A 32-year-old man was referred to our medical center for chest pain and dyspnea of 2 days duration. Past medical history was unremarkable. Vital signs and physical examination were normal. ECG (Figure 1) showed a right bundle-branch block but no changes suggestive of ischemia. Troponin T level was 4.01 ng/mL (normal: <0.10) with a total creatine kinase of 534 U/L (normal: 40 to 200). Coronary angiography revealed nonobstructive disease. Transthoracic echocardiography demonstrated asymmetrical septal hypertrophy with mildly depressed left ventricular function and moderately depressed right ventricular function (Figure 2 and online-only Data Supplement Movies I and II).

Serum electrolytes, blood urea nitrogen, and creatinine were normal. Aspartate transaminase was 81 U/L, but the remainder of the patient’s liver function tests were normal. Total white blood cell count was 2600/mm³ with a normal cell differential; the rest of the complete blood count was normal. Serum and urine protein electrophoresis and immunofixation were normal. Anti-nuclear antibody screen was 1:40. HIV-1,2 antibodies were nonreactive. Angiotensin-converting enzyme level was normal at 10 U/L. Abdominal fat pad biopsy did not demonstrate amyloidosis. Bone marrow biopsy revealed a mildly hypocellular marrow. Acid fast stain and culture as well as viral cultures failed to grow any microorganisms. Extensive serological screening for infectious organisms was unrevealing.

Cardiac magnetic resonance imaging (Figure 3) showed abundant delayed hyperenhancement in the left ventricular myocardium. Magnetic resonance imaging–targeted biopsy of the interventricular septum revealed giant cell myocarditis.
Over the next 3 days, the patient developed overt congestive heart failure with frequent runs of monomorphic ventricular tachycardia and progressive right and left ventricular dysfunction (Figure 5 and online-only Data Supplement Movies III and IV). Treatment commenced with an aggressive immunosuppressive regimen that included methylprednisolone, cyclosporine A, and OKT3. Despite this treatment, his condition continued to deteriorate. On hospital day 20, he underwent successful orthotopic heart transplantation. Follow-up biopsies over the next 12 months documented episodes of mild cellular rejection but no recurrence of giant cell myocarditis.

First described in 1905, idiopathic giant cell myocarditis (IGCM) is a rare inflammatory disorder of the myocardium. Clinical features can include chest pain, fever, rapidly progressive heart failure, refractory ventricular arrhythmias, and heart block. These adverse events are known to occur with...
much greater frequency when compared with lymphocytic myocarditis. Most patients experience a fulminant course with a median survival from symptom onset of 3 months without treatment. Prognosis of non-GCM fulminant myocarditis appears to be significantly better, and distinction between these forms of myocarditis as well as infectious causes is important.

Classic histopathological description includes serpiginous regions of myonecrosis with multinucleated giant cells at the margins of necrosis and absence of well formed granuloma. The sensitivity of right ventricular endomyocardial biopsy in a subset of patients in the Multicenter Giant Cell Myocarditis Registry was 80% to 85%.

A scant amount of information exists in the medical literature on magnetic resonance imaging findings in GCM. A sensitive test to diagnose GCM is vitally needed because of the fulminant course of the disease and its poor short-term prognosis. The ability to noninvasively risk stratify patients presenting with acute heart failure will assist the clinician in decisions on endomyocardial biopsy, immunosuppressive therapy, ventricular assist devices and transplantation. Cardiac magnetic resonance imaging may provide this information. As experience with this technique in nonischemic cardiomyopathies and myocarditis evolves, patterns and extent of myocardial involvement, as demonstrated with delayed-enhancement techniques, may provide diagnostic and prognostic information. In the case described, the impressive amount of myocardial heterogeneity, edema, and delayed hyperenhancement, not initially suspected based on the appearance of his transthoracic echocardiogram, accelerated plans for an endomyocardial biopsy and treatment.

Disclosures

Dr Stolpen has received honoraria (<$10 000) from GE Healthcare, Bracco, and Siemens. He has served as a consultant to GE Healthcare and Bracco. The remaining authors report no conflicts.

References


Figure 5. Echocardiogram 14 days after admission. Long- and short-axis images demonstrate resolution of septal hypertrophy, overt chamber dilation, and systolic dysfunction. Cine images can be viewed in online-only Data Supplement Movies III and IV.
Evanescent Asymmetrical Septal Hypertrophy and Rapidly Progressive Heart Failure in a 32-Year-Old Man

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