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CASE REPORT

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A rare case of Capmatinib/Pikray sensitive metastatic intimal cardiac sarcoma

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Abstract

Primary cardiac tumors are infrequently encountered, with a higher prevalence of secondary tumors. Among primary cardiac tumors, myxomas are benign and more commonly observed, while angiosarcomas constitute the most frequently encountered malignant tumors. Intimal sarcomas, the least prevalent among primary cardiac tumors, are seldom reported. This case report details the unique instance of a patient diagnosed with spindle cell neoplasm, a type of intimal sarcoma mimicking atrial myxoma with distant metastases, manifesting complications such as new onset atrial fibrillation and heart failure. A male in his 70s, presented with subacute Dyspnea on minimal exertion, orthopnea, paroxysmal nocturnal dyspnea. Initial investigations, revealed new onset AFIB, heart failure with cardiomegaly and atrial masses suggestive of thrombi. Echocardiograpgy findings was consistent with atrial myxoma. Subsequent PET scan indicated malignancy with metastases to the spine. The patient opted for chemotherapy and palliative resection, which was incomplete with positive margins. Tumor biopsy revealed intimal sarcoma (spindle cell neoplasm) which was sensitive to Capmatinib and Pikray. Patient expired nine months later.

Keywords: Intimal sarcomas. Thrombi. Metastases. Atrial masses. PET scan. Capmatinib. Pikray.

Introduction

Cardiac tumors are rare, constituting less than 0.33% [1] of cases, and are classified as primary or secondary based on the origin. Primary tumors, originating from the heart, make up only 5% of cases, with sarcomas being the predominant malignant type. Among primary tumors, myxomas are common and benign, while sarcomas, particularly angiosarcomas, represent the majority of malignancies. Of all primary cardiac tumors, around 75% are benign, and 50% of these are myxomas, while the rest include fibroelastoma, lipoma, fibroma, hemangioma, rhabdomyoma, paraganglioma, and teratoma [2-4].

Secondary cardiac tumors are more common and are the results of metastasis from other organs [1]. Sarcomas account for 95% of the malignant tumors, with different histology observed. The most frequent types are angiosarcomas, undifferentiated sarcomas, and undifferentiated pleomorphic sarcomas, with percentages of 37%, 24%, and 11-24%, respectively [2].

In this case, it was emphasized the rarity and diagnostic challenges of primary cardiac tumors, specifically intimal sarcomas.

Methods

Study Design

The present study was prepared according to the rules of the CARE case report Available at: https://www.care-statement.org/. Accessed in: 11/11/2025. This case study was carried out after the patient's authorization through the Informed Consent Form. The patient's medical records were analyzed, correlating the findings with the literature.

Ethical Approval

This study was approved by the Research Ethics Committee of University of Abomey-Calavi, Cotonou, Benin, Africa.

Case report

Patient Information and Clinical Findings, Timeline, Diagnostic Assessment, Therapeutic Intervention, and Follow-up

A male in his early 70s with a medical history of hypertension, non-sustained ventricular tachycardia status post-AICD placement, and prior basal cell carcinoma of the forehead presented with a two-month history of dyspnea on minimal exertion, orthopnea, paroxysmal nocturnal dyspnea, productive cough with greenish sputum, pleuritic chest pain, and subjective fevers. On initial evaluation, vital signs were stable, and laboratory findings were grossly unremarkable. Chest X-ray (Figure 1) revealed pulmonary congestion, cardiomegaly, and moderate bilateral pleural effusions. Electrocardiogram (EKG,) showed new-onset atrial fibrillation with rapid ventricular response.

A CTangiogram of the chest demonstrated two well-circumscribed left atrial masses measuring 3.9 x 3.7 cm and 4.6 x 3.6 cm (Figure 2), suggestive of thrombus versus tumor. Additionally, a new 2.1 x 1.6 cm lytic lesion in the T11 vertebral body was noted. Transthoracic echocardiogram (TTE) revealed a dilated left atrium with a circumscribed obstructive mass suggestive of cardiac myxoma (Figure 3, with video link) and normal ejection fraction. He was admitted to the ICU for management of new onset atrial fibrillation and decompensated heart failure in the setting of a left atrial mass more likely to be a thrombus vs atrial myxoma. Despite minimal oxygen requirements, dyspnea persisted.

