CASE REPORT

Vol. 11 No. 3 July-September 2026

doi: 10.35366/123484

Cardiac intimal sarcoma in the right atrium. Case report and literature review

Sarcoma intimal cardíaco en aurícula derecha. Reporte de caso y revisión de la literatura

Felipe G. Rendón-Elías, Cesar Castillo-Romero, Jorge Leyva-Villegas, Carlos M. Salas-Ríos, Urias Puentes, Arturo Carranza-Hernández and Luis H. Gómez-Danés

Department of Thoracic and Cardiovascular Surgery, University Hospital "Dr. José Eleuterio González". Monterrey, Nuevo León, México.

ABSTRACT

Primary cardiac intimal sarcoma is a very rare subtype of cardiac tumor that is often misdiagnosed. It can present in several ways and is known to mimic other conditions. Due to the advances in immunohistochemistry of the cardiac tumors each day is more frequent the diagnosis of cardiac intimal sarcoma. We present the case of a 42-years old patient, who was found to have a right atrial mass during the protocol of acute dyspnea in the emergency room, and after further investigation with enhanced computed tomography imaging confirmed the presence of right atrial mass that rule out the diagnosis of myxoma or cardiac thrombus. The objective of this article is to present the first case report in the country of the authors of an intimal cardiac sarcoma in the right heart and provide a comprehensive analysis of the medical literature about these rare primary cardiac sarcomata, including details on its pathogenesis, clinical presentation, current treatments, prognosis, and follow-up.

Keywords: heart surgery, dyspnea, cardiac sarcoma, immunohistochemistry, Murine Double Minute 2 Protein, genetic testing.

RESUMEN

El sarcoma intimal cardíaco primario es un subtipo muy raro de tumor cardíaco, que a menudo se diagnostica erróneamente porque este tumor cardíaco puede presentarse de diversas maneras y se sabe que es un excelente imitador de otras afecciones. Debido a los avances en la inmunohistoquímica de los tumores cardíacos, el diagnóstico de sarcoma intimal cardíaco es cada vez más frecuente. Presentamos el caso de un paciente de 42 años, que se encontró con una masa auricular derecha durante el protocolo de disnea aguda en el servicio de urgencias, y tras una investigación adicional con tomografía computarizada mejorada confirmó la presencia de una masa auricular derecha que descarta el diagnóstico de mixoma o trombo cardíaco. El objetivo de este artículo es presentar el primer caso diagnosticado de sarcoma cardíaco intimal del corazón derecho en el país de los autores y proporcionar una revisión exhaustiva de la literatura médica sobre estos sarcomas cardíacos primarios raros, incluyendo detalles sobre su patogénesis, presentación clínica, tratamientos actuales, pronóstico y seguimiento.

Palabras clave: *cirugía cardíaca, disnea, sarcoma cardíaco, inmunohistoquímica, proteína MDM2, pruebas genéticas.*

How to cite: Rendón-Elías FG, Castillo-Romero C, Leyva-Villegas J, Salas-Ríos CM, Puentes U, Carranza-Hernández A et al. Cardiac intimal sarcoma in the right atrium. Case report and literature review. Cir Card Mex. 2026; 11 (3): 104-110. https://dx.doi.org/10.35366/123484

Received: 06-10-2025. Accepted: 27-11-2025.

Correspondence: Dr. Felipe G. Rendón-Elías. E-mail: felipe.rendonels@uanl.edu.mx

Abbreviations:
CT = Computed Tomography
MDM2= Murine Double Minute 2 Protein
TEE = Transesophageal Echocardiography
TTE = Transthoracic Echocardiography
UEAs = Ultrasound-Enhancing Agents

Primary cardiac tumors are very uncommon, can be primary or metastatic, occurring in about 0.001-0.3% of autopsies[1] and diagnosis may be difficult, as symptoms and cardiac imaging may mimic other cardiac diseases. They can be classified into three clinicopathological groups: 1) benign congenital tumors; 2) benign acquired tumors; and 3) malignant tumors.[2] Nearly 75-90% of primary cardiac tumors are benign (myxoma the most common), and 10-25% are malignant (angiosarcoma is the most common type of sarcoma). Cardiac sarcomas account for 75-95%, with a severe prognosis, having a five-year survival rate of 14%.[3] Generally, sarcomas represent a heterogeneous group underlying a variety of aetiologies and anatomical features.[4] Intimal sarcomas are extremely rare mesenchymal tumors and is a subtype of sarcoma that occurs more commonly in the great vessels and pulmonary veins but rarely involves the heart and less common in the right heart.[5]

The objective of this article is to present the first case report in the country of the authors of an intimal cardiac sarcoma in the right heart and provide a comprehensive analysis of the medical literature about these rare primary cardiac sarcomata, including details on its pathogenesis, clinical presentation, current treatments, prognosis, and follow-up.

