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Case Report

A rare case of multimetastatic cardiac angiosarcoma [☆]

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ABSTRACT

Cardiac malignant tumors are rare entities with nonspecific clinical presentation and poor prognosis. Here, we report a case of about a 30-year-old man who was admitted for right thoracic pain. Imaging indicated a cardiac malignant tumor, and pathology confirmed the diagnosis. Our case highlights the value of multimodal imaging in the differential diagnosis of a cardiac mass.

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Introduction

According to autopsy series, the incidence of cardiac tumors is 0.02%, of which 75% are benign and 25% are malignant [1]. The most frequent malignant cardiac tumors are metastases (from lung, breast, kidney, and skin cancers), angiosarcomas, and poorly differentiated sarcomas [2]. An international multicentric study reported only 61 cases of cardiac sarcomas in 6 institutions across 3 different continents over a 10-year period [3], highlighting the rarity of the disease. The clinical presentation of cardiac sarcomas is unspecific, depending

on the localization of the tumor and its effects on adjacent structures (cardiac tamponade, embolization, and valvular involvement); it can also cause arrhythmias [4]. Cardiac sarcomas are diagnosed on the basis of echocardiography, computed tomography (CT), and magnetic resonance imaging (MRI) findings. Positron emission tomography-CT (PET-CT) is also useful in the initial extension evaluation and follow-up [4]. Furthermore, the treatment for cardiac sarcomas comprise surgical resection possibly associated with chemotherapy [2]. However, the prognosis remains very poor, with a median survival between 5 and 10 months [3]. Here, we report about a rare case of a multimetastatic cardiac angiosarcoma.

Abbreviations: CT, Computed Tomography; FDG-PET/CT, FluoroDeoxyGlucose - Positron Emission Tomography/Computed; MRI, Magnetic Resonance Imaging.

[☆] Competing interest: The authors have nothing to disclose.

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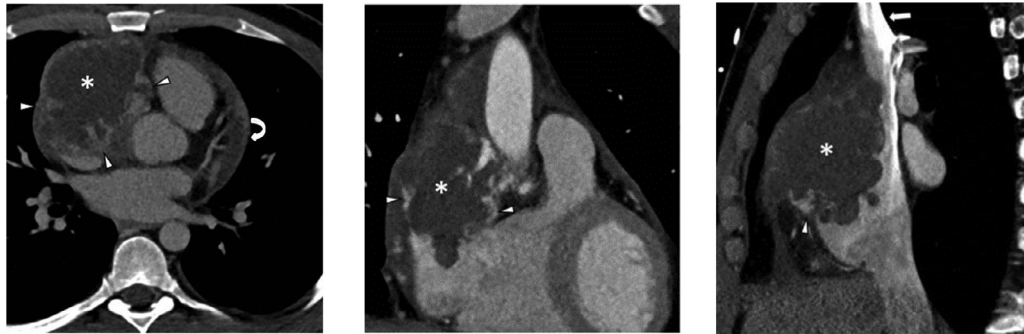


Fig. 1 – Axial, coronal, and sagittal views of an enhanced computed tomography showing a multilobular process centered on the right atrium (asterisk) and expanding to the superior cava vein (arrow). The lesion presents a nodular peripheral enhancement (arrowhead). Pericardial effusion is also visible (curved arrow).

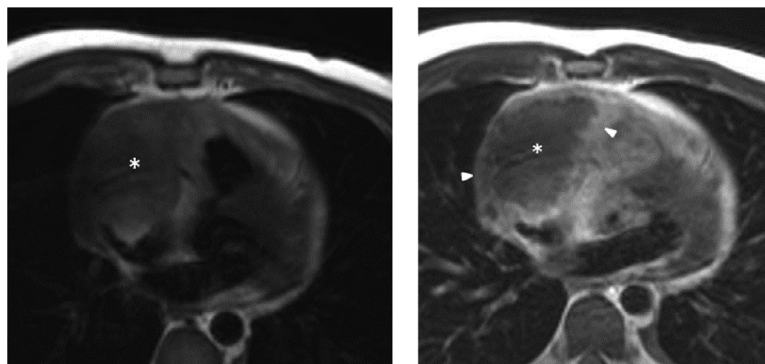


Fig. 2 – Axial T1 magnetic resonance images before (left) and after (right) intravenous injection of gadolinium showing nodular peripheral enhancement (arrowhead) of the tumor centered in the right atrium (asterisk).

Case report

A 30-year-old man presented with dyspnea and right thoracic pain. He had no medical history except for a motorbike accident 4 months earlier that caused a left clavicle fracture. Plain thorax radiography findings were normal.

A CT pulmonary angiogram excluded pulmonary embolism; however, a large mass was visible in the right atrium. A cardiac CT scan (Fig. 1) showed a multilobular process centered on the right atrium and expanding to the superior cava vein. The lesion presented a nodular peripheral enhancement and had an estimated diameter of 70 mm. Pericardial effusion was present, and many enlarged lymph nodes were noted in the mediastinum. Cardiac MRI findings were also the same (Fig. 2). A PET-CT scan (Fig. 3) revealed a lesion with a hypometabolic center and peripheral hypermetabolism.

