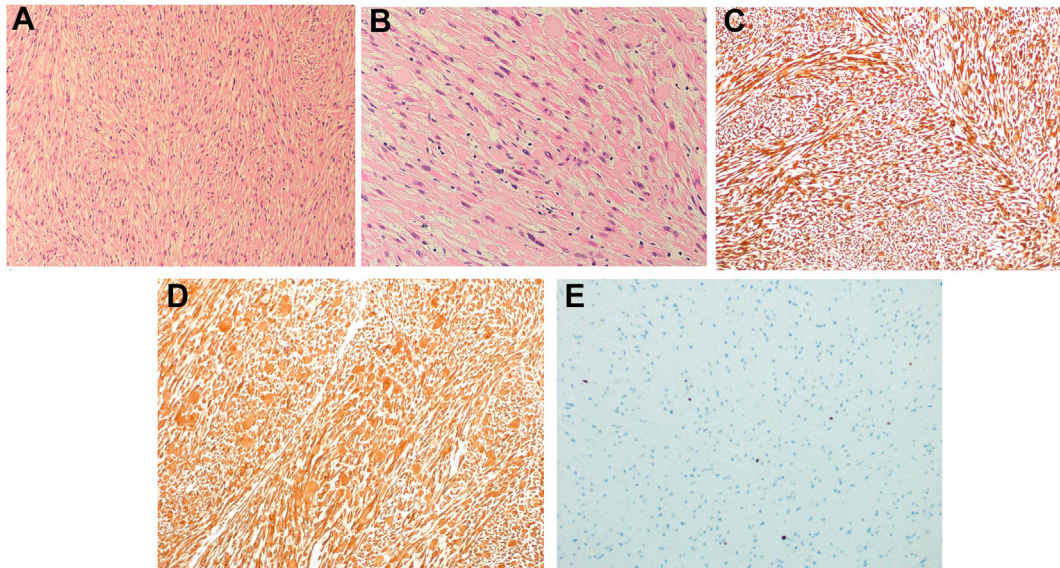


Fig. 3. Histopathological examinations: A: Leiomyosarcoma with spindle cell proliferation in intersecting fascicular pattern. B: Spindle cells with fibrillary eosinophilic cytoplasm and Cigar-shape nuclei. Mild cellular atypia is noted. C: The tumor cells show diffuse and strong immunoreactivity to smooth muscle actin (SMA). D: The tumor cells show diffuse and strong immunoreactivity to Desmin. E: Ki-67 proliferative index factor: < 2%.

survival in patients with cardiac sarcoma [6]. Recently, two cases with survival times of 9 and 8 years after combined surgical and adjuvant chemoradiotherapy were reported [7, 8]. These are the longest survival times reported to date, giving hope for promising outcomes in such cases. Generally, malignant cardiac tumors have a poor prognosis, with overall median survivals ranging from 6 months to a few years [9]. After complete surgical resection, our patient still has no evidence of recurrence for 24 months. We recommend that a multidisciplinary team of cardiac surgeons, pathologist, medical oncologists, and radiation oncologists should manage all cardiac leiomyosarcoma cases. There is wide agreement that surgery is the cornerstone of treatment when possible.

Learning Points

- 1- Not every right atrial mass is a myxoma.
- 2- complete resection should be considered when possible and may increase survival.

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Conflict of Interest

None declared.

References

- [1] Shapiro LM. Cardiac tumours: diagnosis and management. *Heart* 2001;85(2):218–22. <https://doi.org/10.1136/heart.85.2.218>.
- [2] Hoffmeier A, Sindermann JR, Scheld HH, Martens S. Cardiac tumors—diagnosis and surgical treatment. *Dtsch Arztebl Int* 2014;111(12):205–11. <https://doi.org/10.3238/arztebl.2014.0205>.
- [3] Careddu L, Foa A, Leone O, Agostini V, Gargiulo GD, Rapezzi C, et al. Primary Cardiac Leiomyoma Causing Right Ventricular Obstruction and Tricuspid Regurgitation. *Ann Thorac Surg* 2017;104(3):e231–3. <https://doi.org/10.1016/j.athoracsur.2017.03.068>.
- [4] Wang JG, Cui L, Jiang T, Li YJ, Wei ZM. Primary cardiac leiomyosarcoma: an analysis of clinical characteristics and outcome patterns. *Asian Cardiovasc Thorac Ann* 2015;23(5): 623–30. <https://doi.org/10.1177/0218492315574197>.
- [5] Nakashima K, Inatsu H, Kitamura K. Primary Cardiac Leiomyosarcoma: A 27-Month Survival with Surgery and Chemotherapy. *Intern Med* 2017;56(16):2145–9. <https://doi.org/10.2169/internalmedicine.6965-15>.
- [6] Bakaeen FG, Jaroszewski DE, Rice DC, Walsh GL, Vaporciyan AA, Swisher SS, et al. Outcomes after surgical resection of cardiac sarcoma in the multimodality treatment era. *J Thorac Cardiovasc Surg* 2009;137(6):1454–60. <https://doi.org/10.1016/j.jtcvs.2008.11.026>.
- [7] Glaoui M, Benbrahim Z, Belbaraka R, Naciri S, Errihani H, Lescene A. An uncommon long-term survival case of primary cardiac leiomyosarcoma. *World J Surg Oncol* 2014;12:338. <https://doi.org/10.1186/1477-7819-12-338>.
- [8] Muthukumar L, Jan MF, Taylor R, Crouch J, Neitzel G, Tajik AJ. Biatrial High-Grade Leiomyosarcoma With Nine-Year Post-Surgical Survival. *Ann Thorac Surg* 2017;103(1): e43–5. <https://doi.org/10.1016/j.athoracsur.2016.06.063>.
- [9] Orlandi A, Ferlosio A, Roselli M, Chiariello L, Spagnoli LG. Cardiac sarcomas: an update. *J Thorac Oncol* 2010;5(9): 1483–9. <https://doi.org/10.1097/JTO.0b013e3181e59a91>.