CASE REPORT

A 42-year-old male patient with past medical history of hypertension, non-insulin dependent diabetes mellitus, obesity, dyslipidemia, history of smoking (eight packs/year), and family history of colon and breast cancer, was referred to our emergency room for evaluation of sudden rest dyspnea without thoracic pain accompanied by constitutional symptoms. The patient referred a two-month history of progressive worsening effort-related dyspnea, fatigue and dry cough. On admission, he was unstable, with an arrhythmic heart rate (150 bpm), respiratory rate of 30 breaths/minute, and body temperature of 36.5 °C, the blood pressure and the peripheral oxygen saturation were not possible measure (because of the arrythmia). The electrocardiogram showed atrial fibrillation with high ventricular response that was treated by cardioversion. With the patient hemodynamically stable a transthoracic echocardiography was performed, which revealed presence of a well-defined right atrial mass of approximately 7 × 5 cm, attached to the auriculo-ventricular septum prolapsing to the right ventricle *(Fig. 1)*. The patient was subsequently referred for a contrast enhanced computed tomography (CT) scan to further visualize the mass and rule out pulmonary embolism. The CT revealed a well-circumscribed mass within the right atrium, measuring 7 × 5 cm fixed in the auriculo-ventricular septum near the tricuspid valve which was partially occluded by the mass and pass through it to the right ventricle *(Fig. 1)*, pericardial and peritoneal effusion accompanied it.

The patient was subsequently translated to intensive care unit and presented to cardiac surgery team that decided to program the patient to realized resection of the cardiac mass. An intraoperative transesophageal echocardiography performed prior to placing the patient on Cardiopulmonary Bypass (CPB) revealed a 6 × 10 cm, right atrial mass attached to the septum interatrial that prolapsed the tricuspid valve.

In the operating room after a median longitudinal sternotomy, cardiopulmonary bypass under normothermia was routinely instituted and Del Nido cardioplegic solution was administered via the aortic root after aortic cross clamping. A right atriotomy was used to expose and realize the resection of the cardiac tumor. The tumor was glistening, white, hard,

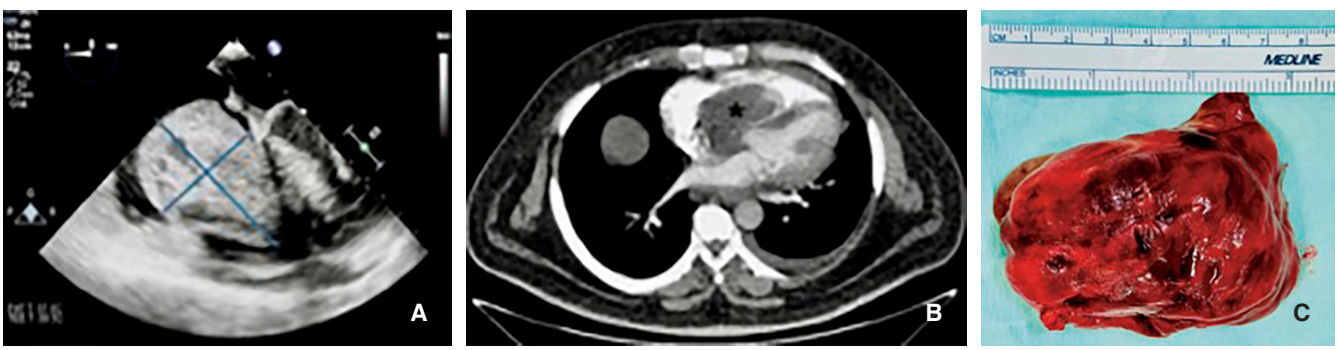

Figure 1: Preoperative echocardiogram showing the tumor in the right atrium (crisscrossing lines) prolapsing to the right ventricle **(A)**. Computed tomography image demonstrating a large non-enhancing, irregularly shaped mass with homogenous central hypodensity, originating from the fossa ovalis (asterisk) and protruding into the ventricle **(B)**. Intraoperative photograph of the excised specimen **(C)**.

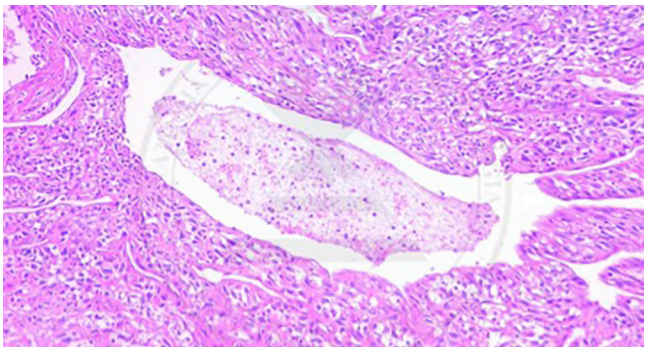

Figure 2: A delineated endothelio space is observed, surrounded by pleomorphic epithelioid and spindle cells (hematoxylin and eosin, ×200).

smooth, not lobed, adherent to auriculo-ventricular septum, and prolapsing to tricuspid valve. The tumor was resected circumferentially *en bloc*. The tumor did not appear to invade another cardiac chamber, the myocardium and any major vessel. The patient during the postoperative period showed an atrial fibrillation and right heart failure that was treated without complications.

