Characterization of Cardiac Sarcoma With 2- and 3-Dimensional Echocardiography, Myocardial Contrast Echocardiography and Cardiac Magnetic Resonance Imaging

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A 57-year–old woman had previously undergone a right hemicolectomy for Duke B adenocarcinoma of caecum. She presented with acute back pain. A magnetic resonance imaging scan of her thoracic spine showed destruction of a vertebra with a tumor deposit at T11 (Figure 1A). She underwent a course of radiotherapy (20 Gray in 5 fractions) to this area. Good symptomatic relief was obtained. Biopsy demonstrated a sarcomatoid tumor suggestive of leiomyosarcoma. Over the next 2 months she developed dyspnea on exertion, fatigue, and leg edema. Examination revealed a
regular tachycardia, raised jugular venous pressure, and bilateral leg edema. On auscultation, an added early diastolic sound (consistent with tumor plop) was audible. A 3-dimensional echocardiogram identified a large mass protruding from the right atrium into the right ventricle (Figure 1B and online-only Data Supplement Movie I). The mass prolapsed through the tricuspid valve into the right ventricle during diastole, intermittently obstructing right ventricular inflow and almost completely occluding the tricuspid valve annulus (Figure 1C and online-only Data Supplement Movie II). To assess the vascularity of the mass, the patient underwent myocardial contrast echocardiography. Limited passage of microbubbles into mass (arrow shows partial enhancement) suggests low vascularity tumor (Figure 1D). A trans-

Figure 2. A, Transesophageal echocardiogram. Mass is seen filling the superior vena cava (SVC) and extending into right atrium (RA). LA indicates left atrium. B, Cardiac magnetic resonance imaging (CMR). Mass is seen originating from superior vena cava almost occluding the vessel. RV indicates right ventricle; PA indicates pulmonary artery. C, CMR demonstrated large, mobile, multilobulated mass extending from RA into RV. LV indicates left ventricle. D, Gross morphological of surgically excised mass shows well-circumscribed, lobulated, firm mass.

Figure 3. A, Histopathologic examination of the mass identified malignant pleomorphic cells (hematoxylin-eosin stain, original magnification, ×600). B, Immunostaining for smooth muscle actin in malignant cells (brown color) confirmed the diagnosis of leiomyosarcoma.
esophageal echocardiogram identified the mass originating from the superior vena cava (Figure 2A). Cardiac magnetic resonance imaging demonstrated that the mass almost completely filled the superior vena cava (Figure 2B) and was continuous with a large, mobile multilobulated mass within the right atrium and ventricle (Figure 2C and online-only Data Supplement Movie III). Because the mass was obstructing right ventricular inflow and producing signs and symptoms of right heart failure, it was surgically excised for symptomatic relief. The resected mass was well circumscribed and lobulated (Figure 2D). Histology of the excised mass showed pleomorphic spindle cell tumor with extensive necrosis, thrombosis, and infarction (Figure 3A and 3B). The tumor cells were positive for smooth muscle actin, which was consistent with a pleomorphic sarcoma with myofibroblastic differentiation. The patient made an excellent postoperative recovery with resolution of symptoms.

Sarcomas affecting the heart or great vessels are extremely rare, with a reported prevalence of 0.017%. The most common histological subtype is angiosarcomas. Tumors may grow up the inferior vena cava to invade the right atrium and ventricle, typically intravascular leiomyomatosis. Tumors of the superior vena cava or azygous vein can also occupy the right heart and, as in this case, potentially obstruct the right ventricular outflow tract. The optimal management strategy for these patients is not well defined, partly because of a paucity of data. Management strategies (including surgical resection, chemotherapy, and radiotherapy) are individualized depending on tumor size, location (including origin and extension), mobility, vascularity, and local invasion. Newer echocardiographic techniques may provide incremental information over 2-dimensional techniques. Three-dimensional echocardiography offers the ability to view structures from multiple imaging planes and allows assessment of morphology, including mass size, shape, location, attachments, and origin. Myocardial contrast echocardiography allows for improved definition of intracavity structures and assessment of vascularity. Together with cardiac magnetic resonance imaging, this case demonstrates how the integration of these techniques allows for tissue characterization and detailed morphological evaluation of intracardiac tumors, which are essential for determining suitability for different management strategies and planning of surgical techniques.

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Disclosures
None.

Reference
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