Metastatic Ewing's sarcoma involving the right ventricle

Sağ ventrikülü tutan metastatik Ewing sarkomu

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Cardiac metastasis of Ewing's sarcoma is rare. A 22-year-old woman was admitted with complaints of palpitation and fatigue on exertion. She had a seven-year history of radical right tibial resection for Ewing's sarcoma and was also receiving chemotherapy for lung metastasis of Ewing's sarcoma. Both transthoracic and transesophageal echocardiography demonstrated a single, large (3x3.5 cm) inhomogeneous mass located in the free wall of the right ventricle. To differentiate the mass from a massive thrombus, contrast-enhanced magnetic resonance imaging was performed. The mass showed partial contrast enhancement, suggesting a malignant metastatic mass. Surgical resection was not considered due to accompanying lung metastasis and potentially poor outcome of the operation.

Key words: Diagnosis, differential; echocardiography; heart neoplasms/secondary; sarcoma, Ewing's/secondary.

Ewing sarkomunda kalp metastazı nadirdir. Yirmi iki yaşında bir kadın hasta egzersizle ortaya çıkan çarpıntı ve halsizlik yakınmalarıyla başvurdu. Hastaya yedi yıl önce Ewing sarkomu nedeniyle radikal sağ tibia rezeksiyonu uygulandığı ve aynı nedenli akciğer metastazı için halen kemoterapi gördüğü öğrenildi. Transtorasik ve transözofajiyal ekokardiyografide, sağ ventrikül serbest duvarında büyük bir inhomojen kitle (3x3.5 cm) görüldü. Kitleyi masif trombüsten ayırt etmek için yapılan kontrastlı manyetik rezonans görüntülemede kısmi kontrast tutulumu izlenmesi üzerine kitlenin malign metastatik lezyon olduğu düşünüldü. Eşlik eden akciğer metastazı ve cerrahinin fazla yararı olmayacağı göz önüne alınarak hasta için cerrahi rezeksiyon önerilmedi.

Anahtar sözcükler: Tanı, ayırıcı; ekokardiyografi; kalp neoplazileri/ikincil; sarkom, Ewing/ikincil.

In principle, every malignant tumor can metastasize to the heart. So far, only central nervous system tumors have not been reported to cause cardiac metastasis.[1] The most common tumors associated with cardiac metastasis are carcinomas of the lung, breast, esophagus, malignant lymphoma, leukemia, and malignant melanoma.[2-6] Ewing's sarcoma is a highly aggressive, round cell neoplasm of uncertain origin, mostly affecting children and adolescents. It accounts for about 5% of all bone tumors, and most are located in the long bones.[7] Cardiac metastasis of Ewing's sarcoma is rare. In a comprehensive literature search, we could find only four cases of Ewing's sarcoma with cardiac metastasis.[8-11] We present an unusual case of Ewing's sarcoma with metastasis to the right ventricle which was diagnosed by transthoracic (TTE) and transesophageal echocardiography (TEE).

CASE REPORT

A 22-year-old woman was admitted with complaints of palpitation and fatigue on exertion. She had a seven-year history of radical right tibial resection for Ewing's sarcoma. At the time of admission, she was also under chemotherapy for lung metastasis of Ewing's sarcoma, confirmed histologically by a lung biopsy. On admission, physical examination and laboratory data revealed no abnormality. In the apical four-chamber view, TTE demonstrated a single, large (3 x 3.5 cm) inhomogeneous mass with a regular margin, located in the free wall of the right ventricle (Fig. 1a). Left ventricle function was normal. The mass did not hinder tricuspid valve motion. To rule out the presence of smaller masses that might have been overlooked on TTE and to depict the mass in detail, TEE was performed with a 6.5 MHz multiplane probe (Vivid 7, GE, Norway) (Fig. 1b), which showed no coexisting masses, no involvement of any other cardiac structure, nor any evidence for metastasis to the inferior vena cava. To differentiate the mass from a massive thrombus, magnetic resonance imaging (MRI) with contrast media was

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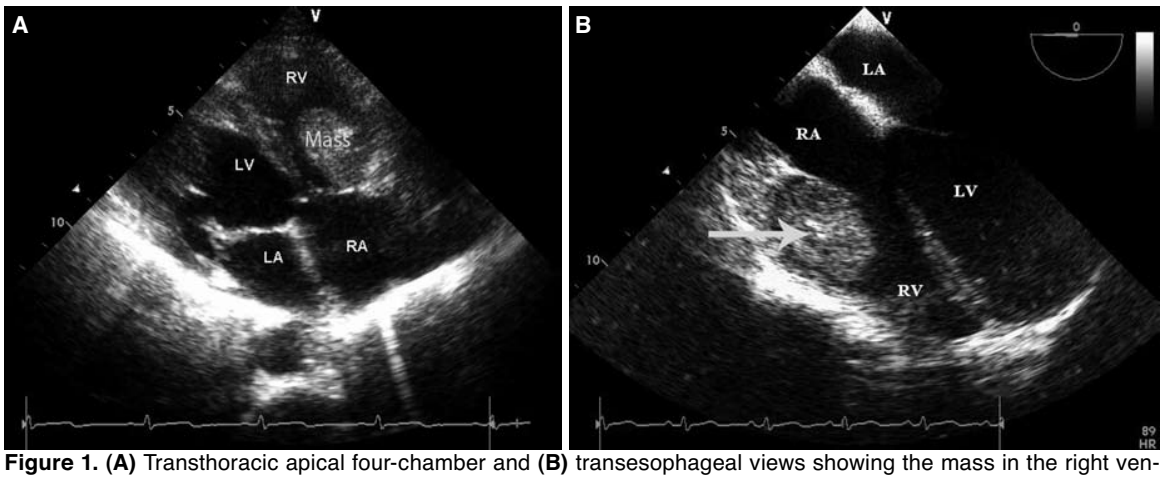