The patient's resected cardiac tumor was sent to pathologic examination in formalin. The macroscopy appearance of the resected tumor was ovoid, smooth surface, light brown color, hard consistency, measured 7 × 5 × 5 cm and weight of 100 grams. On section, the tumor was solid-cystic, light brown and areas of hemorrhage *(Fig. 1)*.

Microscopically, the mass was composed of atypical spindle cells disclosing the presence of sarcoma with areas of necrosis and chondroid differentiation. Immunohistochemical analysis revealed positive expression of Murine Double Minute 2 Protein (MDM2) and focal positivity for smooth muscle actin and sarcomeric actin in neoplastic cells. Combined morphological, immunological and molecular studies performed on the resection specimen were consistent with a diagnosis of primary cardiac intimal sarcoma *(Fig. 2) (Fig. 3)*.

The postoperative course was uneventful, and the patient was discharged home on the 11th postoperative day and immediately started the oncologic protocol. Seven months after the surgery the patient does not present tumoral activity, Eastern Cooperative Oncology Group (ECOG) 1; Visual Analogue Scale for pain (VAS) 0, and received six doses of chemotherapy with Adriamycin.

COMMENTARY

Cardiac tumors have been reported since the 16th century however due to the incidence of malignant cardiac tumors are a very rare, they still pose a unique diagnostic and therapeutic challenge. Cardiac masses and tumors have always fascinated pathologists and cardiovascular physicians/surgeons alike. Before the mid-1950s, primary cardiac tumors were usually reported at autopsies. With the development of echocardiography, the imaging and diagnosis of tumors became a possibility. With advancements in cardiopulmonary bypass, their resection and thus a cure became an exciting reality. The first successful resection of a cardiac tumor was carried out by Crafoord in 1954,[6] but the experience in treating these aggressive and lethal tumors is minimal and management protocols are not well defined.

Primary cardiac tumors are rare; the reported prevalence is between 0.0017 and 0.028%[7] that has increased in recent decades. Primary cardiac tumors occur along a pathologic spectrum that includes benign, malignant, and intermediate with uncertain biological behavior. Cardiac sarcomas affect more men than women at a 2.5:1 ratio, usually presenting in the third and fourth decades of life.[8]

In 2015, the World Health Organization (WHO) wrote the classification of cardiac neoplasms including benign tumors, tumor-like lesions, malignant tumors, and pericardial tumors. Cardiac tumors are divided into primary and secondary forms.[9] Updates to each of the main categories of cardiac tumors were made in the fifth edition in 2021,[10] resulting in some entities being combined with others and some being newly established. Malignant primary cardiac neoplasms have been an area of growing controversy in the past decade. As detailed subsequently, the entities of intimal sarcoma and undifferentiated pleomorphic sarcoma have been formally separated, reflecting a better understanding of their biology and a better reflection of their anatomical sites of occurrence. Intimal sarcoma has been relocated to the Lung Section of the Thoracic Volume and the Intimal Sarcoma chapter of the WHO Classification of Soft Tissue and Bone Tumors.[11] Although many sarcomas have been described in the heart, only a few are encountered with regularity.

Approximately 10% of primary cardiac tumors are malignant and 90% benign.[12] Myxomas account for

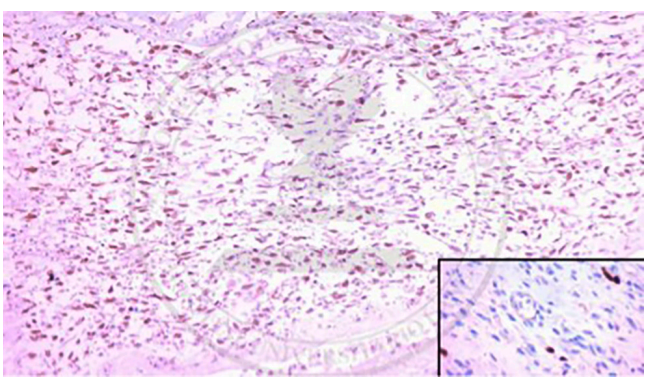

Figure 3: Murine double minute 2 protein immunohistochemistry marker positive.

approximately 50% of all benign cardiac tumors in adults and only for a small percentage in children. Rhabdomyoma is the most common benign tumor in children, accounting for 40 to 60% of the cases. Other benign cardiac tumors that have been described include fibromas, lipomas, hemangiomas, papillary fibroelastomas, cystic tumors of the atrioventricular node, and paragangliomas. The remaining 10-20% of primary cardiac tumors are malignant and usually are pathologically described as sarcomas.[13]

Primary cardiac sarcomas constitute approximately 1% of all soft tissue sarcomas and are the most common malignant primary cardiac tumor.[14] Angiosarcomas and unclassified sarcomas account for approximately 76% of all cardiac sarcomas, of which angiosarcomas are the most common. Rhabdomyosarcoma is the most common form of cardiac sarcoma in children. Leiomyosarcoma, synovial sarcoma, osteosarcoma, fibrosarcoma, myxoidsarcoma, liposarcoma, mesenchymal sarcoma, neurofibrosarcoma, and malignant fibrous histiocytoma are other cardiac sarcomas observed. Most cardiac sarcomas have been published as single case reports or institutional small series, with only a few studies examining large sample sizes.[15]

Different histological subtypes of cardiac sarcomas include, but are not limited to angiosarcoma, leiomyosarcoma, fibrosarcoma, rhabdomyosarcoma, synovial sarcoma, osteosarcoma, undifferentiated pleomorphic sarcoma and intimal sarcoma.

