A 59-year-old male was admitted to Massachusetts General Hospital, Boston, Mass, with a 2-month history of exertional dyspnea (New York Heart Association class II to III). The patient denied dyspnea at rest, chest pain, palpitations, or syncope. There was no history of fevers or recent weight loss.

An outpatient echocardiogram (Figure 1), performed as part of the workup of the patient’s dyspnea, demonstrated normal left ventricular size and function. The right ventricle (RV) was normal in size but diffusely hypokinetic. There was evidence of segmental RV dysfunction, with 2 discrete aneurysmal areas in the RV free wall at the base and apex, which measured 1.5 and 3.0 cm in width. Both areas appeared thinned and dyskinetic. The echocardiographic appearance was suggestive of arrhythmogenic RV dysplasia/cardiomyopathy (ARVD/C). A CT scan ruled out the presence of pulmonary embolism but was notable for marked mediastinal lymphadenopathy (Figure 2).

One week later, the patient developed intermittent Mobitz type II second-degree atrioventricular block that prompted his admission to the hospital for monitoring and further workup. An admission ECG (Figure 3) showed normal sinus rhythm with a heart rate of 45 bpm and Mobitz type II second-degree atrioventricular block with 2:1 conduction. Chest radiograph findings (Figure 4) were unremarkable. All laboratory tests were normal, including troponin and ACE levels.

Cardiac magnetic resonance imaging (Figures 5 and 6) confirmed the presence of the RV aneurysms and segmental hypokinesis. Delayed hyperenhancement of the basal portion of the RV free wall, the RV border of the midventricular septum, and the subepicardial portion of the basal and apical inferolateral walls of the left ventricle was noted. These findings placed into doubt the diagnosis of ARVD/C and suggested the possibility of cardiac sarcoidosis.

An endomyocardial biopsy (Figure 7) was performed that targeted the locations of delayed hyperenhancement, observed by MRI, within the RV. It revealed extensive fibrosis with giant cells, compact noncaseating granulomas, and an accompanying lymphocytic infiltrate consistent with cardiac sarcoidosis. 2-[18F]fluoro-2-deoxy-D-glucose positron emission tomography scanning was conducted (Figure 8) to evaluate the extent of disease, which demonstrated intense uptake in the RV and patchy uptake in the left ventricle. Prominent uptake in multiple thoracic lymph node groups...
was also noted. A course of steroid therapy was initiated, with an interval resolution of symptoms and conduction abnormalities after 10 days of therapy.

ARVD/C is a rare condition characterized by fibrofatty infiltration, typically of the RV, although rarely, the left ventricle can also be involved. The disease has a predilection for the RV outflow tract, the RV apex, and the posterior subtricuspid region, often referred to as the “triangle of dysplasia.” These regions may demonstrate hypocontractility or even aneurysm formation. ARVD/C is suspected in individuals who have ventricular arrhythmias or arrhythmic sudden death and when there is generalized dilatation or localized abnormalities of the RV in the presence of preserved left ventricular size and function. The diagnosis can be difficult, however, when there are subtle structural changes in the RV.

It is seldom appreciated that myocardial involvement in sarcoidosis can mimic some of the clinical and structural abnormalities of ARVD/C. We report a case of a patient diagnosed echocardiographically as having RV dysplasia but confirmed pathologically as having cardiac sarcoidosis.

Disclosures

None.

References


Figure 2. CT of the chest demonstrating marked mediastinal lymphadenopathy (arrows).

Figure 3. ECG performed on admission to hospital demonstrating sinus rhythm and Mobitz type II second-degree atrioventricular block with 2:1 conduction.
Figure 4. Chest radiograph demonstrating normal lung parenchyma, cardiopericardial silhouette, and mediastinal structures.

Figure 5. Four-chamber view with steady-state free precession acquisition (SSFP) MRI demonstrating 3 discrete aneurysmal portions of the RV free wall (arrows).

Figure 6. Delayed gadolinium-enhanced MRI demonstrating delayed enhancement primarily of the RV border of the interventricular septum (short arrows) and the inferolateral wall of the left ventricle (long arrow). Pooling of gadolinium is demonstrated in an aneurysmal portion of the RV (arrowhead).

Figure 7. Endomyocardial biopsy of the RV septal wall. Hematoxylin-and-eosin–stained histological sections at 200× magnification showing extensive fibrosis with giant cells (arrow) and an accompanying lymphocytic infiltrate.

Figure 8. 2-[18F]fluoro-2-deoxy-D-glucose positron emission tomography scan demonstrating intense uptake in the RV and patchy uptake in the left ventricle.
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