

Case report

Cardiac involvement of Churg Strauss syndrome demonstrated by magnetic resonance imaging

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Key words: Churg Strauss syndrome, cardiac magnetic resonance, myocarditis, small vessel disease.

Abbreviations

CSS: Churg Strauss syndrome
ECG: electrocardiogram
ceCMR: contrast-enhanced cardiac magnetic resonance imaging

ABSTRACT

Churg Strauss syndrome (CSS) may lead to cardiac involvement in up to 60% of patients. The myocardium, coronary vasculature, valves and pericardium may be affected. This results in significant morbidity and mortality, accounting for 48% of deaths due to CSS. Cardiac magnetic resonance imaging (CMR) is used to evaluate cardiac structure and function, and is able to evaluate myocardial perfusion and delineate scar tissue. We are the first to demonstrate these features in a 53-year-old CSS patient who presented with palpitations and atypical chest pains, and was found to have myocardial perfusion defects and scar tissue, most likely secondary to vasculitis of the small myocardial vasculature and myocardial infiltration.

Introduction

Churg Strauss syndrome (CSS) may lead to cardiac involvement in up to sixty percent of patients (1). CSS may affect the myocardium, coronary arteries, valves and pericardium (Table I).

Cardiomyopathy may result from vasculitis-related ischemia affecting small myocardial vessels and coronary arteries, and from eosinophilic or from granulomatous myocardial infiltration (2-6). Cardiac involvement is the major cause of mortality, accounting for 48% of deaths due to CSS (7).

Cardiac magnetic resonance imaging is used to evaluate cardiac structure and function, and is able to assess myocardial perfusion, and delineate myocardial scar tissue (8). So far the role of CMR in CSS was highlighted in only two case-reports, in which CMR was used to evaluate left ventricular function and delineate subendocardial fibrosis (9, 10).

We present a 53-year-old Churg Strauss patient, who complained of palpitations, and demonstrate the diagnostic potential of contrast-enhanced CMR in the evaluation of myocardial perfusion and myocardial scar in this syndrome.

Case-report

A 53-year old female, who had been

Table I. Cardiac manifestations of CSS.

Vasculitis of the epicardial coronary arteries
Vasculitis of the small intra-myocardial vasculature (i.e. arterioles, capillaries, venules)
Granulomatous myocardial infiltration
Eosinophilic myocarditis
Myocardial fibrosis
Right ventricular endomyocardial fibrosis
Right ventricular an/or left ventricular intra-cavitary thrombus
Restrictive cardiomyopathy
Dilated cardiomyopathy
Mitral regurgitation due to papillary muscle dysfunction
Acute pericarditis
Pericardial tamponade
Constrictive pericarditis
Supra-ventricular arrhythmias
Ventricular arrhythmias
Atrio-ventricular or intra-ventricular conduction disorders
Sudden cardiac death



Fig. 1. The 12-lead ECG demonstrates diffuse ST-Tsegment deviations.

diagnosed with Churg Strauss syndrome (CSS) 7 years earlier, developed complains of palpitations. The diagnosis of CSS was based on a history of bronchial asthma, the presence of hypereosinophilia >10% on blood counts, sinusitis and non-fixed pulmonary infiltrates by X-ray, and evidence of extra-vascular eosinophils and vasculitis on pulmonary and dermal biopsies (11).

Her palpitations occurred mainly at rest, lasted several seconds and were not accompanied by dyspnoea, dizziness or chest pain. Apart from her age, no additional cardiovascular risk factors existed. Her medication consisted of maintenance immunosuppressive therapy, low dose steroids and azathioprine, and ranitidine. Physical examination did not reveal abnormalities, except for a Cushingoid appearance. Laboratory assessment showed a total blood eosinophil count of 2400×10^6 (normal < 440) CRP 2.1 mgr/l (normal < 2), and negative ANCA.

The 12-lead electrocardiogram (ECG) showed a sinus rhythm, with normal electrical axis', and mild generalized ST-segment depressions, with mild ST-segment elevation in lead aVR (Fig. 1). The 24-hour ambulatory electrocardiogram showed frequent premature multi-focal ventricular contractions, and one short non-sustained monomorphic ventricular tachycardia.

Echocardiography revealed hyperechoicity of the interventricular septum and a local, small dyskinetic segment (Fig. 2).