Intimal sarcoma is a mesenchymal tumor that is more commonly encountered in the great vessels and pulmonary veins. It rarely involves the heart and less frequent the right atrium. To our knowledge, our case is the first cardiac intimal sarcoma reported in our country and around the world no more than thirty cases have been reported till date.[16]

Intimal sarcoma are poorly differentiated malignant tumors characterized by the formation of tightly packed spindle shaped cells with fascicular growth pattern, and often show areas of necrosis. These tumors can frequently resemble smooth muscle neoplasms such as leiomyosarcoma. They exhibit variable degrees of necrosis, cellular atypia and nuclear polymorphisms and rarely show areas with morphological features like angiosarcoma, rhabdomyosarcoma, and osteosarcoma. It was believed that angiosarcoma and undifferentiated pleomorphic sarcoma were the most common cardiac sarcomas but in recent study, intimal sarcoma has been reported as the most frequently occurring primary cardiac sarcoma (42%) with MDM2 gene amplification being its characteristic pathological feature. Immunohistochemical analyses of these tumors typically show positive reactivity for MDM2, osteipontin and vimentin. Variable positivity is seen for SMA, desmin, CD117, CD68, P53 and BCL-2. CD31, CD34, and factor VIII are typically negative, but may be positive in areas with angiosarcomatous differentiation.[17-19]

The cardiac tumors may be symptomatic or found incidentally during evaluation for seemingly unrelated problem or physical finding. Symptoms are usually related to its cardiac location producing mass effect interfering with myocardial function or blood flow, causing arrhythmias, cardiac valves disfunction, pericardial effusion with or without tamponade, and these are manifested by dyspnea, chest discomfort, pre-syncope, or syncope. Also, may be present like systemic signs and symptoms (fever, arthralgias, weight loss, fatigue among others) and thromboembolic pulmonary or systemic. Cardiac sarcoma can present in several ways and are known to be great mimickers of other conditions. Patients may often remain asymptomatic until advances stages or present with dyspnea, orthopnea, paroxysmal nocturnal dyspnea, or lower extremity edema. In our case, the patient had a history of dyspnea, and dry cough and the mass was seen on transthoracic echocardiography that was performed as a part of the emergency protocol for acute dyspnea and chest pain.[20,21]

For an appropriate diagnostic approach, the first step is differentiating between cardiac tumors and other cardiac masses such as thrombi or vegetations. Four steps should be followed to orient the diagnosis:[21] 1) the clinical setting provides critical diagnostic clues in helping establish the etiology of a cardiac mass or lesion and may be able to realize the differential diagnostic (cardiac thrombi secondary to atrial fibrillation, myocardial infarction or infective endocarditis);[22-27] 2) the tumor location, the intracardiac tumors can arise in every cardiac cavity or in every cardiac wall, even though each type of cardiac tumor prefers a well-defined location; 3) the histology-based likelihood and the age of the patient at the time of presentation; 4) the morphological and functional characteristics: size, shape, mobility, tissue characterization, vascular supply, and metabolic activity. Points 3 and 4 may be investigated using different imaging techniques: echocardiography, CT, magnetic resonance imaging, and positron emission tomography/computed tomography. Each technique has advantages and disadvantages in terms of costs, availability, and diagnostic power: often they must be used in combination.[28,29]

The role of multimodality imaging is very important and crucial in planning further evaluation and management. The goals of the initial evaluation are to ascertain the presence of a cardiac tumor, the location of the lesion within cardiac structures, and when possible, whether a tumor is benign or malignant.

Two-dimensional Transthoracic Echocardiography (TTE) is often the first diagnostic approach in terms of imaging modality because of its wide availability, low cost, and portability. TTE allows for the assessment of size, location, mobility, understanding the hemodynamic patron provoked by the tumor and the pericardial involvement of a tumor.

The limitations of TTE include poor acoustic windows, particularly in obese patients (like our patient) and those with chronic lung disease,[30] and lack of potential for tissue characterization. In addition, the extent and origin of the mass may not be distinguishable by echocardiography if it arises from outside of standard imaging views (e.g., in the superior vena cava or inferior vena cava or branch pulmonary vessels).

