A 39-year-old man, with a temperature of 38.5°C and sinus tachycardia, was admitted for work-up of chest pain. He had a history of asthma, recurrent pneumonia, sinusitis, and nasal polyposis. Clinical examinations, ECG (Figure 1) and chest x-ray (Figure 2) on admission were suspicious for perimyocarditis. Routine blood analysis revealed an elevated erythrocyte sedimentation rate (88 mm/h; normal <15 mm/h) and a normal leukocyte count (7200/mm³ with 21% eosinophilic granulocytes (normal 1% to 6%). Levels of C-reactive protein and immunoglobulin E were elevated at 14.0 mg/dL (normal 0.1 to 0.5 mg/dL) and 237 U/mL (normal <100 U/mL), respectively. Thus, Churg-Strauss syndrome with perimyocardial involvement was suspected.

Because echocardiography could not provide any information on myocardial involvement in this case (Figure 3; for full-motion images, see Movie I in the online-only Data Supplement), the patient was referred for cardiovascular magnetic resonance imaging (CMR; 1.5 Tesla Sonata, Siemens Medical Systems, Erlangen, Germany). Cine images were acquired using fast-gradient echo steady-state free precession sequences that demonstrated increased pericardial thickness (5 mm), as well as small amounts of pericardial effusion (Figure 4). Systolic left ventricular (LV) function was mildly impaired (LV end-diastolic volume 144 mL, LV end-systolic volume 59 mL, and LV ejection fraction 59% (Figure 4; Movie II in the online-only Data Supplement). Ten minutes after injection of 0.2-mmol/kg gadodiamide (Omnis...
can, Amersham-Health, Braunschweig, Germany) contrast CMR was performed using an inversion recovery gradient echo technique (IR-FLASH), constantly adjusting inversion time to null normal myocardium. Late gadolinium enhancement was present in the basal and midventricular lateral wall, as well as in the septum, and distributed in the subendocardial, intramural, and subepicardial myocardium, as demonstrated in Figure 4.

Before initiating immunosuppressive therapy, coronary artery disease, as well as myocardial infection with cardiotropic viruses or bacteria, was ruled out by coronary angiography and CMR-guided endomyocardial biopsies. Histopathological work-up of biopsy samples demonstrated eosinophilic infiltrates in combination with tissue edema and myocardial necrosis (Figure 5), confirming the suspected diagnosis of Churg-Strauss syndrome with cardiac involvement as defined by American College of Rheumatology criteria. Interestingly, the subepicardial, intramural, and subendocardial pattern of late gadolinium enhancement demonstrated by contrast CMR nicely matches the pattern of scattered patches of reddish-gray discoloration representing eosinophilic infiltrates and hemorrhagic necrosis on the necropsy sample of a different patient who died suddenly from Churg-Strauss syndrome with cardiac involvement (Figure 6).1

Figure 2. Frontal posterior–anterior chest x-ray on admission. Note the enlarged pulmonary hila, as well as the relation of the cardiac silhouette to the lateral distance (16 cm/32.5 cm), suggesting mild cardiac enlargement, pericardial effusion, or a combination of these conditions.

Diastole

Systole

TTE-4CH  TTE-SAX

Figure 3. Transthoracic echocardiogram (TTE) after chest x-ray. Systolic as well as diastolic images of apical long axis (4CH) and parasternal short axis (SAX) are displayed. No pericardial effusion could be detected, and no information on myocardial involvement in suspected Churg-Strauss syndrome could be obtained by echocardiogram.
Figure 4. CMR study at initial presentation. Steady-state free precession CMR images of multiple short- and long-axis views are displayed in the upper 2 rows (diastole and systole). Note the increased pericardial thickness in combination with a small pericardial effusion (black arrows). Contrast CMR images are displayed in the bottom row. Contrast CMR (normal myocardium is black) reveals contrast enhancement, located in the basal and midventricular lateral as well as septal wall and distributed in the subendocardial, intramural, and also subepicardial myocardium (white arrows).

Figure 5. Histopathological work-up of LV endomycocardial biopsy specimen (Giemsa staining), displaying normal myocytes (A), numerous infiltrating clusters of eosinophilic granulocytes (B, revealing red cytoplasmic granules), fibrosis (C, curly collagen fibers), and myocyte necrosis (D).
After 2 cycles of intravenous cyclophosphamide (750 mg) and oral steroids (1.0 mg/kg daily) the patient’s symptoms had resolved. Two months after initiation of treatment, the erythrocyte sedimentation rate had decreased from 88 mm/h to 8 mm/h, and the eosinophilic count had dropped from 21% to 1.6% of granulocytes. Follow-up CMR imaging at this time still demonstrated mildly impaired LV function (LV end-diastolic volume 160 mL, LV end-systolic volume 65 mL, and LV ejection fraction 60%) but normalization of pericardial thickness (2 mm; Figure 7). As in certain patterns of viral myocarditis, late gadolinium enhancement decreased over time in good correlation to clinical improvement (Figure 7; Movie III in the online-only Data Supplement). A quantitative plot of the extent of late gadolinium enhancement using signal intensity detection in the American Heart Association/American College of Cardiology–recommended 17-segment model before and after treatment can be viewed in Figure 8.

Six months after initiation of treatment, LV function had also normalized (LV end-diastolic volume 135 mL, LV end-systolic volume 42 mL, LV ejection fraction 69%). At this time point, ECG abnormalities were decreasing. How-

Figure 7. Follow-up CMR after 2 months of immunosuppressive therapy using the same CMR protocol as described above. After treatment, pericardial thickness (black arrows), pericardial effusion, and contrast enhancement have decreased in good correlation to clinical improvement.
ever, the ECG remained abnormal despite normalization of LV function (Figure 9).

To our knowledge, this is the first case demonstrating that CMR is not only an important tool for diagnosing cardiac involvement in Churg-Strauss syndrome3,4 but may also be useful for directly monitoring myocardial response to medical treatment, independent of LV ejection fraction or ECG abnormalities.

**Disclosures**

None.

**References**

Magnetic Resonance Assessment and Therapy Monitoring of Cardiac Involvement in Churg-Strauss Syndrome
Hannibal Baccouche, Ali Yilmaz, Dominik Alscher, Karin Klingel, Jose Fernando Val-Bernal and Heiko Mahrholdt

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