Tumor involvement of the left atrium and mitral valve: imaging features of intra-cardiac malignancy


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Learning Point for Clinicians

We presented pre-treatment imaging of rare primary intimal sarcoma involving the left atrium and mitral valve. Despite the rarity of primary cardiac intimal sarcoma, imaging features of wide-based involvement of the tumor with intra-tumoral edema, post-gadolinium enhancement and a secondary tumor nodule with similar imaging findings involving the mitral valve were suggestive of intra-cardiac malignancy. Preoperative imaging evaluation is particularly beneficial to determine the tumor extent and can help to make optimal therapeutic decision and surgical planning.

Introduction

Although the majority of these tumors arise from the aorta or central pulmonary arteries, cardiac intimal sarcoma is a rare primary cardiac neoplasm with only three reported cases. We presented pre-treatment imaging of rare primary intimal sarcoma involving the left atrium and mitral valve. Despite the rarity of primary cardiac intimal sarcoma, imaging features of wide-based involvement of the tumor with intra-tumoral edema, post-gadolinium enhancement and a secondary tumor nodule with similar imaging findings involving the mitral valve were suggestive of intra-cardiac malignancy. Preoperative imaging evaluation is particularly beneficial to determine the tumor extent and can help to make optimal therapeutic decision and surgical planning.

Case presentation

A 57-year-old female presented with persistent chest pain and exertional dyspnea for one and a half month. A two-dimensional transthoracic echocardiography (TTE) revealed a large isoechoic mass in the left atrium attached to its posterior wall with another subcentimeter nodular lesion attached to the
posterior leaflet of mitral valve (Figure 1A) resulting in mitral valve regurgitation. Cardiac magnetic resonance imaging (CMR) demonstrated a well-circumscribed, ovoid mass within the left atrium with broad base of attachment to the posterior wall of left atrium (Figure 1B). The mass was intermediate signal intensity on T1-weighted image (T1WI), hyperintense on T2 fat saturation image (Figure 1B) with intact fat plane between the mass and interatrial septum and epicardium on T1WI. Contrast-enhanced T1WI done after perfusion imaging revealed persistent tumor enhancement. In addition, CMR confirmed the small nodular lesion attached to the posterior leaflet of the mitral valve with similar signal intensity to the left atrial mass (Figure 1B) suggesting similar histopathology and most likely malignant neoplastic etiology.

Based on the multimodality imaging findings of primary malignant cardiac tumor, the patient underwent surgical resection. Intraoperatively, a large left atrial mass with intramural invasion of the posterior wall of the left atrium was removed completely. However, the small posterior leaflet of mitral valve tumor seen grossly invading into the posterior wall of left ventricle was technically difficult to resect completely with subsequent reconstruction of the mitral valve thus rendering it unresectable. Postoperatively, the patient was placed on combination chemotherapy. Subsequent histopathological examination of the tumor confirmed intimal sarcoma of the left atrium.

**Discussion**

Primary malignant cardiac tumors usually have a dismal prognosis; without surgical resection, the reported survival rate at 9–12 months is only 10%. When possible, surgical excision is the best method of therapy for these cardiac tumors, and curative resection can significantly improve the prognosis.

TTE is the most commonly used initial imaging technique, which however is limited by the small field of view and poor tissue characterization. On the contrary, CMR can describe the tissue characteristics, such as presence of edema in T2-weighted imaging, preservation of fat plane in T1-weighted imaging and post-gadolinium enhancement. Malignant cardiac sarcomas, such as undifferentiated sarcoma or rhabdosarcoma, can be single or multiple, which has a tendency to involve cardiac valve.

**Conclusion**

Despite the rarity of primary cardiac intimal sarcoma, imaging features of wide-based involvement of the tumor with intra-tumoral edema, post-gadolinium enhancement and a secondary tumor nodule with similar imaging findings involving the mitral valve were suggestive of intra-cardiac malignancy. Preoperative imaging evaluation also particularly beneficial to determine the tumor extent, knowledge of which is the key for optimal therapeutic decision making and surgical planning.

![Figure 1. Imaging of the intimal sarcoma involving the left atrium and mitral valve. (A) Apical four-chamber TTE view reveals a 2.9 cm x 3.5 cm tumor with broad attachment to the posterior wall of the left atrium (bold arrow). Another small tumor is seen fused with the posterior leaflet of mitral valve (thin arrow). (B) T2-weighted double inversion recovery image with fat saturation in four-chamber view shows high signal intensity as compared with myocardium in both the tumors (bold and thin arrows).](https://academic.oup.com/qjmed/article-abstract/107/9/747/1569327)
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