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Editorial

A Comprehensive Perspective of Clinical and Echocardiographic Features in the Differential Diagnosis of Cardiac Myxomas and Myxoma-Like Masses

Dong-Soo Kim, MD, PhD

Division of Cardiology, Department of Internal Medicine, Busan Paik Hospital, Inje University College of Medicine, Busan, Korea

▸ See the article "Comparison of Clinical and Echocardiographic Characteristics between Cardiac Myxomas and Masses Mimicking Myxoma" in volume 50 on page 822.

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Correspondence to
Dong-Soo Kim, MD, PhD
Division of Cardiology, Department of Internal Medicine, Busan Paik Hospital, Inje University College of Medicine, 75, Bokji-ro, Busanjin-gu, Busan 47392, Korea.
E-mail: dongskim@inje.ac.kr

ORCID iDs
Dong-Soo Kim
https://orcid.org/0000-0003-4463-1085

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Primary cardiac tumors (PCTs) are rare and the incidence is from 0.001 to 0.3% by autopsy.[1] Approximately 90% of PCTs are benign, and it was reported that nearly 80% of these tumors were cardiac myxomas (CMs).[2][3] Although there is no systematic study on the incidence of PCTs in Koreans, the comprehensive studies reported so far have shown that the frequency of benign tumors is higher than that of foreign countries (92%), and CMs are higher among them (89%).[4]

CMs are usually solitary and are attached to the interatrial septum (IAS) at the border of the fossa ovalis, usually protruding into the left atrium (LA) but occasionally into the right atrium (RA) and rarely into the ventricles.[3][5] The mobility of the tumor depends upon the extent of attachment to the IAS and the length of the stalk. CMs vary in size from 1–15 cm (mean 3–4 cm) in diameter.[3] They are generally polypoid with a smooth or gently lobulated surface, often pedunculated, and characteristically arise from a narrow stalk. On the other hand, the less common villous or papillary myxomas have multiple fine fragile villous extensions at the greatest risk of embolism. It is known that embolism is about 10–20%, but is not related to tumor size.[5][6] The clinical features of CMs are determined by their location, size, and mobility.[3] Although clinical manifestations vary among studies, dyspnea due to intracardiac obstruction, constitutional symptoms, and embolism are the most frequent clinical manifestations of CMs ("myxoma triad"). Occasionally, early diagnosis may be a challenge because there are no symptoms, particularly with small tumors.

Transthoracic echocardiography (TTE) is the most commonly used imaging modality for cardiac tumor detection. It can usually provide adequate diagnostic information, such as location, a site of attachment, size, shape, mobility and other morphologic characteristics of a tumor as well as any hemodynamic consequences.[1][2][5] However, suboptimal image quality due to poor echogenicity blocked by bone and lung tissue in TTE increases the probability of false positives or false negatives, so transesophageal echocardiography (TEE) is often required. TEE is excellent for imaging the RA myxoma, small cardiac lesions, extracardiac involvement and also for determination of the exact location of tumor attachment which is an important consideration in the planning of surgical excision.[2][5] Meng et al.[7] reported that the diagnostic sensitivity of TTE and TEE was 93.3% and 96.8%, respectively. Thereafter, the cardiac mass can be checked in detail by computed tomography (CT) or cardiac magnetic

resonance (CMR) imaging for further characterization and differential diagnosis. As CMR is superior to evaluate tumor location and extent as well as tissue characterization, it is increasingly used in the diagnostic work-up of CMs.[2)5)] Moreover, it is thought that the real-time 3D echocardiography will be more useful than other areas to accurately determine the overall characteristics of the mass. Recently, incidental detection of CMs is not infrequent because increasing use of imaging modalities (e.g., echocardiography, CMR, and CT) has led to an increasing number of incidental findings of PCTs.

Lee et al.[8)] reported valuable information on the comparison of the clinical and echocardiographic characteristics of CMs, myxoma-mimicking other tumors, and thrombi in 265 patients who were diagnosed with CMs by TTE. Of the 265 patients (mean age, 61±16 years; 63.8% for female), 195 patients (73.6%) underwent surgical resection and 174 (65.7%) were pathologically confirmed with CMs. Among 186 patients whose pathological diagnosis was confirmed, 174 (93.5%) were identified as CMs, 8 (4.2%) of patients were established to have other cardiac tumors, and 4 (2.2%) of the masses were proven to be atrial thrombi. Compared with previous reports of CMs diagnosed in the cardiac masses,[6)7)9)] this study was similar in terms of clinical manifestation, tumor location and characteristics on echocardiography. Lee et al.[8)] also remarked on different features in characteristics of CMs and myxoma-mimicking other tumors. Non-myxoma tumors were significantly smaller (20.4×12.6 mm vs. 41.4×27.6 mm, $p<0.01$) and less frequently found at the LA (87.5% vs. 10.5%, $p<0.001$) than CMs. All thrombi had comparable or larger size than CMs. Atrial fibrillation (AF) on electrocardiogram was more frequent (75% vs. 7.0%, $p<0.001$) and LA diameter was larger (55.0±14.6 mm vs 41.3±7.7 mm, $p=0.001$) in patients with thrombi than in those with CMs. As they reported, compared with CMs, other non-myxoma tumors were smaller, more frequently found in non-atrial sites, and thrombi were associated with AF and larger LA diameter.

This study is the largest retrospective study that has been recently published by investigating 265 patients with echocardiographic diagnosis of CMs in 4 teaching hospitals in Korea, but the number of non-myxoma cardiac mass was too small to evaluate the different characteristics from CMs with statistical significance. However, this study is meaningful to summarize the prevalence and characteristics of primary cardiac masses in Korea and compare the characteristics of pathologically proven myxoma and other masses mimicking myxoma on echocardiography.

In conclusion, if a relatively small-sized tumor is found at a site other than atria, the first presumptive diagnosis should be one of non-myxoma cardiac tumors, and in patients with AF, a thrombus should be considered primarily, even if it mimics myxoma.

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