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CASE REPORT

Churg–Strauss syndrome presenting with eosinophilic myocarditis: A diagnostic challenge

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Abstract Churg–Strauss syndrome (CSS) is an unusual disease that presents as systemic vasculitis and peripheral eosinophilia in patients with an atopic constitution. Cardiac involvement is unusual and often not prominent on initial presentation, but is an important cause of morbidity and mortality in patients with CSS. We report the case of a young woman with severe acute myocarditis. Coronary arteriography demonstrated extensive focal vasculopathy, consistent with coronary vasculitis, and myocardial biopsy showed eosinophilic myocarditis. This presentation led to an initial diagnosis of CSS in this patient and appropriate therapy resulted in a spectacular remission of disease activity.

PALAVRAS-CHAVE
Síndrome de Churg–Strauss;
Vasculite coronária;
Miocardite eosinofílica;
Reversão completa do envolvimento cardíaco

Síndrome de Churg Strauss complicada com miocardite eosinofílica: um desafio diagnóstico

Resumo A Síndrome de Churg-Strauss é uma doença rara que se caracteriza por vasculite sistémica associada a eosinofilia periférica, tipicamente em doentes com constituição atópica. O envolvimento cardíaco é incomum e geralmente discreto na apresentação inicial mas constitui uma importante causa de morbilidade e mortalidade nestes doentes. Descrevemos o caso de uma mulher jovem admitida por miocardite aguda. A angiografia coronária mostrou lesões coronárias difusas sugestivas de vasculite coronária e a biópsia miocárdica mostrou a presença de miocardite eosinofílica. Esta apresentação clínica permitiu o diagnóstico inaugural de síndrome

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de Churg-Strauss nesta doente e a instituição de terapêutica adequada resultou na remissão completa da atividade da doença.

Introduction

Churg–Strauss syndrome (CSS) is a rare primary systemic vasculitis characterized by peripheral eosinophilia in patients with an atopic condition, typically with a previous history of asthma or allergy.[1] It affects both small and medium-sized blood vessels of nearly all organ systems. The pathophysiology of this syndrome can be divided into three stages: first, a prodromal stage characterized by asthma and allergic manifestations; second, eosinophilic infiltration into tissues, predominantly the lungs and myocardium; and finally, a systemic stage, associated with the development of necrotizing vasculitis.[2,3] Although cardiac involvement is unusual and often not prominent on initial presentation, subclinical myocardial abnormalities are frequent and are found in more than 50% of post-mortem examinations.[1] Symptomatic cardiomyopathy carries a poor prognosis, accounting for about 50% of deaths, and is thus the major cause of mortality in CSS.[2] It can present with eosinophilic vasculitis, myocarditis, pericarditis, pericardial effusion, fibrosis, valvular heart disease, conduction disorders, intracavitary thrombi, and cardiomyopathy.[4–6] Death is usually due to myocardial infarction, heart failure, malignant ventricular arrhythmias, and/or cardiac tamponade.[4]

We describe the case of a patient with newly diagnosed CCS presenting with pronounced eosinophilic myocarditis and extensive focal vasculopathy on coronary angiography consistent with vasculitis. After appropriate and timely pharmacologic therapeutic intervention the symptoms resolved completely, as did the coronary lesions and myocardial infiltrates.

Case report

A 22-year-old woman with a history of allergic rhinitis, asthma and repeated upper respiratory tract infections in the previous two years developed progressive asthenia, dizziness and left leg paresthesias. One month after the beginning of these non-specific symptoms she was hospitalized because of pleuritic chest pain. She had had no recent flu-like symptoms, either of the upper respiratory or gastrointestinal tract, or other symptoms suggestive of a previous infectious disease. Physical examination was unremarkable; the electrocardiogram (ECG) revealed poor anteroseptal R-wave progression and diffuse T-wave inversion (Figure 1A). Laboratory tests showed eosinophilia (3.83×10^9/l), elevated erythrocyte sedimentation rate (43 mm/h) and elevated biochemical markers of myocardial injury (peak troponin I 155.6 ng/ml). Serologic and PCR tests were negative for cardiotropic viruses, *Aspergillus*, *Toxoplasma*, *Chlamydia psittaci* and *Mycoplasma pneumonia*. Specific study for parasites was also negative.

Transthoracic echocardiography revealed a small pericardial effusion with no other abnormalities. A diagnosis of myopericarditis was assumed and anti-inflammatory therapy with ibuprofen was initiated. However, the patient's recurrent chest pain persisted, associated with dynamic ECG abnormalities (transient ST-segment elevation in inferior leads) (Figure 1B) and new wall motion abnormalities on echocardiography (inferior wall hypokinesia). Given the unfavorable clinical evolution associated with a significant rise in plasma troponin I concentration and new ECG and echocardiographic abnormalities, despite anti-inflammatory therapy, cardiac catheterization was performed on the fourth day after admission. Coronary arteriography demonstrated irregularity of the larger arteries with long diffuse stenotic lesions in the left anterior descending and right coronary arteries (Figure 2A and B, arrowed), consistent with diffuse vasculitis. A myocardial biopsy showed eosinophilic myocarditis (Figure 3A) and a concomitant nasal biopsy revealed eosinophilic necrotizing granulomatous vasculitis. Immunologic study, including ANCA antibodies, was negative, while electromyography revealed left saphenous nerve mononeuropathy.

Churg–Strauss syndrome was then diagnosed as four out of six criteria were present in this patient: (1) asthma; (2) eosinophilia; (3) mononeuropathy; (4) extravascular eosinophils.[7] Pulse intravenous (i.v.) corticosteroid

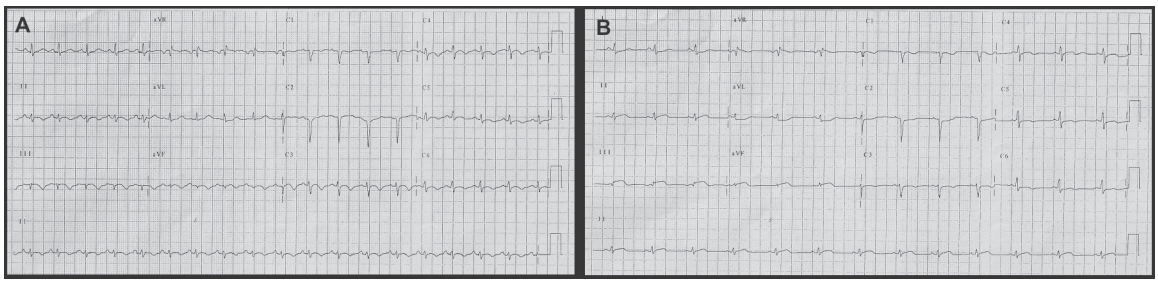

Figure 1 Electrocardiography at presentation with diffuse T-wave inversion (A) and after recurrent chest pain with transient ST-segment elevation in inferior leads (B).