Transesophageal Echocardiography (TEE) is commonly used when a valvular lesion is suspected, particularly in patients with atrial masses or with mobile valvular lesions and is necessary to better characterize a cardiac tumor in terms of size, morphology, attachment site, extension, and hemodynamic affects. When used in combination with real-time three-dimensional echocardiography, TEE offers incremental value for the evaluation of intracardiac masses by providing a more accurate assessment of the anatomical relationships, size, and shape of the mass.[31]

The use of Ultrasound-Enhancing Agents (UEAs) in echocardiography is an important tool for the assessment of myocardial perfusion and can also be used for the evaluation of the relative perfusion of a cardiac mass. It allows for improved definition of intracavity structures and assessment of vascularity.[32] The difference in perfusion of cardiac masses may help distinguish between vascular and nonvascular tumors or thrombus because there are both qualitative and quantitative differences in perfusion among the various pathologies.[33] For example, malignant tumors, which are highly vascular due to abnormal neovascularization, demonstrate greater enhancement than the adjacent myocardium. In contrast, benign tumors (myxomas) often have poor blood supply and therefore demonstrate lower perfusion on visual inspection and have quantitatively less perfusion than the surrounding myocardium. Because thrombi are avascular, they do not perfuse with UEAs echocardiography. Therefore, UEAs echocardiography perfusion imaging may aid in the early identification of thrombi and guide subsequent diagnostic and/or treatment strategies.

Once a cardiac mass or tumor is suspected, patients may be referred for cardiovascular magnetic resonance, imaging study that offer a complete multiplanar and noninvasive evaluations of the mass (vascularity, adjacent infiltration) and its potential involvement with cardiac chambers and pericardium; because it also provides information about extracardiac structures and its surrounding anatomy, this often proves useful in surgical planning.[34]

Cardiac computed tomography has become increasingly used modality for the assessment of cardiac masses, especially when other imaging modalities are nondiagnostic or contraindicated, and is also useful in tumor staging because of its ability to detect metastases in cases suspected malignancies.[35]

In our case the diagnostic approach was to discard pulmonary embolism because of the clinical presentation (acute exacerbation of dyspnea and atrial fibrillation), and the most practical approach was to realize a TTE in the emergency and when the patient was more stable was taken to perform an CT. with this modality of imagines study the medical and surgical plan was designed. After the surgical procedure and with the histopathological diagnosis of intimal cardiac sarcoma the patient was sent to realize and CT and positron emissions tomography for staging malignancies.

Currently, there are no specific treatment guidelines for primary cardiac intimal sarcomas because of the rarity of these cases and their heterogeneous spectrum.[36] Surgical removal of the tumor (potentially associated with auto-transplantation or heart transplant) is still the best treatment with radical intent, in small tumors without loco-regional/distant spreading or severe impairment of cardiac function.[37-41] Nonetheless, the lack of symptoms may delay the diagnosis as the patient might remain for a prolonged period only under primary health care surveillance and thus, the opportunity window for a successful intervention might be missed. Additionally, chemotherapy, radiotherapy, immunotherapy, or molecular treatments with genomic agents are used as adjuvant/neoadjuvant lines, as an alternative to surgery, in addition to complete/incomplete tumor removal or in recurrent postoperatory cases.[42-45]

The prognosis of cardiac intimal sarcoma is generally poor. Factors that are associated with worse prognosis include necrosis, high mitotic count, and metastasis. Another important factor determining the prognosis is the anatomical location of the tumor within the heart (intracavitary vs intramural). It has also been found that the right-side tumors carry a worst prognosis than left sided tumors.[20,46,47]

Due to the frequent involvement of vital structures and early metastasis (up to 50% of patients at diagnosis), cardiac sarcomas are associated with high mortality and morbidity. Intimal sarcomas are highly aggressive with the mean survival being three months to one year, although survival up to 11 years has been reported. Patients with complete tumor resection are known to live twice as long as those without the surgical resection. Thus, early diagnosis and complete resection are of paramount importance in the management of intimal sarcomas.[48-50]

Although the achievement of tumor free margins with surgical resection is the mainstay of treatment in cardiac sarcomas and associated with improved survival, it is important to understand that surgical resection may not be possible in almost 50% of the cases due to the involvement of vital structures. However, in a recent study, about 65% of the patients with planned surgical management could undergo complete resection. Furthermore, local recurrence and metastasis occur frequently and early, usually within one year. Thus, there is a role for other treatment modalities such as chemotherapy and radiation therapy in the management of cardiac sarcomas. Ifosfamide-epirubicin

(or doxorubicin) and CyVADIC (cyclophosphamide, vincristine, doxorubicin, and dacarbazine) are the two main regimens used in adult soft tissue sarcomas and may confer survival advantage in patients with cardiac sarcomas as well.[51] Two meta-analysis have demonstrated that adjuvant chemotherapy improves the time to local and distant recurrence as well as the overall survival.[52,53] The addition of ifosfamide further improves these benefits, but these should be weighed against the toxic effects arising from its use. Gemcitabine based chemotherapy regimen (single agent or in combination with docetaxil) may also be tried in patients with cardiac sarcomas based on the favorable response to these drugs in various trials evaluating their role in the treatment of soft tissue sarcomas.[54] Apart from the conventional chemotherapeutic regimens, there could also be a role for tyrosine kinase inhibitor pazopanib in the treatment of advanced disease, especially in those with poor response to the abovementioned drugs. Pazopanib was approved in the United States in 2012 for treating patients with advanced soft tissue sarcomas who had received prior chemotherapy, based on the data from PALETTE trial.[55]

CONCLUSIONS

Cardiac intimal sarcoma is a rare, but aggressive disease entity with poor prognosis. Owing to its variable presentation, diagnosing requires a high degree of suspicion while encountering patients with characteristic findings on the imaging studies. Although surgical excision with tumor free margins is the mainstay of treatment, complete surgical excision may often not be possible because of the involvement of the vital structures. Owing to this entity could be very challenging, the high rates of recurrence and metastasis, adjuvant chemotherapy must be used in all the patients diagnosed with this disease. Finally, it cannot be stressed enough that early diagnosis and prompt treatment are extremely crucial in improving outcomes in patients with this tumor.