A critical preoperative bleeding limited the surgical exploration during biopsy, resulting in an insufficient amount of tissue for a complete pathological analysis. The follow-up revealed a progressive increase in the size of the mass, and the patient later developed pulmonary, cerebral, and maxillary metastases. Histopathological examination of the biopsy of the maxillary lesion, which was easily accessible, revealed a human herpes virus-8-negative kaposiform angiosarcoma (Fig. 4). Chemotherapy, including doxorubicin (Adriamycin,

Pfizer, New York, NY) and olaratumab (Lartruvo, Eli Lilly and Company, Indianapolis, IN), was initiated. However, the patient died 7 months after the mass was detected.

Discussion

Primary cardiac sarcomas are very rare, with an incidence of 0.001%-0.03% [5]. The histopathological subtypes of cardiac sarcomas are angiosarcomas, rhabdomyosarcomas, fibrosarcomas, leiomyosarcomas, liposarcomas, and synovial sarcomas [3,6]. The prognosis remains poor, and survival does not exceed 2 years [3,7]. However, younger age on diagnosis, localized disease, and surgical management of the primary tumor are associated with improved outcomes [3]. Imaging plays a key role in the differential diagnosis between benign and malignant lesions, which has important implications for treatments and follow-ups. On both CT and MRI, the following features indicate a malignant process [8,9]: tumor size larger than 5 cm; irregular mass with ill-defined borders; direct tumor invasion of adjacent tissues; localization of the mass in the right side of the heart (typically in the right atrium for angiosarcomas); pericardial and pleural involvement, including thickening, effusions, and nodular masses; presence of multiple lesions; tissue heterogeneity indicating necrosis and hemor-

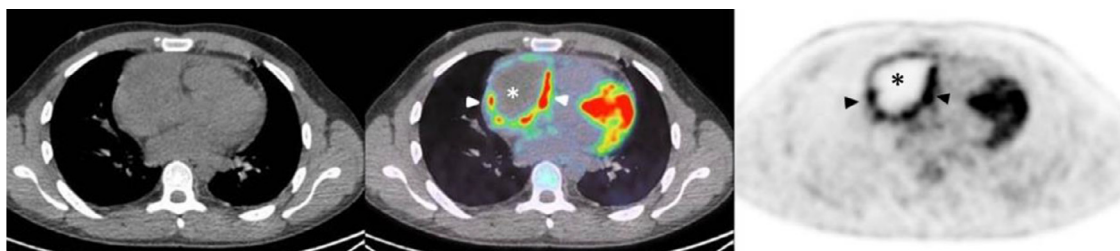


Fig. 3 – Axial 18F-fluorodeoxyglucose positron emission tomography-computed tomography showing the right atrial tumor with hypometabolic center (asterisk) and peripheral hypermetabolism (arrowhead).

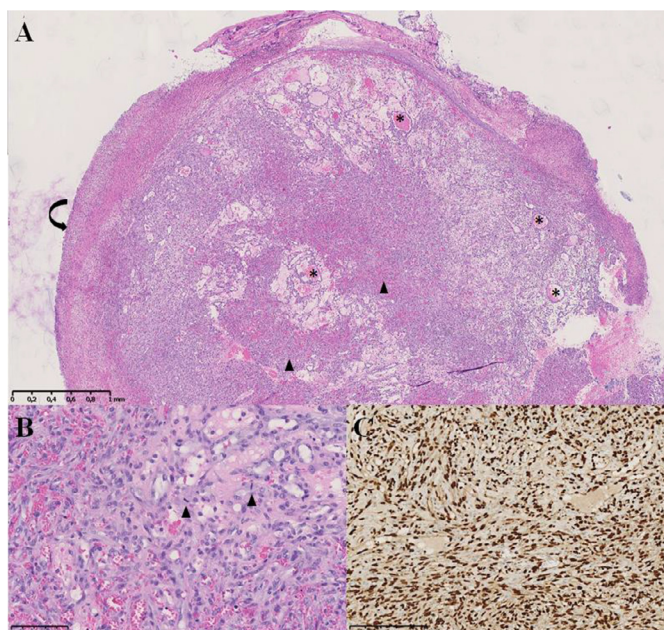


Fig. 4 – Microscopic examination of the maxillary metastasis (A: hematoxylin-eosin [HE] staining- $\times 2.5$, B: HE- $\times 20$, C: Erythroblast transformation specific related gene [ERG] immunohistochemical staining- $\times 10$). The lesion is surrounded by a Malpighian epithelium (A, curved arrow) and has many capillaries (A, asterisk) associated with high cellularity areas (A, arrowhead). These areas are composed of fusiform cells (B), and many mitotic figures are observed (B, arrowhead). ERG immunohistochemistry (C) showing diffuse nuclear staining consistent with a vascular malignant tumor.

rhage in the mass; and contrast enhancement depicting high vascularity.

The superior tissue characterization achieved with MRI makes it the best technique for predicting the malignancy of a cardiac mass [10]. 18F-Fluorodeoxyglucose-PET/CT assists the evaluation of cancer extension at initial diagnosis and follow-up and differentiates benign from malignant tumors with a sensitivity of over 90%; however, its availability remains limited [11].

In conclusion, cardiac angiosarcomas are rare malignant tumors with poor prognosis. Imaging, specifically CT and MRI, plays a key role in the differential diagnosis between benign and malignant mass, and the association of CT with PET can enhance sensitivity and specificity.

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