He was transferred to a tertiary facility for further evaluation. There, a PET scan showed FDG-avid left atrial masses and lytic lesions at L4 (Figure 4). The L4 lesion biopsy revealed a spindle cell neoplasm. Surgical resection of the cardiac mass was deferred due to suspicion of metastatic malignancy, and the patient was discharged. One week later, he returned with worsening dyspnea, irregular heartbeats, dizziness, and occasional vomiting of undigested food. He was hypotensive in the emergency department. Repeat CT angiography revealed significant interval enlargement of both left atrial masses (4.1 x 4.0 x 6.6 cm and 4.9 x 3.9 x 2.6 cm). The T11 lesion remained unchanged. He was readmitted for pulmonary edema and atrial fibrillation management. Subsequently, the patient developed hemoptysis and persistent hypotension. Palliative care consultation supported palliative surgical debulking.

He underwent open heart surgery at a tertiary center, where partial resection of the atrial mass was performed. Pathological examination revealed high-grade intimal sarcoma with spindle cell morphology, marked nuclear pleomorphism, high mitotic index (48 mitoses per 10 HPFs), and extensive necrosis (50%). Genomic testing revealed the presence of two actionable mutations: CAPZA-2-MET fusion and PIK3CA which were sensitive to Capmatinib and PIQRAY respectively along with amplification MDM2 (Figure 5). Postoperatively, the patient opted for chemotherapy. He underwent left femur rod placement surgery to prevent pathological fracture. Was initially on Capmatinib , but later changed to PIKray due to side effects. He survived an additional 11 months before passing away from progressive disease.

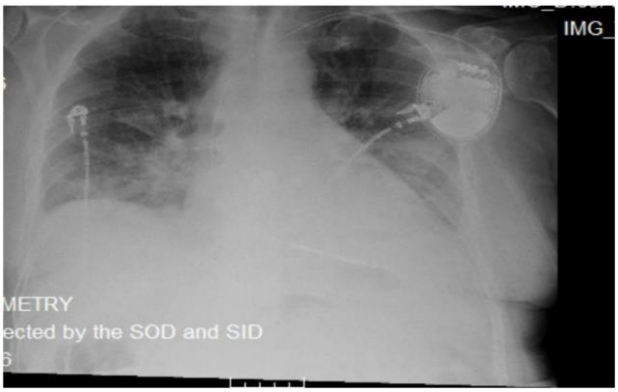

Figure 1. CXR showing cardiomagaly, pulmonary edema with B/L pleural effusions. Source: Own authorship

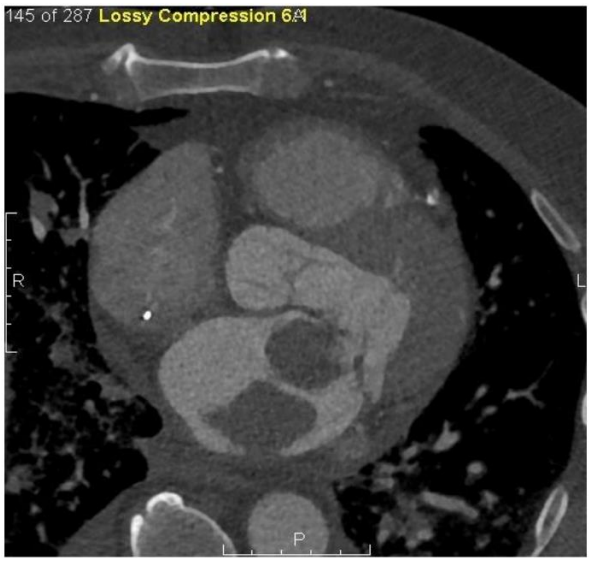

Figure 2. CT angiogram showing two tumors in left atrium. Source: Own authorship.