REFERENCES

1. Raphael KL, Martinez AP, Clements SD, Isiadinso I. Role of multimodal cardiac imaging in diagnosing a primary intimal sarcoma of the left atrial appendage. Tex Heart Inst J. 2019;46(1):28-31. doi: 10.14503/THIJ-16-5896.
2. Moeri-Schimmel R, Pras E, Desar I, Krol S, Braam P. Primary sarcoma of the heart: case report and literature review. J Cardiothorac Surg. 2020;15(1):104. doi: 10.1186/s13019-020-01157-4.
3. Thuraisingam A, Fletcher N, Mckay G. Primary cardiac intimal sarcoma. Ann Surg Case Rep. 2019;2(1):1013. doi: 10.1016/j.hlc.2019.02.113.
4. Valecha G, Pau D, Nalluri N, Liu Y, Mohammad F, Atallah JP. Primary intimal sarcoma of the left atrium: an incidental finding on routine echocardiography. Rare Tumors. 2016;8(4):6389. doi: 10.4081/rt.2016.6389.
5. Nomoto N, Tani T, Konda T, Kim K, Kitai T, Ota M, et al. Primary and metastatic cardiac tumors: echocardiographic diagnosis, treatment and prognosis in a 15-years single center study. J Cardiothorac Surg. 2017;12(1):103. doi: 10.1186/s13019-017-0672-7.
6. Chitwood WR Jr. Clarence Crafoord and the first successful resection of a cardiac myxoma. Ann Thorac Surg. 1992;54(5):997-8. doi: 10.1016/0003-4975(92)90676-u.
7. Patel J, Sheppard MN. Pathological study of primary cardiac and pericardial tumors in a specialist UK centre: surgical and autopsy series. Cardiovasc Pathol. 2010;19:343-352. doi: 10.1016/j.carpath.2009.07.005.
8. Oliveira GH, Al-Kindi SG, Hoimes C, Park SJ. Characteristics and survival of malignant cardiac tumors: a 40-year analysis of > 500 patients. Circulation. 2015;132:2395-2402. doi: 10.1161/CIRCULATIONAHA.115.016418.
9. International Agency for Research on Cancer, World Health Organization(WHO), International Academy of Pathology. WHO classification of tumours of the lung, plura, thymus and heart. 4a ed. Travis WD, editor. IARC; 2015.
10. Maleszewski JJ, Basso C, Bois MC, Glass C, Klarich KW, Leduc C, et al. The 2021 WHO Classification of Tumors of the Heart. J Thorac Oncol. 2022;17(4):510-518. doi: 10.1016/j.jtho.2021.10.021.
11. Thoracic tumours: WHO Classification of Tumors. WHO Classification of Tumours Editorial Board (editor). WHO editorial, 5th edition; 2021.
12. Basso C, Valente M, Poletti A, Casarotto D, Thiene G. Surgical pathology of primary cardiac and pericardial tumors. Eur J Cardiothorac Surg. 1997;12:730-737. doi: 10.1016/s1010-7940(97)00246-7.
13. Ekmektzoglou KA, Samelis GF, Xanthos T. Heart and tumors: location, metastasis, clinical manifestations, diagnostic approaches and therapeutic considerations. J Cardiovasc Med (Hagerstown). 2008;9:769-777. doi: 10.2459/JCM.0b013e3282f88e49.
14. 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. doi: 10.1002/cncr.23459.
15. Bussani R, De-Giorgio F, Abbate A, Silvestri F. Cardiac metastases. J Clin Pathol. 2007;60:27-34. doi: 10.1136/jcp.2005.035105.
16. Nistor C, Carsote M, Cucu AP, Stanciu M, Popa FL, Ciuche A, et al. Primary cardiac intimal sarcoma: multi-layered strategy and core role of MDM2 amplification/co-amplification and MDM2 immunostaining. Diagnostics (Basel). 2024;14(9):919. doi: 10.3390/diagnostics14090919.
17. Ibrahim A, Luk A, Singhal P, Wan B, Zavodni A, Cusimano RJ, et al. Primary intimal (spindle cell) sarcoma of the heart: a case report and review of the literature. Case Rep Med 2013;2013:461815. doi: 10.1155/2013/461815.
18. Butany J, Nair V, Naseemuddin A, Nair GM, Catton C, Yau T. Cardiac tumors: diagnosis and management. Lancet Oncol 2005;6:219-228. doi: 10.1016/S1470-2045(05)70093-0.
19. Neuville A, Collin F, Bruneval P, Parrens M, Thivolet F, Gomez-Brouchet A, et al. Intimal sarcoma is the most frequent primary cardiac sarcoma: clinicopathologic and molecular retrospective analysis of 100 primary cardiac sarcomas. Am J Surg Pathol 2014;38(4):461-469. doi: 10.1097/PAS.0000000000000184.
20. Gupta A. Primary cardiac sarcomas. Expert Rev Cardiovasc Ther. 2008;6(10):1295-1297. doi: 10.1586/14779072.6.10.1295.
21. Bruce C. 42-cardiac tumors. En: Otto CM, editor. The practice of clinical echocardiography. 5th ed. Elsevier; 2016.
22. Kraft C, Schuettfort G, Weil Y, Tirneci V, Kasper A, Haberichter B, et al. Thrombosis of the inferior vena cava and malignant disease. Thromb Res. 2014;134(3):668-673. doi: 10.1016/j.thromres. 2014.07.012.
23. Kojiro M, Nakahara H, Sugihara S, Murakami T, Nakashima T, Kawasaki H. Hepatocellular carcinoma with intra-atrial tumor growth. A clinicopathologic study of 18 autopsy cases. Arch Pathol Lab Med. 1984;108(12):989-992. doi: 10.1007/978-4-431-68177-9_4.

