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Case Report

A rare case of left atrial myxofibromyosarcoma ☆,☆☆

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ABSTRACT

Myxofibrosarcoma is an aggressive soft tissue sarcoma, previously known as myxoid variant of malignant fibrous histiocytoma. Primary cardiac myxofibrosarcomas are the rarest forms of cardiac malignant tumors that often remain asymptomatic until metastasis occurs. In this case report, we describe a rare left atrial cardiac myxofibrosarcoma in a patient with recurrent renal cell carcinoma. We discuss the multimodality imaging approach to diagnose and evaluate cardiac masses as well as imaging characteristics to differentiate cardiac masses from thrombus.

Introduction

Primary cardiac tumors are exceedingly rare, with a reported incidence rate of 1.38 per 100,000 individuals [1], while metastatic tumors are reported to be 20-40 times more common, with a variable published incidence ranging between 1.5% and 20% [2,3]. Malignant primary cardiac tumors account for 25% of all cardiac tumors, of which angiosarcomas, rhabdomyosarcomas, mesothelioma, fibromyxosarcoma, and lymphomas are the most common histological subtypes [4]. The most common primary neoplasms known to have predilection for cardiac metastasis are melanoma, lymphoma, leukemia, and cancers of lung, breast, and esophagus [5].

Myxofibrosarcoma is an aggressive soft tissue sarcoma, previously known as myxoid variant of malignant fibrous histiocytoma [6]. They are commonly located in subcutaneous tissue in elderly; however, less than 3% of myxofibrosarcomas are reported to occur in the retroperitoneum [7]. Primary cardiac myxofibrosarcomas are the rarest forms of malignant cardiac tumors that often remain asymptomatic until metastasis occurs [8]. A recent pooled analysis from 30 case reports revealed they were more common in younger women and more likely to involve the left atrium [9]. Here, we report a rare left atrium myxofibrosarcoma with osseous metastasis in a 65-year-old male with recurrent left renal carcinoma.

Case report

A 65-year-old male with a past medical history of urinary tract infections, benign prostatic hyperplasia, and left clear cell renal carcinoma, status post-partial nephrectomy in March 2018, presented with acute onset of productive cough and dyspnea on exertion in June 2021. He denied chest pain, hemop-

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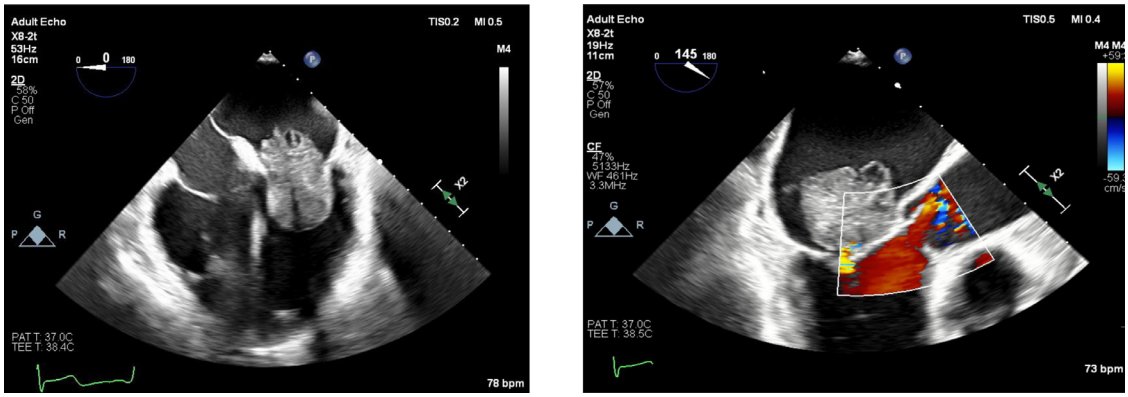

Fig. 1 – Transthoracic echocardiography apical 4-chamber view showing a large heterogeneous mobile mass obstructing mitral opening during diastole.

tysis, dizziness, headache, or leg swelling. There was no pertinent history of cardiovascular, pulmonary, or immunocompromised disease. He was being followed by urology for a left peri-hilar renal mass with a high suspicion of recurrence. At presentation, he was afebrile, HR 90, BP 126/77, RR 18, SpO2 98%. Routine blood work was notable for WBC 10.6, Hg 12.5, and platelets 58. Transthoracic echocardiography (Fig. 1) showed a mildly dilated left ventricle with a large heterogeneous mass attached to the interatrial septum, mobile and obstructing mitral opening during diastole, with evidence of mild mitral stenosis. CT angiography chest revealed mobile left atrial mass measuring 5.0 × 2.8 × 2.8 cm, attached to the interatrial septum of the left atrium extending through the mitral valve into the left ventricle, without evidence of pulmonary embolism, and findings consistent with heart failure. The double and triple inversion recovery sequences indicated no fat. The mass did not appear to be invading the myocardium. Pre contrast MR images, lobulated hypointense left atrial mass was seen (Fig. 2). Post-contrast MR images demonstrated lobulated enhancing left atrial mass (Fig. 3). Cine cardiac CT views showed lobulated left atrial mass prolapsing across the mitral valve into the left atrium (Fig. 4). On post-gadolinium images, there were nodular and linear areas of enhancement within the mass and punctate enhancing internal foci representing neovascularity (Fig. 5). No other intracardiac lesions were identified. Subsequently, he underwent excision of the left atrial mass and closure of patent foramen ovale in June 2021. Histopathological study characterized the mass as intermediate grade atrial myxofibrosarcoma with positive smooth muscle actin and desmin immunohistochemical stains and positive MDM2 12q15 gene amplification.

