A 42-year-old woman presented to the emergency department with recurrent episodes of palpitation. The physical examination and the ECG were unremarkable. Echocardiographic examination revealed a solid mass compressing the right cardiac chambers (Movie I in the online-only Data Supplement). A contrast-enhanced computed tomography of the thorax confirmed the presence of a soft tissue mass measuring 8×6×9 cm in transverse × anteroposterior × craniocaudal dimensions, in the right atrioventricular groove (Figure 1). It contained no calcifications, and no vascular connection with adjacent structures was identified. There was no pericardial effusion.

Cardiac magnetic resonance imaging confirmed an inoperable mass in the right atrioventricular groove surrounding the right coronary artery. The mass was isointense to myocardium on T1-weighted imaging (Figure 2) and hyperintense on T2-weighted imaging and on short TR inversion recovery images (Figure 3). It demonstrated patchy areas of enhancement on first-pass perfusion magnetic resonance imaging (Figure 4 and Movie II in the online-only Data Supplement) that increased in size on delayed gadolinium enhancement with a nodular appearance (Figure 5). Although the mass caused significant compression of the right cardiac chambers (Movie III in the online-only Data Supplement), the right ventricular systolic function was preserved.

A transthoracic computed tomography-guided biopsy was attempted but interrupted because of intraprocedural development of chest pain associated with a small hemopericardium. A surgical biopsy was subsequently performed. The pathology specimen revealed a lesion with vascular channels (Figure 6). The immunohistochemistry stains were consistent with a pericardial lymphangiohemangioma (Figure 7).

During the surgical biopsy the mass was deemed to be unresectable because of its close contact with the myocardium and right coronary artery. The mass has now been followed for >8 years with a minimal increase in size. The patient reports occasional episodes of palpitation not consistent with arrhythmia on Holter monitoring, and the right ventricular systolic function and volumes remain normal.

Lymphangiohemangioma is a very rare slow-flow vascular malformation containing both venous and lymphatic elements.1–3 Its imaging characteristics depend on the amount of both venous and lymphatic components. Lymphatic structures appear hyperintense on T2-weighted images because of prominent cystic spaces and do not demonstrate enhancement.2 Venous components are usually of intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted images sequences and demonstrate progressive enhancement after gadolinium administration, indicating trapped gadolinium within the abnormal venous channels.2,3 To our knowledge, this represents the first case of pericardial lymphangiohemangioma reported in the literature. Thirteen cases of mediastinal lymphangiohemangioma, mostly located in the anterior mediastinum, have been reported.3 These lesions are rarely biopsied because of the risk of bleeding and are most commonly diagnosed and treated surgically.3

Benign vascular malformations should be considered in the differential diagnosis of a vascularized soft tissue mass in the right atrioventricular groove, together with other more aggressive entities, such as angiosarcoma, lymphoma, and metastatic disease. Cardiac magnetic resonance is paramount in providing information about tissue characteristics and cardiac function. Computed tomography and magnetic resonance provide complementary information to help surgical planning for resection or to obtain tissue diagnosis.

Disclosures

None.

References


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Figure 1. Contrast-enhanced multidetector computed tomography image demonstrates a well-circumscribed soft-tissue mass in the right atrioventricular groove. The mass causes significant compression of the right atrium (RA) and right ventricle (RV).

Figure 2. Axial T1-weighted fast spin echo magnetic resonance image demonstrates an isointense mass to myocardium in the right atrioventricular groove. Note that the right coronary artery is embedded within the mass (arrow).

Figure 3. Coronal oblique short T1 inversion recovery magnetic resonance image demonstrates a markedly hyperintense mass. Note the right coronary artery is nicely depicted traversing the mass (arrow). RA indicates right atrium; RV, right ventricle; and RVOT, right ventricular outflow tract.
Figure 4. Sagittal oblique first pass perfusion magnetic resonance image in the coronal oblique plane demonstrates several patchy areas of enhancement (arrows) indicating the vascular nature of the mass (Movie II in the online-only Data Supplement).

Figure 5. Late gadolinium enhancement phase-sensitive inversion recovery image in axial plane shows nodular areas of enhancement within the mass likely attributed to pooled gadolinium within the vascular channels.

Figure 6. Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) from a surgical biopsy of the lymphangiohemangioma. The specimen demonstrates dilated vascular channels (arrowhead) containing blood and lymphoid tissue (arrow) revealing the mixed nature of the lesion.

Figure 7. Immunohistochemistry of lymphangiohemangioma (original magnification, ×100). Top, Blood vessel endothelium immunopositivity for CD31 (arrows). Bottom, Immunopositivity for D2-40, a marker of lymphatic vessel lining cells (arrows).
Pericardial Lymphangiohemangioma: Multimodality Imaging Features and Pathologic Correlation
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