Diffuse Large B-Cell Lymphoma

Presenting as a Cardiac Mass

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A 59-year-old immunocompetent man presented at the emergency department, reporting several days of constant substernal chest pain without recent weight loss, night sweats, or fever. Clinical examination and laboratory results yielded nothing unusual. No adenopathy was identified. An electrocardiogram revealed T-wave inversion in the inferior and anterolateral leads.

A chest computed tomogram showed diffuse thickening of the right ventricular (RV) myocardium. No lymphadenopathy or pleural effusion was noted. Cardiac magnetic resonance (CMR) images revealed diffuse RV myocardial hypertrophy (Fig. 1); the maximum RV myocardial thickness was 2 cm during end-diastole. The RV end-diastolic volume was 213 mL (indexed value, 104 mL/m^2), its mass was 210 g (indexed mass, 102 g/m^2), and the ejection fraction was 0.37. The RV myocardium had an intermediate T2 signal and contained no fat. Mild circumferential pericardial effusion was detected. During first-pass perfusion, gadolinium uptake was biventricular (Fig. 2); delayed spin-echo images showed greater gadolinium enhancement in the RV than in the left ventricle (LV) (Fig. 3). The LV interventricular septum was 1.3 cm thick, but the LV otherwise had normal size, systolic function, and mass. Delayed-enhancement CMR revealed biventricular diffuse patchy enhancement (Fig. 4).

Although we initially suspected biventricular hypertrophic cardiomyopathy, the delayed-enhancement pattern suggested an infiltrative process. Coronary angiograms showed no coronary disease. Biopsy specimens of the RV myocardium had a hard consistency; histologic analysis revealed diffuse large B-cell lymphoma with myocyte necrosis. The patient underwent 6 cycles of chemotherapy with rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone, and remained in remission 2 years after diagnosis.

Comment

Diffuse large B-cell cardiac lymphoma (typically non-Hodgkin) accounts for only 1% of primary cardiac tumors.[1] Primary cardiac lymphoma should be suspected when

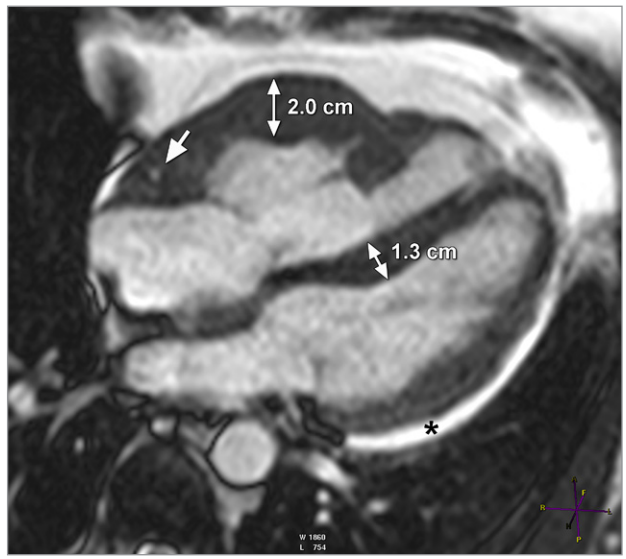

Fig. 1 *Cardiac magnetic resonance steady-state free-precession gradient-echo sequence (4-chamber view in end-diastole) shows 2-cm-thick myocardium at the right ventricular free wall and a 1.3-cm-thick interventricular septum. Encasement of the mid right coronary artery is present, best seen in the right atrioventricular groove (arrow). A small pericardial effusion (*) is also present.*

Supplemental motion image is available for Figure 1.

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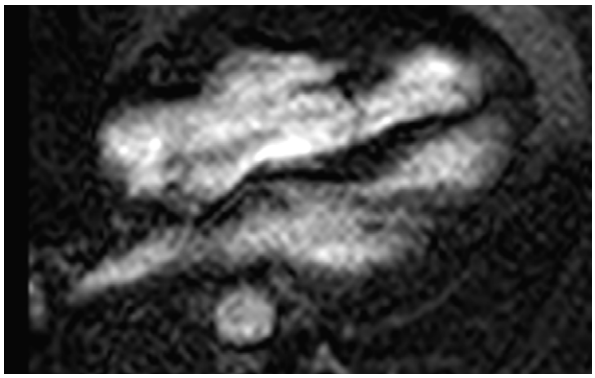

Fig. 2 *Cardiac magnetic resonance first-pass perfusion scan (4-chamber view), obtained with use of a fast gradient-echo sequence and gadolinium enhancement, reveals limited enhancement immediately after gadolinium bolus administration. Biventricular uptake of contrast is better seen in the supplemental motion image.*

Supplemental motion image is available for Figure 2.