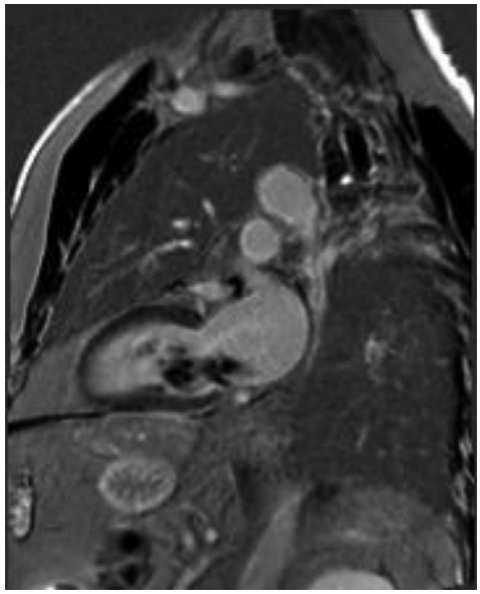

Fig. 2 – MR pre-contrast 2-chamber view demonstrates lobulated hypointense left atrial mass.

Discussion

In this study, we reviewed a case of left atrial myxofibrosarcoma, presenting in a patient with recurrent renal mass 3 years post-partial nephrectomy secondary to renal clear cell carcinoma. Isolated left atrium involvement without renal vein, inferior vena cava, or pulmonary metastasis is exceedingly rare. However, patent foramen ovale could be a potential route of seeding into the left atrium. Nevertheless, both primary and secondary myxofibrosarcomas are rare malignant cardiac tumors with poor prognoses. Additionally, without a histopathological exam of the recurrent renal mass, it is difficult to ascertain if it is indeed renal myxofibrosarcoma.

Cardiac masses are often asymptomatic and may be an incidental finding on imaging. The most frequent cardiac masses are non-neoplastic (clots, vegetations, calcifications, structural lesions), followed by benign and malignant cardiac tumors [10]. Of the benign cardiac tumors, 80% are myxomas; others include papillary fibroelastoma, rhabdomyoma, fibroma, hemangioma, lipoma, and paraganglioma [11]. Sarcomas and lymphomas are among the most common malignant cardiac tumors [12].

Multimodality cardiac imaging plays a complementary role in the evaluation and diagnosis of cardiac masses. Transthoracic or transesophageal echocardiography (TTE and TEE) is

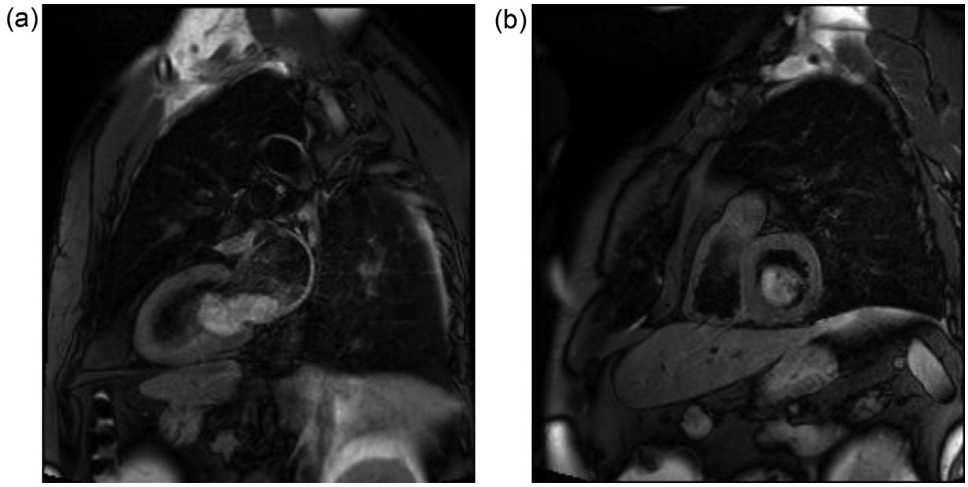

Fig. 3 – (a) MR post-contrast 2-chamber view (b) short axis view demonstrates lobulated enhancing left atrial mass.

