Thrombotic Thrombocytopenic Purpura
Inducible Myocardial Ischemia Depicted on Dipyridamole Stress First-Pass Perfusion Cardiac Magnetic Resonance Despite Normal Myocardial Stress Single-Photon Emission Computed Tomography

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A 26-year-old black woman, without risk factors for coronary artery disease, was admitted in the cardiology department of our institution for acute chest pain. Three months before admission, she had an episode of abdominal pain along with fever, thrombocytopenia (platelet count 23 000/mm³), and hemolytic anemia (hemoglobin 5.1 g/dL). Thrombotic thrombocytopenic purpura was diagnosed with ADAMTS-13 (a disintegrin and metalloproteinase with a thrombospondin type 1 motif, member 13) activity of <5% and presence of anti-ADAMTS-13 antibody activity. Severe thrombocytopenia (<50 000/mm³) persisted despite intensive treatment with high-dose steroids, rituximab (anti-CD20 an-

Figure 1. A, Resting 12-lead ECG on admission showing nonspecific, permanent, and diffuse negative T waves (heart rate 81 bpm, blood pressure 100/70 mm Hg). B, Stress 12-lead ECG showing diffuse 1-mm ST-segment depression in leads V4 through V6 and leads DII, DIII, and VF (heart rate 135 bpm, blood pressure 125/75 mm Hg).
tibody), and vincristine. Splenectomy was then performed, and the patient underwent daily plasma exchange along with repeated intravenous-bolus cyclophosphamide. She was admitted in the cardiology department because of acute chest pain that lasted ≈20 minutes and was resistant to sublingual nitrate, with stable, diffuse, negative T waves on ECG (Figure 1A). The test for troponin I twice showed negative results (<0.04 mg/mL). Transthoracic echocardiography was normal. Hemoglobin (13.2 g/dL) and platelet count (367,000/mm³) were stable, and antiplatelet therapy was administered (aspirin 250 mg/d).

Stress dipyridamole (0.84 mg/kg) ⁹⁹Tc sestamibi single-photon emission CT revealed significant ST-segment depression in leads V₅ and V₆ on stress ECG (Figure 1B) but no evidence of inducible myocardial ischemia or myocardial infarction (Figure 2A). Because of this discrepancy and because of suspected thrombotic microangiopathy, dynamic multislice first-pass perfusion dipyridamole (0.84 mg/kg) stress cardiac magnetic resonance imaging (CMR; 1.5 T Magnetom Avanto, Siemens, Erlangen, Germany) was performed, which showed diffuse areas of hypoenhancement predominating in the subendocardium that did not correspond to any coronary territory (Figure 2B). These patterns persisted over the minute of acquisition of stress CMR perfusion images (Movie 1). Matching short-axis views of cine CMR showed normal global and segmental LV function (Movie 1). Contrast-enhanced inversion-recovery gradient echocardiography CMR images acquired 10 minutes after intravenous injection of 0.1 mmol/kg gadolinium chelates (gadoterate meglumine, Dotarem, Guerbet, Aulnay, France) showed no late enhancement anywhere in the left ventricular myocardium (Figure 3). Black-blood spin-echo T2-weighted CMR images were acquired before contrast injection at the same location and showed no myocardial edema (Figure 4). Because of inducible myocardial ischemia and low risk for coronary artery disease, a 64-row multidetector coronary CT was performed, which ruled out the presence of significant coronary stenosis. The patient was treated with aspirin (250 mg/d), and cardiac follow-up was uneventful.

Acute myocardial ischemic events may occur during the course of thrombotic thrombocytopenic purpura due to the thrombotic occlusion of the coronary microvasculature. Microvascular thrombi in arterioles and capillaries of the myocardium consist mainly of platelet aggregates, with only small amounts of fibrin. In thrombotic thrombocytopenic purpura, the histological picture of extensive microvascular thrombotic lesions occurs frequently in the myocardium, but clinical cardiac manifestations are few, and objective findings of myocardial injury have thus far been limited to ECG changes. After acute coronary syndrome occurring in a

Figure 2. A, Left ventricular short-axis dipyridamole ⁹⁹Tc sestamibi single-photon emission CT images from base to apex showing no evidence of inducible myocardial ischemia or myocardial infarction. B, Left ventricular short-axis diastolic still frames extracted from dipyridamole stress dynamic first-pass perfusion CMR showing multiple and diffuse foci of hypoenhancement in the subendocardium (arrows).

Figure 3. Corresponding inversion-recovery fast-gradient echocardiography CMR images obtained 10 minutes after gadolinium injection, showing no focal area of hyperenhancement, which indicates the absence of myocardial infarction or fibrosis.
patient with thrombotic thrombocytopenic purpura, small foci of inducible myocardial ischemia were depicted on dipyridamole stress perfusion CMR despite an unremarkable dipyridamole stress SPECT study. As is commonly seen in diseases with thrombotic microangiopathy, myocardial ischemia was diffuse throughout the left ventricle, in a noncoronary distribution, and predominant in the subendocardium. Late-enhancement CMR showed an absence of myocardial infarction. This observation illustrates the potential of stress perfusion CMR, because of its good spatial resolution and contrast, to be useful in the challenging imaging of myocardial ischemia in conditions with thrombotic microangiopathy. The implications of these findings in terms of therapy are still unknown.

Disclosures

None.

Reference

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