24. Wang Y, Yuan L, Ge RL, Sun Y, Wei G. Survival benefit of surgical treatment for hepatocellular carcinoma with inferior vena cava/right atrium tumor thrombus: results of a retrospective cohort study. Ann Surg Oncol. 2013;20(3):914-922. doi: 10.1245/s10434-012-2756-0.
25. Wassef J, Xu S. Hepatocellular carcinoma with tumor thrombus to the hepatic veins and the right atrium: a case report and review exploring various presentations and treatment options. Cureus. 2020;12(6):e8405. doi: 10.7759/cureus.8405.
26. Lee JM, Lee KW, Hong SK, Yoon KC, Cho JH, Yi NJ, et al. Unusual techniques for preserving surgical and oncologic safety in hepatectomy of advanced adrenal malignancy with vena cava and liver invasion. Ann Surg Oncol. 2018;25(11):3324-3325. doi: 10.1245/s10434-018-6545-3.
27. Sun N, Zhang J, Li B, Li A, Lv M, Zhang C. Favorable response to multimodal treatment in hepatocellular carcinoma with inferior vena cava and right atrial tumor thrombus and left adrenal gland metastasis: a case report and literature review. Medicine (Baltimore). 2021;100(49):e27987. doi: 10.1097/MD.0000000000027987.
28. Lestuzzi C. Primary tumors of the heart. Curr Opin Cardiol. 2016;31(6):593-598. doi: 10.1097/HCO.0000000000000335.
29. Tyebally S, Chen D, Bhattacharyya S, Mughrabi A, Hussain Z, Manisty C, et al. Cardiac tumors: *JACC CardioOncology* state-of-the-art review. JACC CardioOncol. 2020;2(2):293-311. doi: 10.1016/j.jaccao.2020.05.009.
30. Buckley O, Madan R, Kwong R, Rybicki FJ, Hunsaker A. Cardiac masses, part 1: imaging strategies and technical considerations. AJR Am J Roentgenol 2011;197(5):W837-W841. doi: 10.2214/AJR.10.7260.
31. Zaragosa-Macias E, Chen MA, Gill EA. Real time three-dimensional echocardiography evaluation of intracardiac masses. Echocardiography. 2012;29(2):207-219. doi: 10.1111/j.1540-8175.2011.01627.x.
32. Bhattacharyya S, Khattar R, Senior R. Characterisation of intracardiac masses by myocardial contrast echocardiography. Int J Cardiol. 2013;163(1):e11-e13. doi: 10.1016/j.ijcard.2012.06.098.
33. Kirkpatrick JN, Wong T, Bednarz JE, Spencer KT, Sugeng L, Ward RP, et al. Differential diagnosis of cardiac masses using contrast echocardiographic perfusion imaging. J Am Coll Cardiol. 2004;43(8):1412-1419. doi: 10.1016/j.jacc.2003.09.065.
34. Beroukhim RS, Prakash A, Buechel ER, Cava JR, Dorfman AL, Festa P, et al. Characterization of cardiac tumors in children by cardiovascular magnetic resonance imaging: a multicenter experience. J Am Coll Cardiol. 2011;58810):1044-1054. doi: 10.1016/j.jacc.2011.05.027.
35. Kassop D, Donovan MS, Cheezum MK, et al. Cardiac masses on cardiac CT: a review. Curr Cardiovasc Imaging Rep. 2014;7(8):9281. doi: 10.1007/s12410-014-9281-1.
36. Alama L, Agrawala K, Kankanala K, Fishberg R, Powell D. Primary cardiac undifferentiated high-grade intimal pleomorphic sarcoma: a case series report. Cardiol Res. 2020;11(2):129-133. doi: 10.14740/cr1029.
37. Tamaki T, Suzuki M, Umezu R, Utanohara Y, Mahara K, Takanashi S, et al. Clinicopathological features of bi-ventricular cardiac intimal sarcoma-Report of an autopsy case. J Cardiol Cases. 2017;16(4):113-115. doi: 10.1016/j.jccase.2017.06.001.
38. Hudzik B, Szkodzinski J, Zubelewicz-Skodzinska B, Gasior M. Primary sarcoma of the heart. Pol Arch Intern Med. 2017;127(10):694-695. doi: 10.20452/pamw.4121.
39. Lennerz C, O'Connor M, Schunkert H, Deutsch MA. A case report of primary cardiac sarcoma: A diagnostic and therapeutic challenge. Eur Heart J Case Rep. 2018;2(4):1-4. doi: 10.1093/ehjcr/yty143.
