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Editorial

Image modalities to assess cardiac tumors: Echocardiography, multidetector CT, and MR imaging

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Primary cardiac tumors of the heart are rare. The prevalence of such neoplasms has been reported to range from 0.001% to 0.03% in post-mortem series [1]. On the other hand, metastatic involvement of the heart is approximately 30 times more prevalent than primary cardiac tumors. Primary cardiac neoplasms include both benign and malignant histological types. Approximately 75% of primary cardiac tumors are benign and nearly half of these are myxomas. The majority of primary malignant tumors of the heart are sarcomas, most often vascular in origin [2]. Angiosarcoma is the most common, whereas osteosarcoma and myxofibrosarcoma are the least common of the various primary cardiac sarcomas. Thus, the histological types of cardiac tumors are varied, accordingly characterization of cardiac tumors using imaging techniques has been challenging.

Recently, image resolution and tissue characterization of various imaging modalities have been developed, therefore, familiarity with certain imaging features could help to diagnose cardiac tumors. Up to now, transthoracic echocardiography has been the most widely used imaging modality for the evaluation of cardiac masses. Echocardiography is the best imaging modality to depict small masses that arise from the cardiac valves because of high resolution. In addition, it allows assessment of hemodynamic alteration by cardiac tumors, such as valve obstruction and tamponade. 3D echocardiography can provide the opportunity of true spatial assessment of entire tumors (rather than individual slices of tumors) and help to understand the relationship between tumors and adjacent cardiac structures. However, although the use of transesophageal echocardiography overcomes the limited acoustic window of the transthoracic mode, visualization of extracardiac extension is suboptimal since the airways and lungs can be obstacles for imaging of the aortic arch, pulmonary arteries, and some systemic and pulmonary veins. On the other hand, multidetector computed tomography (MDCT) can provide anatomical information without blind spots and be useful in staging of malignant tumors. The use of electrocardiogram-gated MDCT has better soft tissue contrast than echocardiography and can definitely characterize fatty content and calcifications. A wide field-of-view with MDCT helps to assess the extent of a cardiac malignancy and helps to detect metastatic lesions. However, MDCT also has disadvantages including radiation exposure and need for nephrotoxic contrast material. Streak artifacts can occur when the contrast material remains in the right heart during image acquisition. Magnetic resonance imaging (MRI) is useful for tissue characterization of cardiac masses. High contrast resolution and multiplanar capability allow a specific diagnosis and optimal evaluation of myocardial infiltration, pericardial involvement, and extracardiac extension. The use of MRI allows characterization of a mass due to fibromas and hemangiomas. Homogeneity of a mass due to compact cellularity may be characteristic of a lymphoma [3]. Acquisition of post-contrast sequences enables better depiction of tumor vascularity and can be used to define tumor borders. In addition, differentiation between cardiac tumors and thrombi by MRI can be helpful to avoid unnecessary surgical procedures. Disadvantages of the use of MRI include a long scan time and the inability to demonstrate calcification and small masses, especially in cardiac valves due to limited spatial resolution. MRI is not available to the patients with non-MRI compatible pacemaker and implantable cardioverter-defibrillator devices, and those with non-sinus rhythm and claustrophobia. Thus, each imaging modality has different strengths and limitations. Combination of echocardiography, MDCT, and MRI could aid differential diagnosis, and allow determination of the resectability of tumors and planning for the reconstruction of cardiac chambers in pre-surgical assessment.

Myxofibrosarcoma is a tumor of mesenchymal origin that occurs most commonly in the extremities of the elderly [2,4]. Tumors typically arise in the lower and upper extremities, trunk, retroperitoneum, as well as the head and neck region. More than half of these are located in dermal or subcutaneous tissues. Myxofibrosarcoma infrequently occurs in other tissues but has been reported to arise in the tunica albuginea of the testis, cervix, vulva, breast, esophagus, maxillary, and sphenoid sinuses, hypopharynx, intra-articular region of the hip, tibia, mandible, external auditory canal, and possibly pancreas. Low-grade myxofibrosarcoma tends to metastasize locally, requiring wide surgical resection. Repeated local recurrences with progressively poor differentiation may occur in such cases. High-grade myxofibrosarcoma also invade local and contiguous tissues but are also associated with distant metastasis (lymph nodes, lungs, brain, and bone). The cell origin of myxofibrosarcoma is the myofibroblast. Some experts believe that this tumor is the myxoid variant of malignant fibrous histiocytoma [2,4].

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Primary cardiac myxofibrosarcoma rarely arises in the heart. To our knowledge, the case reported by Iglesias et al. [5] is the 15th case since 1963 in the PubMed database [6–15]. Patients' age ranged from 6 to 66 years (mean age 39.6 years), which is younger than those with myxofibrosarcoma in extracardiac organs (60.5 years), and there were 6 males and 9 females. The most common symptom was dyspnea, which was similar to myxomas. Tumors involved predominantly the left atrial cavities (10 cases), mitral valve in 2 cases, right atrium in 1 case, and left ventricle in 1 case (information on 1 case was not available). Tumors may be unicentric or multicentric. Low-grade tumors invade local tissue including valvular structures. High-grade tumors also invade locally, but may also metastasize to contiguous and distant sites. Left atrial myxofibrosarcoma may simulate left atrial myxoma in its clinical presentation. The outcomes of the patients were unfavorable. Of all patients, only 2 achieved complete recovery: a 66-year-old woman after tumor resection and radiation therapy [10] and a 6-year-old girl after heart transplantation [14]. The remaining 13 patients died or had a recurrence 3–50 months after diagnosis despite surgical resection, radiation therapy, and chemotherapy. The case reported by Iglesias et al. is surviving 46 months after diagnosis with only few symptoms fortunately [5].

At the current moment, cardiac myxofibrosarcoma is difficult to differentiate from myxomas. Heterogeneity is a common feature of myxomas and myxofibrosarcomas, and is characterized by hemorrhage, necrosis, cyst formation, fibrosis, or calcification. Both typically have lobular contours. However, the prognoses of the two tumor types are quite different. Further improvement in imaging techniques is required before myxofibrosarcoma can be diagnosed before surgery.

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