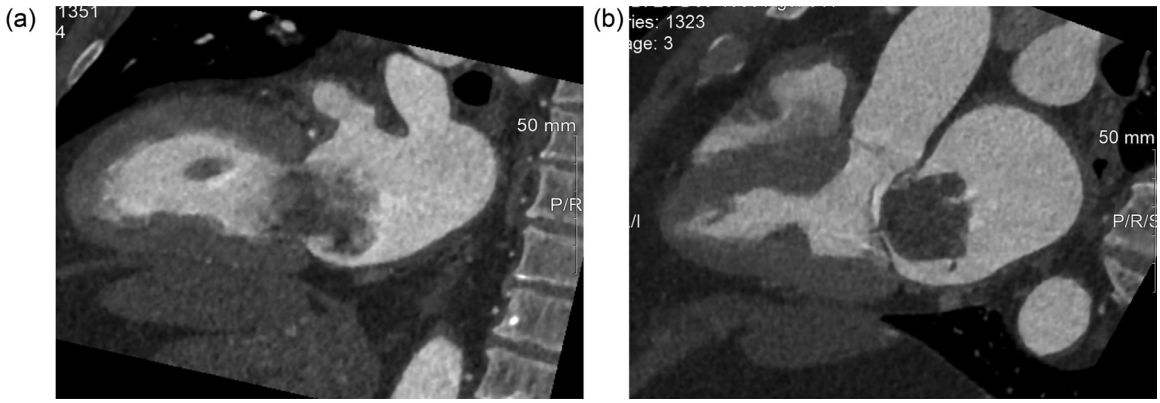

Fig. 4 – Cine (a) short axis and (b) long axis views of cardiac CT demonstrates lobulated left atrial mass prolapsing across the mitral valve into the left atrium.

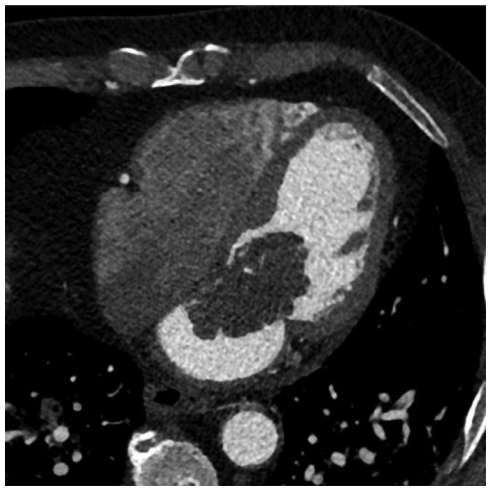

Fig. 5 – CTA cardiac image demonstrating left atrial mass broad based towards posterior wall. Punctate internal enhancing foci representing neovascularity.

often the first-line imaging modality that provides crucial information regarding mass location, shape, attachment, size, mobility, and hemodynamic impact. However, echocardiography is limited by poor acoustic window, and its ability to characterize soft tissue masses and extracardiac involvement [13]. Computed tomography (CT) provides volumetric data and is optimal for the evaluation of extracardiac involvement as well as calcified masses. Contrast-enhanced CT studies provide information on tumor neovascularization. Additionally, in those with contraindications to MR, CT is the modality of choice. Limitations of CT include radiation exposure, a small risk of contrast-induced nephropathy, and poor temporal resolutions compared to MR [14]. Multiparametric cardiac MR (CMR) provides multiplanar non-invasive assessment of cardiac mass with superior tissue characterization, unrestricted field of view, and higher temporal resolution than CT. However, CMR has limited availability, and it may not be compatible with a few implantable devices [15].

Specific imaging characteristics of CMR to differentiate a thrombus from cardiac tumor include:

(1) T1 hypo/iso intensity with T2 hyperintensity is a characteristic feature of the majority of cardiac tumors. Whereas acute clot has isointense to high signal intensity on T1and T2, subacute has high T1 and low T2 intensity, and chronic clot has low T1 and T2 signal intensity.
(2) First pass contrast enhancement is indicative of neovascularization.
(3) Malignant tumors have early post-contrast enhancement that is not seen with thrombus.
(4) T1 nulling time on gadolinium enhancement is shorter than myocardium for malignant tumors but longer for thrombus.
(5) Cine gradient-echo (GRE) and cine steady-state free precession (SSFP) is hyperintense in tumors, whereas thrombus has hypointense signal.

Lastly, ^{18}F FDG-PET studies help to differentiate benign from malignant tumors and predicts survival. Malignancy can be excluded with confidence in studies with no ^{18}F-FDG uptake in a lesion. Infection, surgical, or radiation changes may at times lead to false positive ^{18}F-FDG uptake [16]. Hybrid modalities such as PET/MRI, PET/CT add incremental value and enhance the diagnostic accuracy [17].

Conclusion

Cardiac myxofibrosarcomas are rare malignant tumors. Although primary myxofibrosarcomas have been documented in a few published case reports, metastatic cardiac myxofibrosarcoma without inferior vena cava involvement is unusual. Even though echocardiography is the first imaging modality of choice in cardiac masses, cardiac MR provides useful information to delineate thrombus from malignant tumor. Given the aggressive nature of this tumor, surgery along with chemotherapy and radiation is inevitable in the management of myxofibrosarcomas.

Patient consent

We complied with our institution policies of IRB approval for case reports or image submissions. Written informed consent was obtained from the authorized legal representative of the patient as the patient is now deceased. The informed consent is now part of patient records. All the images included are non-identifiable images consistent with Elsevier policies.

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