40. Beller JP, Maddalo S, Zamuco R, Axel L, DeAnda A, Balsam LB. Right ventricular undifferentiated pleomorphic sarcoma: a case report. J Cardiol Cases. 2015;13(2):60-62. doi: 10.1016/j.jccase.2015.10.008.
41. Muturi A, Kotecha V, Ruturi J, Muhinga M, Waweru W. High-grade spindle cell sarcoma of the heart: a case report and review of literature. J Cardiothorac Surg. 2015;29(10):46. doi: 10.1186/s13019-015-0245-6.
42. Mito JK, Mitra D, Doyle LA. Radiation-associated sarcomas: an update on clinical, histologic, and molecular features. Surg Pathol Clin. 2019;12(1):139-148. doi: 10.1016/j.path.2018.10.010.
43. Chen E. Pathogenesis of sarcomas. Diagn Pathol. 2016;01(01). doi: 10.4172/2476-2024.1000e103.
44. Salminen SH, Sampo MM, Bohling TO, Tuomikoski L, Tarkkanen M, Blomqvist CP. Radiation-associated sarcoma after breast cancer in a nationwide population: increasing risk of angiosarcoma. Cancer Med. 2018;7(9):4825-4835. doi: 10.1002/cam4.1698.
45. Orlandi A, Ferlosio A, Roselli M, Chiariello L, Spagnoli LG. Cardiac sarcomas: an update. J Thorac Oncol. 2010;5(9):1483-1489. doi: 10.1097/JTO.0b013e3181e59a91.
46. Burke AP, Cowan D, Virmani R. Primary sarcomas of the heart. Cancer. 1992;69(2):387-395. doi: 10.1002/1097-0142(19920115)69:2<387:aid-cncr2820690219>3.0.co;2n.
47. Ye N, Lan L, Hu H, Liu J, Xu H. Case report: The diagnostic challenge of primary cardiac intimal sarcoma. Front Cardiovasc Med. 2023;10. doi: 10.3389/fcvm.2023.1089636.
48. Blackmon SH, Patel A, Reardon MJ. Management of primary cardiac sarcomas. Expert Rev Cardiovasc Ther. 2008;6(9):1217-22. doi: 10.1586/14779072.6.9.1217.
49. Modi A, Lipnevicius A, Moorjani N, Haw M. Prolonged survival with left atrial spindle cell sarcoma. Interact Cardiovasc Thorac Surg. 2009;8(6):703-704. doi: 10.1510/icvts.2009.203562.
50. Fu B, Yu H, Yang J. Primary intimal (spindle cell) sarcoma of the left atrium. Echocardiography. 2015;32(1):192-4. doi: 10.1111/echo.12777.
51. Adjuvant chemotherapy for localised resectable soft-tissue sarcoma of adults: meta-analysis of individual data. Sarcoma meta-analysis collaboration. Lancet. 1997;350(9092):1647-1654.
52. Pervaiz N, Colterjohn N, Farrokhyar F, Tozer R, Figueredo A, Ghert M. et al. A systematic meta-analysis of randomized controlled trials of adjuvant chemotherapy for localized resectable soft-tissue sarcoma. Cancer 2008;113:573-581. doi: 10.1002/cncr.23592.
53. Maki RG, Wathen JK, Patel SR, Priebat DA, Okuno SH, Samuels B, et al. Randomized phase II study of gemcitabine and docetaxel compared with gemcitabine alone in patients with metastatic soft tissue sarcomas: results of sarcoma alliance for research through collaboration study 002. J Clin Oncol. 2007;25(19):2755-2763. doi: 10.1200/JCO.2006.10.4117.
54. Pautier P, Floquet A, Penel N, et al. Randomized multicenter and stratified phase II study of gemcitabine alone versus gemcitabine and docetaxel in patients with metastatic or relapsed leiomyosarcomas: a Federation Nationale des Centres de Lutte Contre le Cancer (FNCLCC) French Sarcoma Group Study (TAXOGEM study). Oncologist 2012;17(9):1213-1220. doi: 10.1634/theoncologist.2011-0467.
55. Van der Graaf WT, Blay JY, Chawla SP, Kim DW, Bui-Nguyen B, Casali PG, et al. Pazopanib for metastatic soft-tissue sarcoma (PALETTE): a randomised, double- blind, placebo-controlled phase 3 trial. Lancet. 2012;379(9829):1879-1886. doi: 10.1016/S0140-6736(12)60651-5.

Funding: none.

Disclosure: the authors have no conflict of interest to disclose.