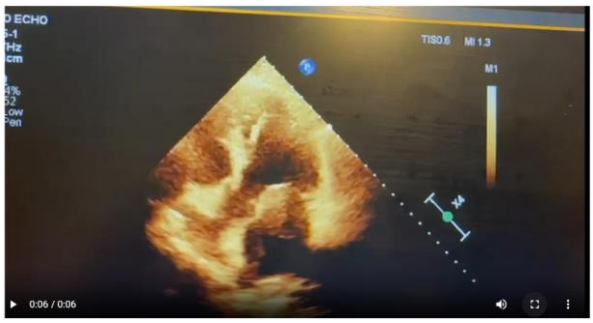

Figure 3. Transthoracic echocardiogram (TTE) revealed a dilated left atrium with a circumscribed obstructive mass suggestive of cardiac myxoma (Figure 3, with video link) and normal ejection fraction. VIDEO-2025-09-13-21-07-49.mp4 . Source: Own authorship.

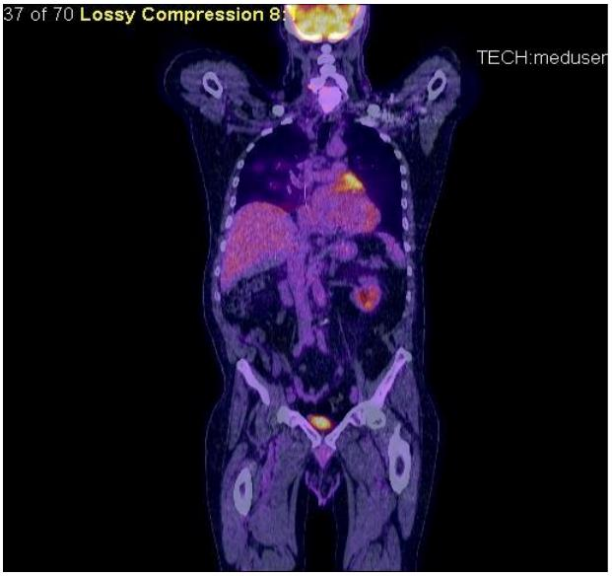

Figure 4. PET SCAN. Left atrium tumor with L4 metastasis. Source: Own authorship

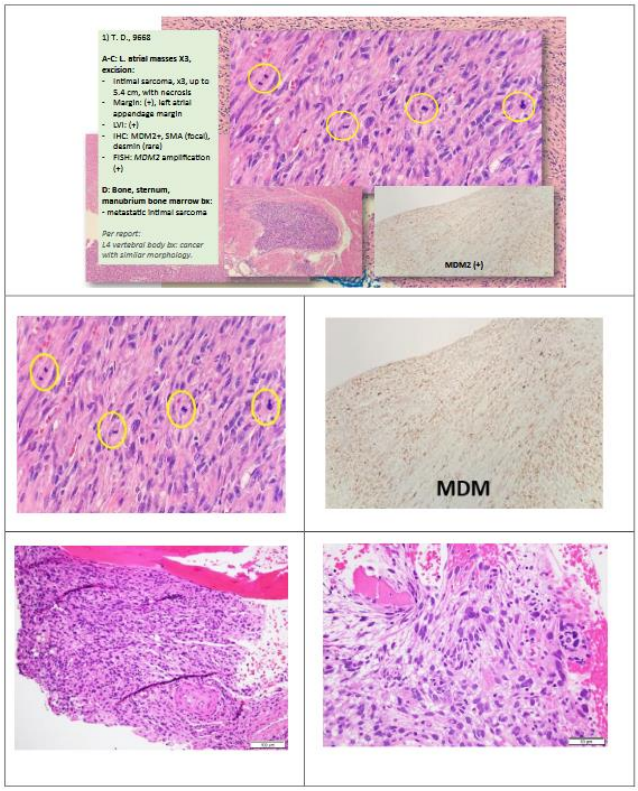

Figure 5. Genomic testing revealed the presence of two actionable mutations: CAPZA-2-MET fusion and PIK3CA which were sensitive to Capmatinib and PIQRAY respectively along with amplification MDM2. Source: Own authorship.