Figure 1. (A) Transthoracic apical four-chamber and **(B)** transesophageal views showing the mass in the right ventricle. LA: Left atrium; LV: Left ventricle; RA: Right atrium; RV: Right ventricle.

performed (Fig. 2). The mass showed partial contrast enhancement, suggesting a malignant metastatic mass. We did not recommend surgical resection due to accompanying lung metastasis and potentially poor outcome of the operation.

DISCUSSION

The frequency of cardiac metastases was generally underestimated before the advent of echocardiography. In different series, cardiac metastases were found in up to 25% of postmortem patients with malignancies.[1-5] Metastasis of Ewing's sarcoma to the heart is rare and its incidence is not known. It usually metastasizes to the lung, bone, pleura, lymph node, and nervous system. Metastasis to other organs, such as liver, breast, kidney, and heart is rare.[3,7,12] We only found four cases of cardiac metastasis from Ewing's sarcoma; among these, only one involved the right ventricle, and another was solitary metastasis to the heart.[8-11] In our patient, the tumor metastasized to both the lung and the right ventricle.

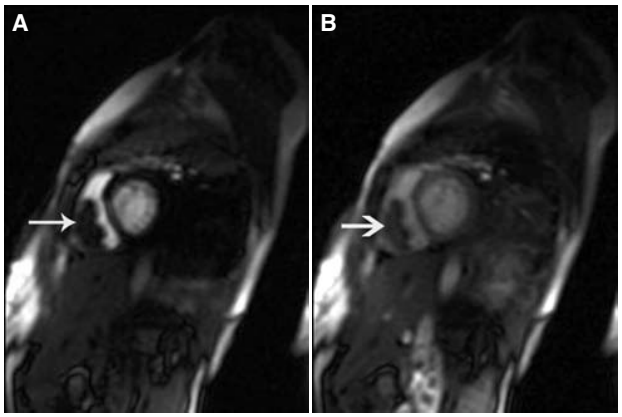

Figure 2. (A) Early and **(B)** late phase magnetic resonance scans with contrast media in the basal short-axis view. The long arrow shows the mass in the right ventricle and the short arrow indicates contrast enhancement of the mass.

Sarcomas or mesotheliomas are considered metastatic if an extracardiac tumor site is documented.[1] Despite the lack of a biopsy result, we considered the mass in the right ventricle to be metastatic Ewing's sarcoma because two extracardiac tumor sites, the right tibia and the lung, had been shown previously and the mass also showed contrast enhancement. Whether benign or malignant, the majority of primary cardiac tumors are intracardiac and usually lead to serious cardiac symptoms.[1,13,14] Intracavitary location of secondary heart tumors is unusual and, since the symptoms of disseminated tumor disease outweigh, signs of cardiac involvement are often overlooked and metastatic heart tumors rarely gain clinical attention.[1,3]

In all reported cases of Ewing's sarcoma, metastasis to the heart was diagnosed at surgery or autopsy. Two-dimensional echocardiography is the method of choice to detect cardiac metastasis and associated complications.[15,16] Cardiac metastatic lesions are usually small and multiple; however, a single large tumoral lesion may also be observed.[1-6] Masses as small as or smaller than 0.5 cm in diameter make the diagnosis rather difficult on TTE examination. On the other hand, masses large enough to distort cardiac structures may be difficult to visualize by TTE. In both situations, and in cases of peri- or paracardiac lesions, transesophageal approach is superior to transthoracic examination.[16] In our case, the metastatic tumor was single and large, and TEE only confirmed the findings obtained by TTE examination.

Computed tomography (CT) and MRI acquire images of cardiac structures in any plane without overlapping, providing additional informa-

tion.[17,18] The differential diagnosis of intracavitary mass lesions includes benign and malignant primary cardiac tumors, intracardiac metastases, thrombus, vegetation, and a foreign body. Intracavitary metastatic heart tumors are often covered by thrombotic material.[1] In contrast to echocardiography, both CT and MRI partly enable tissue differentiation between solid, liquid, hemorrhagic, or fatty lesions, and thus metastases can be better depicted.[16-8] In our case, the intracardiac mass was inhomogeneous, but partly resembled thrombus in both TTE and TEE images. However, the mass showed partial enhancement on MRI examination. Therefore, we considered the mass metastatic intracardiac Ewing's sarcoma covered with thrombotic material. The definite diagnosis can only be made by pathological examination of a biopsy sample, but in some cases this may not be feasible. In such subjects, TTE, TEE, CT, and MRI may provide diagnostic information. In our case, TTE was the primary screening technique and MRI provided additional information.

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