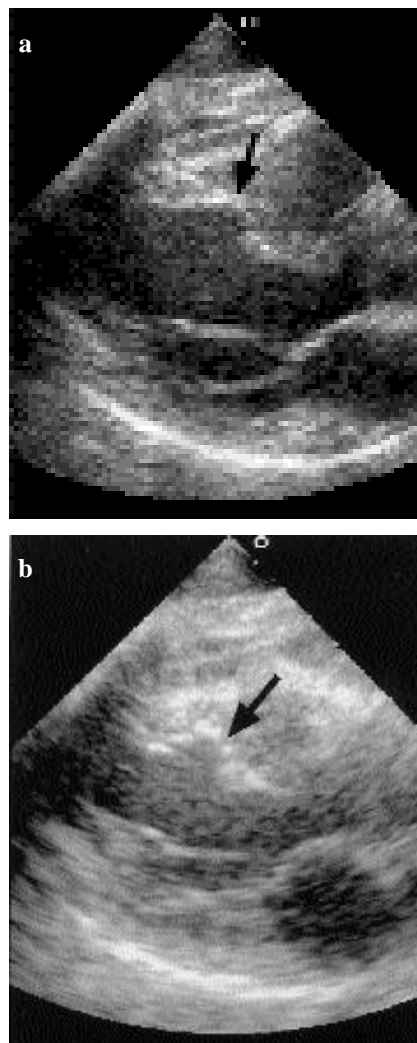


Fig. 2. (a) Echocardiography (parasternal long axis view) demonstrates thinning and dyskinesia of the mid-segment of the inter ventricular septum in early systole. (b) Echocardiography (parasternal long axis view) demonstrates thinning and dyskinesia of the mid-segment of the inter ventricular septum in late systole.

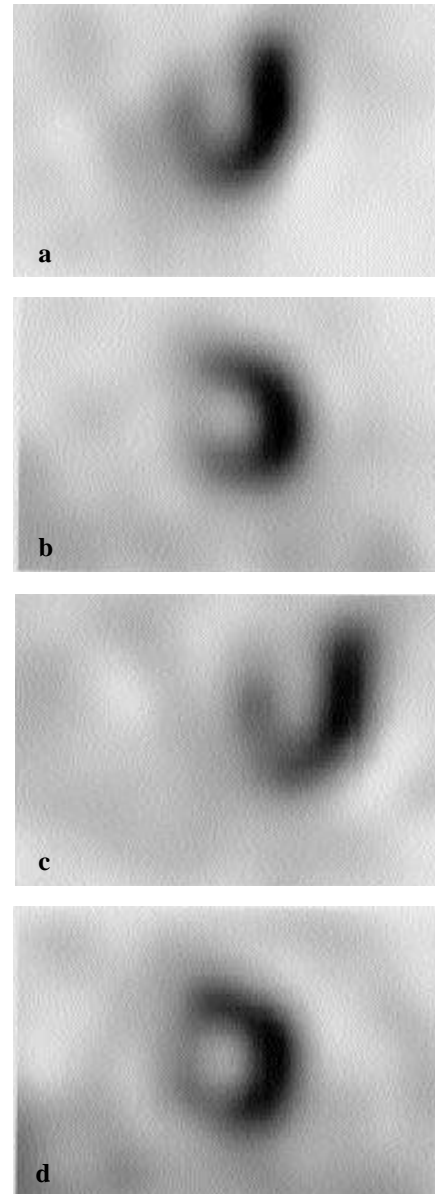


Fig. 3. (a) Thallium scintigraphy after administration of dipyridamol demonstrates a defect in the interventricular septum (4 chamber view). (b) Thallium scintigraphy after administration of dipyridamol demonstrates a defect in the interventricular septum (short axis view). (c) Thallium scintigraphy at rest demonstrates a partly reversible defect in the interventricular septum (4 chamber view). (d) Thallium scintigraphy at rest demonstrates a partly reversible defect in the interventricular septum (short axis view).

Dipyridamole 201 thallium myocard scintigraphy demonstrated a small, partly reversible, defect in the interventricular septum (Fig. 3).

Multi-slice computed tomography did not reveal any calcium deposition in the coronary arteries. Cine MR showed the thin, dyskinetic distal septal seg-

ment, with a normal left ventricular ejection fraction. The T2 weighted study did not reveal signs of active inflammation. First-pass perfusion magnetic resonance imaging confirmed the presence of an irreversible perfusion defect in the interventricular septum and also demonstrated a reversible perfusion defect in the anterolateral wall (Fig. 4).

Ten minutes after administration of gadolinium-DTPA (Schering, Berlin), myocardial enhancement persisted in the interventricular septum, signifying local fibrosis (Fig. 5).