Discussion

Cardiac masses can be divided into two categories: neoplastic and non-neoplastic masses. Neoplasms refer to abnormal growths of tissue, which can be classified clinically as either benign or malignant [5]. These growths can also be further categorized as primary or secondary depending on their site of origin. Primary masses are those that arise directly from cardiac tissue, while secondary masses are those that migrate to the heart from nearby (mediastinal) or distant organs. Primary cardiac tumors can be either benign or malignant, whereas secondary masses are always malignant and are metastases [6].

Secondary masses are far more common [5]. Primary cardiac tumors are not common and their autopsy incidence ranges from 0.001% to 0.030% whereas metastatic cancer to the heart is far from uncommon, about twenty to forty times [6,7]. Among primary cardiac tumors, 25% are malignant, with 95% of these being sarcomas and the remaining 5% being lymphomas. The most prevalent type of cardiac sarcoma is angiosarcoma, accounting for 37% of cases [5]. Other types include undifferentiated sarcoma, malignant fibrous histiocytoma, leiomyosarcoma, and osteosarcoma. Some primary cardiac tumors are extremely rare, with intimal sarcoma, also called spindle cell sarcoma being the least reported [5].

Cardiac intimal sarcomas are endocardial mesenchymal tumors that are essentially encountered in great vessels like the pulmonary artery or thoracic aorta and are far less commonly seen in the heart. When originating in the heart, they are usually from the left heart [8,9] as opposed to angiosarcomas that occur in the right heart. Cardiac intimal sarcoma is not only very rare, underreported, but is very aggressive, therefore prognosis is usually very poor. Genomic profile tends to show recurrent 12q13-14 amplicon involving MDM2. The latter is almost always unique to cardiac intimal sarcoma, thus allowing differentiation from other subgroups of sarcomas [8].

Manifestations of the tumor are usually from intracardiac blockage and are dependent on many factors which include: the location, size, invasiveness, and how fast the tumor grows. Affected patients most often present with dyspnea, and constitutional symptoms among which are fever, fatigue, and weight loss. Further symptoms like splenic, renal infarcts, TIA/Stroke may be also encountered. Less often, patients may also present with arrhythmias/conduction disorders, heart failure and pericardial effusions. Unfortunately, most of the affected individuals remain asymptomatic until the later stages of the disease [10].

In this case report, we present a 71-year-old with a very aggressive intimal sarcoma with distant metastases to the spine. The patient presented in the late stage of the disease. The tumor was causing functional mitral stenosis, and he developed new onset atrial fibrillation and heart failure. Echocardiography findings was initially suggestive of atrial myxoma. The mass exhibited exponential growth within a matter of weeks, accompanied by uncontrolled atrial fibrillation and worsening heart failure.

Early diagnosis is largely dependent upon imaging techniques. The initial diagnostic workup should include Echocardiography (first-line imaging with sensitivity and specificity reaching 90% and 95% respectively) [7], contrast-CT, contrast MRI, and FDG-PET [5,7,11]. Most definitive diagnosis along with grading of the tumor requires a biopsy [7]. Recent studies have shown that patients with successful surgical resection with tumor-free margins have improved survival [10].

The average survival in patients with cardiac intimal sarcoma is 3-12 months [12]. Those that have successful tumor-free margin surgical resection live twice as long which explains how crucial early diagnosis can impact prognosis and therapeutic measures [7]. The average survival rate for patients treated with Chemotherapy and radiotherapy alone is less than a year [13,14]. The patient described in our case underwent multi-modality imaging including CT-angiography, Echocardiography, and PET-CT. Those were crucial in making an appropriate diagnosis. The patient had palliative surgery but without successful complete resection as the margins were not tumor-free. The surgery helped improve the patient's hemodynamics initially but quickly decompensated despite aggressive chemotherapy.