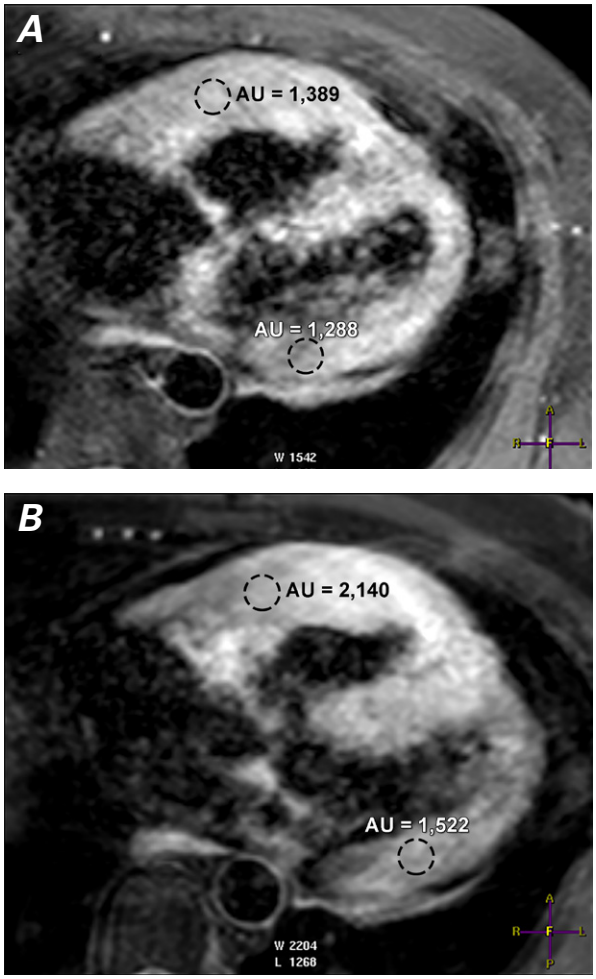

Fig. 3 *Cardiac magnetic resonance spin-echo images obtained **A**) before and **B**) after gadolinium-chelate administration. The latter image shows more enhancement in the right ventricle than in the left ventricle by region-of-interest measurement.*

AU = arbitrary unit

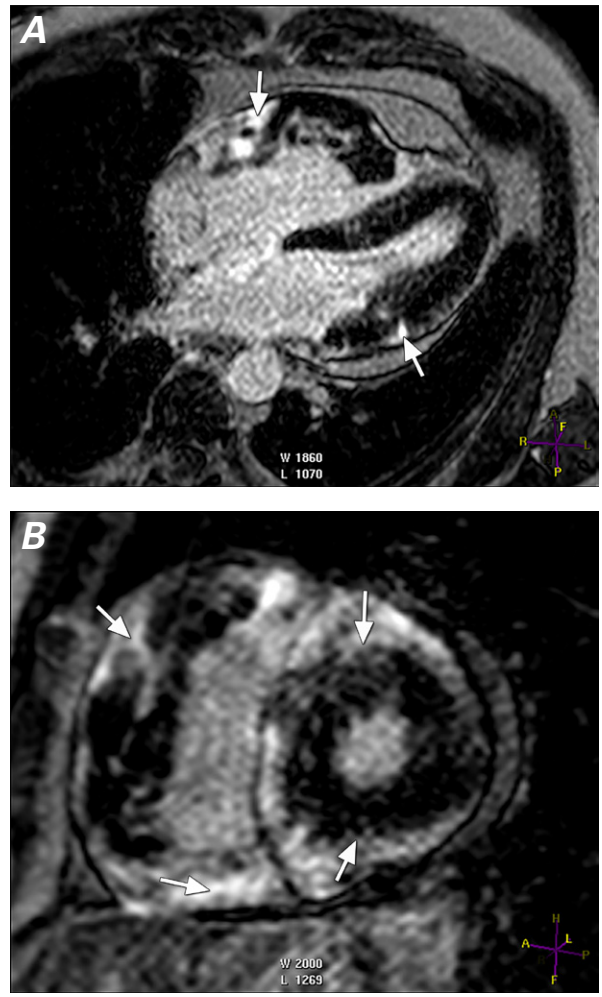

Fig. 4 *Delayed-enhancement cardiac magnetic resonance images in the **A**) 4-chamber and **B**) short-axis views, acquired approximately 15 minutes after gadolinium-chelate administration, show patchy enhancement (arrows) in both ventricles, suggesting biventricular involvement.*

no lymphoma is detected outside the pericardial sac, the tumor is largely inside the pericardium, or there is symptomatic lymphomatous cardiac infiltration at diagnosis. Typically, when patients present with symptoms, the diagnosis is late and the prognosis is poor.[2] Primary cardiac lymphoma most often involves the right-sided cardiac chambers and can present as cardiac failure or pericardial effusion. Primary cardiac lymphoma involving the vena cava can cause obstructive symptoms, as well as valvular and conduction disturbances and embolism.

Echocardiography, the preferred imaging method, frequently shows the tumor mass well. Because CMR requires no geometric assumptions and can characterize tissue, CMR is also well-suited for evaluating cardiac masses, especially in the RV. In this case, both computed tomography and CMR revealed RV abnormalities, and delayed-enhancement CMR showed biventricular patchy enhancement, confirming ventricular lymphomatous infiltration.

References

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2. Ikeda H, Nakamura S, Nishimaki H, Masuda K, Takeo T, Kasai K, et al. Primary lymphoma of the heart: case report and literature review. Pathol Int 2004;54(3):187-95.