Nitrates and beta-blockade were added to her medication. While awaiting elective coronary angiography, she developed complaints of atypical chest pain at rest and was hospitalized. The ECG was unchanged, and cardiac enzymes, including troponin T, remained normal. Coronary angiography did not reveal abnormalities in the epicardial coronary arteries (Fig. 6).

She is currently followed up at the cardiac outpatients clinic, and remains free from cardiovascular symptoms.

Discussion

CSS is classically characterized by hypereosinophilia and systemic necrotizing vasculitis in patients with previous allergic rhinitis or bronchial asthma, and consists typically of three phases. The prodromal phase consists of allergic rhinitis, nasal polyposis, and airway irritability. This is followed by a period of peripheral and tissue eosinophilia, commonly associated with pulmonary infiltrates. The third phase involves systemic, necrotizing vasculitis (2, 4). Cardiac involvement occurs frequently in CSS, and results in significant morbidity and mortality (1-4, 6, 12). CSS may affect the epicardial and intramyocardial vasculature, the myocardium, valves and pericardium, and leads to structural changes and functional impairment. (1-4, 12) (Table I).

Coronary arteritis is generally part of systemic vasculitis, but rarely an isolated manifestation of CSS, and can cause myocardial infarction (5, 13).

Eosinophil infiltration of the myocardium with the release of toxic inflammatory mediators, cationic protein, major basic protein, eosinophil-derived neu-

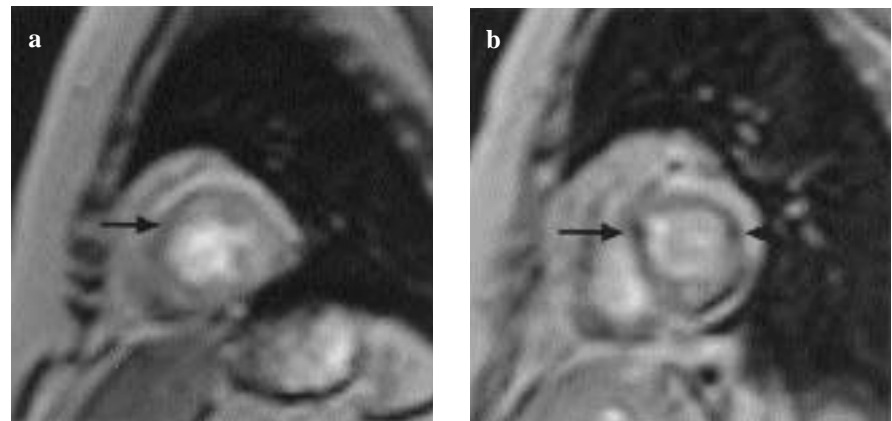


Fig. 4. (a) First-pass perfusion CMR at rest demonstrates an irreversible perfusion defect in the septum (left arrow). (b) First-pass perfusion CMR during administration of adenosine demonstrates perfusion defects in the septum (left arrow) and the anterolateral wall (right arrow).