There are many complications associated with cardiac intimal sarcoma. Among them are heart failure, arrhythmias/conduction disorders, cardiac rupture, sudden cardiac death, and pericardial effusion [15]. Furthermore, necrosis, increased mitotic activity, metastasis, and intracavitary location are associated with worse prognosis. [10,16]. Our patient presented with complications and the biopsy of the tumor was positive with most factors that are associated with worse prognosis including but not limited to high mitotic activity, more than 50% necrosis and metastases to the spine. He expired within a year after the diagnosis.

Cardiac intimal sarcoma is an extremely rare and very aggressive tumor. Patients often present with constitutional symptoms; however, the majority are diagnosed in the late stages of the disease, often with complications such as metastases. Early diagnosis is crucial for improving survival rates, underscoring the importance of implementing appropriate diagnostic measures. Management is typically extrapolated from soft tissue sarcoma protocols and involves a combination of surgery, chemotherapy, and in select cases, targeted therapy.The mainstay treatment remains surgical resection to achieve tumor-free margins. The latter is more likely to happen if the tumor is caught in the early stage which is far from realistic except if the tumor is benign (Myxomas). When a tumor-free margin is achieved post-resection, the survival time is twice as long, and the quality of life is improved. For patients with positive margins, the addition of chemotherapy and radiation might be required even though the increase in survival remains uncertain. Currently, there are no specific, standardized treatment guidelines for primary cardiac intimal sarcomas due to their rarity and histological heterogeneity [17].

Chemotherapy is often used in both adjuvant and palliative settings, although its benefit is not well established. In recent literature, commonly used agents include anthracycline-based regimens, gemcitabine-based therapies, and pazopanib, a multi-tyrosine kinase inhibitor. Anthracyclines, in particular, have shown relatively better response rates in retrospective studies, though the overall prognosis remains poor. In our case, the patient was treated with Capmatinib, a selective MET inhibitor, and Piqray (alpelisib), a PI3K-alpha inhibitor. This regimen was selected based on molecular profiling reflecting the emerging role of targeted therapies in managing aggressive sarcomas. While evidence for these agents in intimal sarcoma is scarce, their use highlights the growing emphasis on personalized medicine in rare malignancies. Nonetheless, the patient experienced disease progression and ultimately succumbed to the illness, underscoring the urgent need for prospective clinical trials and molecular characterization to guide therapy in this challenging subset of cardiac tumors.

Conclusion

Intimal cardiac sarcomas are extremely rare and very aggressive, with a high rate of local recurrence and metastasis. Although surgical resection remains the mainstay of therapy, complete resection is often not feasible due to anatomic constraints and the infiltrative nature of the tumor. This case highlights the diagnostic and therapeutic challenges associated with these types of tumors. The absence of specific treatment guidelines and the tumor's variable presentation underscore the need for heightened clinical suspicion, early tissue diagnosis, and multidisciplinary management. Future research for targeted therapies and case aggregation are essential to better define optimal treatment strategies and improve patient outcomes.

CRediT

Author contributions**: Conceptualization**; **Data curation**; **Formal Analysis**; **Investigation**; **Methodology**; **Project administration**; **Supervision**; **Writing - original draft**; **Writing-review & editing-** Claudiane Mouafo, Ravi Akula, Rabie Shahzad, and Mina Ibulubo.

Acknowledgment

Not applicable.

Ethical Approval

This study was approved by the Research Ethics Committee of University of Abomey-Calavi, Cotonou, Benin, Africa.

Informed Consent

It was applicable.

Funding

Not applicable.

Data Sharing Statement

To safeguard the privacy of participant, the data is not made publicly available. Subject to conformity with institutional ethics procedures and data-sharing agreements, the corresponding author can make available, upon reasonable request, anonymized data supporting the conclusions of this work.

Conflict of Interest

The authors declare no conflict of interest.

Similarity Check

It was applied by Ithenticate®.

Application of Artificial Intelligence (AI)

Not applicable.

Peer Review Process

It was performed.

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