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Donor transmitted left atrial myxoma 13 years after heart transplantation

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Summary

Background: Left atrial cardiac myxomas are among the most common cardiac masses. However, occurrence of left atrial myxomas in post-transplant patients is very rare and often misdiagnosed as left atrial thrombus formation.

Case Report: We report the case of a 67-year old female, who was referred due to suspected left atrial thrombus but was found to have a pediculated mass at the suture line of the left atrium on cardiac MRI. After resection, the diagnosis myxoma was confirmed histologically and the donor origin of the myxoma was proven by tissue typing.

Conclusions: Despite a rare entity, atrial myxomas may occur in post cardiac transplant patients and may therefore support the role of advanced imaging techniques in patients with suspected left atrial masses.

Key words: heart transplantation • cardiac myxoma • heart transplantation – adverse effects

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Background

Left atrial cardiac myxomas are among the most common cardiac masses. However, occurrence of left atrial myxomas in post-transplant patients is very rare and often misdiagnosed as left atrial thrombus formation.

Case Report

A 67-year-old female patient, who had undergone orthotopic heart transplantation due to dilative cardiomyopathy 13 years ago, was admitted due to a suspected intraatrial thrombus diagnosed in an external echocardiography study. The heart was donated by a 47-year-old female who deceased due to subarachnoidal bleeding. After transplantation, the patient had an uncomplicated recovery and immunosupression was maintained after steroid withdrawal with tacrolimus, mycophenolate mofetil.

There was no history of cardiac tumors neither in the subject nor the donor family. Due to the suspected atrial thrombus anticoagulation with phenprocoumone was initiated six months ago. On admission, the patient was asymptomatic with no cardiac murmurs. She revealed a slightly elevated blood pressure of 140/90 and renal impairment with a serum creatinine of 172 µmol/l.

A contrast-enhanced CTA was omitted due to her underlying renal failure and the patient was referred for cardiac mangetic resonance imaging (MRI, 1.5T Avanto, Siemens Medical Solutions, Forchheim, Germany). Single shot fast spin echo images ("Black blood") through the left atrium revealed a hyperintense ovally configured pediculated cardiac mass of 15 mm diameter attached to the basal segment of the left atrium (Figure 1A, B). In cine sequences, a characteristic movement at the border of the right inferior pulmonary vein was detected. After contrast administration of gadolinium, there was a moderate contrast enhancement consistent with a cardiac myxoma (Figure 2).

Due to the suspected cardiac myxoma, the patient was scheduled for elective surgery. Aortobicaval cardiopulmonary bypass was established and the right atrium was incised. To assure access to the mass the interatrial septum was incised (Figure 3). Intraoperatively, the pedicle was attached to the previous suture line and extended to the base of the left atrium. Given the extent of the typical morphologic appearance, the atrial wall was excised transmurally with a 5 mm margin and subsequently closed with direct sutures. The postoperative course was uneventful.

Macroscopically, the mass was smooth with a size of 1.5×1 cm with a gelatinous appearance and an irregular surface (Figure 4). On histology mycoid stroma consistent with a polypoid atrial myxoma was found. The resection margins were free from tumor. HLA typing obtained of the fresh tumor proved its donor origin since the mass was HLA-identical to the donor's tissue typing obtained at the time of transplant and had no matches with the recipient's HLA-genotype (A2, A19, A31, B5, B51, B15, B62, DR4, DR53).

Discussion

We herein report the rare case of a patient with a cardiac mass who underwent multi-modality imaging and surgical resection for the diagnosis of a post-transplant left atrial myxoma. Cardiac myxomas are the most prevalent cardiac tumors [1]. The majority are located in the left atrium and most of the myxomas originate from the interatrial septum. Cardiac myxomas usually present as benign tumor, however serious complications include embolisation and mitral valve obstruction and even with complete excision there is a recurrence rate of 3% [2]. To our knowledge, there are very rare

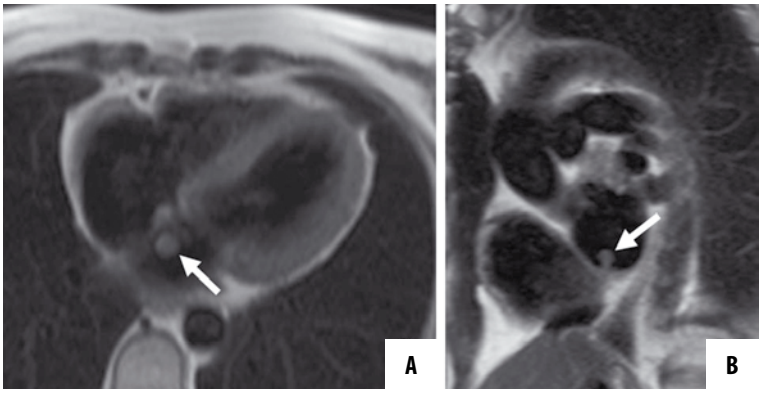

Figure 1. Axial (**A**) and oblique (**B**) Single shot fast spin echo images through the left atrium demonstrating a hyperintense ovally configured pediculated cardiac mass of 15 mm diameter attached to the basal segment of the left atrium (arrow).