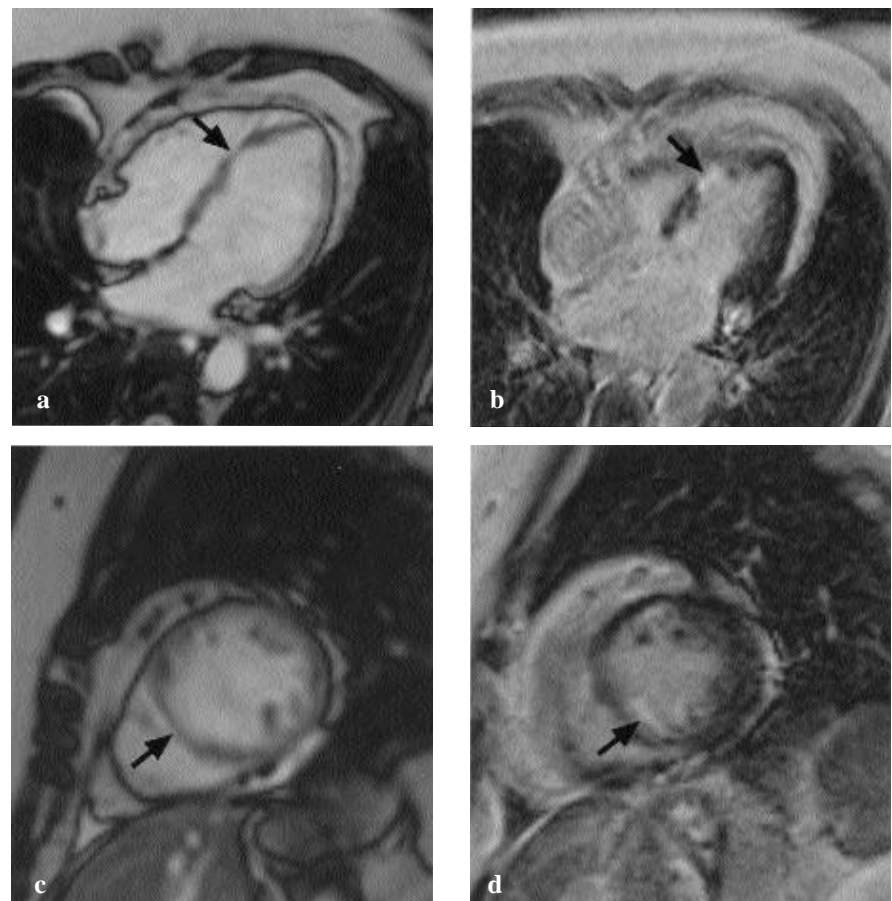


Fig. 5. (a) Left upper panel (SSPF, 4 chamber view): thinning of the distal interventricular septum. (b) Left lower panel (SSPF, short axis view): thinning of the interventricular septum. (c) Right upper panel (4 chamber view, 3-D breath hold IR GRE, inversion time 240 msec, 10 minutes after administration of 0.2 mmol/kg gadolinium-DTPA): enhancement of the interventricular septum. (d) Right lower panel (short axis view, 3-D breath hold IR GRE, inversion time 240 msec, 10 minutes after administration of 0.2 mmol/kg gadolinium-DTPA): enhancement of the interventricular septum.

rotoxin and eosinophil peroxidase, causes myocyte destruction, and results in loss of systolic function and (lethal) arrhythmias (14). Eventually replace-

ment fibrosis leads to restrictive or dilated cardiomyopathy (9). Churg and Strauss reported in their original autopsy series that many of their patients

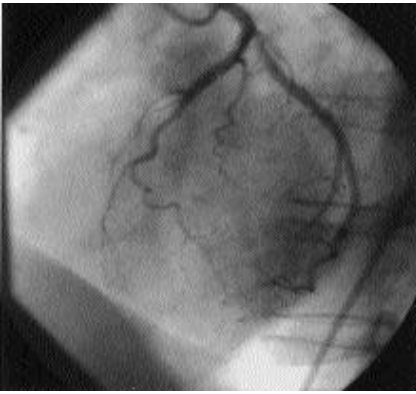


Fig. 6. Diagnostic coronary angiography demonstrates a normal left coronary artery.

died of congestive heart failure (1). Aggressive management with corticosteroids can result in restoration of pump function and resolution of pericardial effusions (2,4,12). The addition of cyclophosphamide or azathioprine serve as steroid sparing agents (6, 9). Cardiac evaluation has traditionally been performed by echocardiography, angiography and myocardial biopsy. Contrast-enhanced cardiac magnetic resonance imaging (ceCMR) has been shown to be of great diagnostic benefit in the assessment of patients with coronary artery disease, cardiomyopathy and myocarditis (8). Particularly the ability of ceCMR to assess pericardial thickness and effusion, regional myocardial contractile (dys)function, perfusion and fibrosis make it a unique and very potent tool in the evaluation of patients with CSS. T2-weighted imaging might help differentiate active inflammation from fibrosis (15). So far two case-reports highlighted the value

of CMR in the assessment of a patient with cardiac involvement in CSS (9, 10). We currently report on the value of first-pass perfusion and late enhancement in the assessment of myocardial perfusion and myocardial scar in CSS. Our patient presented with atypical chest pain and palpitations due to frequent PVCs, in the presence of ECG changes suggestive of myocardial ischemia, (partly) reversible perfusion defects, and myocardial scar in the inter-ventricular septum.

The absence of abnormalities in the epicardial coronaries suggest small vessel vasculitis-related ischemia. Panting *et al.* were the first to demonstrate the value of first-pass perfusion CMR in patients with disease of the small myocardial vasculature (16).

These abnormalities have, to the best of our knowledge, not yet been described in a patient diagnosed with CSS, a disease known to affect the small myocardial vessels. This case report highlights the important role that ceCMR may play in the cardiac assessment of patients with CSS. Future, prospective studies will help us determine the value of this technique in improving the management and outcome of patients with this disease.

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