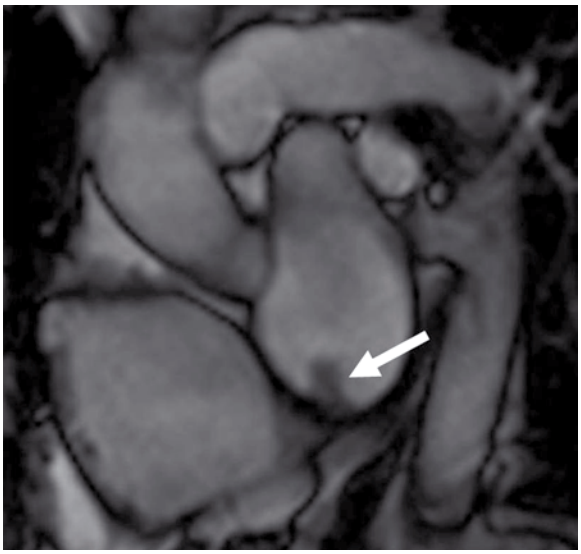

Figure 2. T1w images revealing a moderate contrast enhancement of the pediculated mass in the left atrium (arrow).

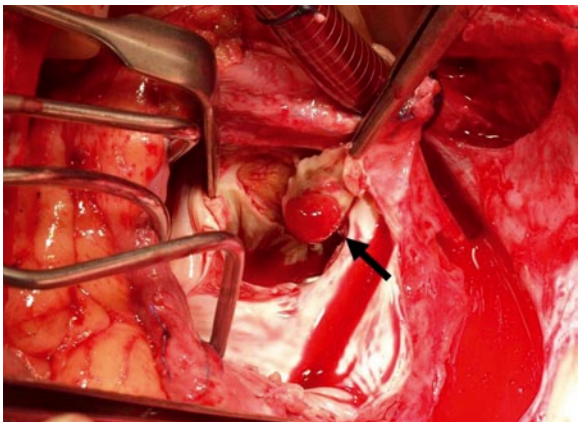

Figure 3. Surgical view after incision of the right and left atrium. Excision of the myxoma (arrow) adjacent to the suture line remaining after HTx.

prior cases that report on the occurrence of cardiac myxomas in transplanted hearts that originate from donor [3] or recipient tissue [4,5]. We provide advanced technology MR images, demonstrating the typical enhancement and hyperintense appearance on T2w images. Usually, Doppler sonography reveals vascularisation and typical perfusion patterns. We confirm earlier reports that myxomas usually arise adjacent to the suture line between the donor and recipient left atrium.

Differentiation between thrombus and masses in the atria remains challenging. The majority of cardiac myxomas can be adequately visualized by transthoracic echocardiogram but a transesophageal echocardiogram is often necessary for accurate delineation of its structure and site of attachment. In contrast, a left atrial thrombus is usually of irregular appearance, has a broad base, layered

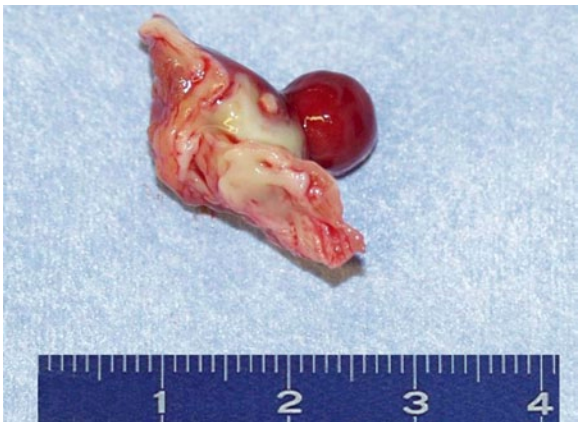

Figure 4. Transatrial resected left atrial myxoma with atrial myocardium.

echocardiographic features and is usually associated with valvular heart disease, LA enlargement, atrial fibrillation and originate from the LA appendage or the posterior atrial wall. Doppler sonography may help differentiating a tumor from a thrombus by demonstrating tissue perfusion but myxomas are not always vascular and the absence of colour flow or contrast uptake does not exclude a tumor. In our case, the patient was put on warfarin due to suspected thrombus formation and only advance imaging revealed the entity of the left atrial mass. Our case emphasizes the need of advanced imaging techniques, such as high resolution doppler sonography or cardiac MRI for accurate diagnosis in unclear cases.

Long term immunosupression is a known risk factor for the growth of secondary tumors, such as lymphoproliferative disorder or squamous cell carcinoma but it remains uncertain weather there is a link between immunosupperessive therapy and the growth of cardiac myxomas. Cyclosporine has been suspected as a causative agent due to an inhibiting effect on cytokine production, i.e. interleukin-6 and -8, which are known to have an association with the development of neoplasms [5] and has been observed previously in a case of cardiac myxoma [6]. Whether the tacrolimus medication in our case contributed to the growth is uncertain since there are alternative hypotheses of a generally suppressed immune system contributing to tumor growth which cannot be attributable to a single agent [7].

Conclusions

Despite a rare entity, atrial myxomas may occur in post cardiac transplant patients and may therefore support the role of advanced imaging techniques in patients with suspected left atrial masses.

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