

# Complete remission of coronary vasculitis in Churg–Strauss Syndrome by prednisone and cyclophosphamide

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**Abstract** The heart is involved in up to 50% of all patients with Churg–Strauss syndrome, but vasculitis of the coronary arteries has only been rarely documented. We present a young patient with severe coronary aneurysms and stenotic lesions due to a Churg–Strauss vasculitis. Prompt therapy with prednisone and cyclophosphamide resulted in the complete resolution of all lesions.

**Keywords** Churg–Strauss syndrome · Coronary vasculitis · Cyclophosphamide · Prednisone

## Introduction

The heart is involved in up to 50% of all patients with Churg–Strauss disease [1, 2]. In contrast, vasculitis of the coronary arteries has only been rarely documented [3–5]. Here, we present images of repeated coronary angiography in a young

patient with Churg–Strauss syndrome demonstrating complete resolution of severe coronary vasculitis after 1 year of therapy with prednisone and cyclophosphamide.

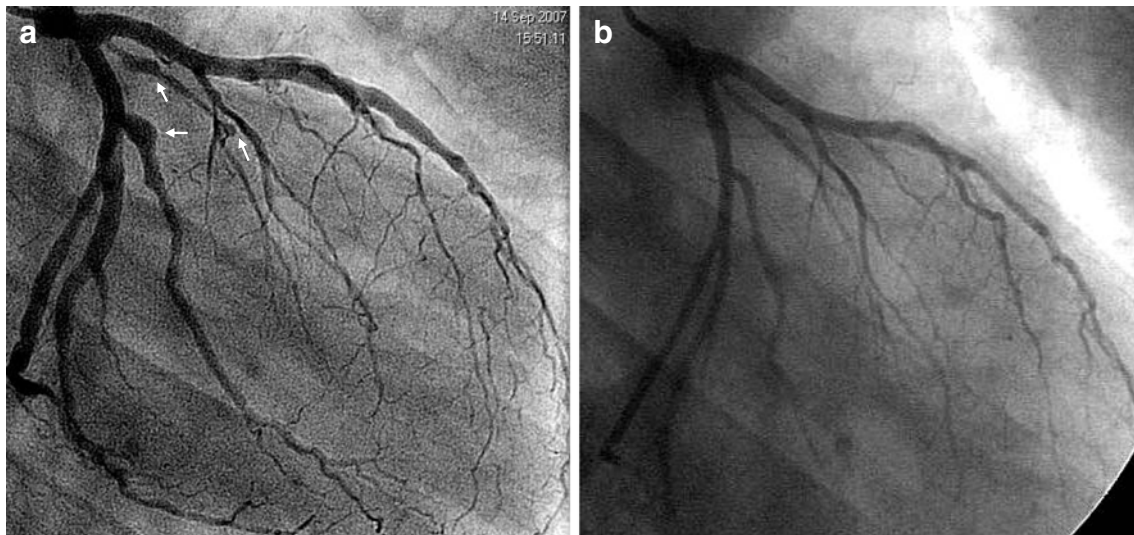
## Case report

An 18-year-old man with a history of asthma was admitted to a local hospital because of fever, profound headache, weight loss, red eyes, blurred vision, and generalized muscle aches. All symptoms resolved spontaneously within 1 month before a classifying diagnosis could be made. Five months later, the headache and blurred vision recurred, and the patient was referred to our outpatient clinic. Laboratory analysis at that moment revealed only mild eosinophilia (11%;  $600 \times 10^6/L$ ). Renal function was normal, and anti-neutrophilic cytoplasmic antibodies and anti-nuclear antibodies were not present. Cranial magnetic resonance imaging showed a unilateral thickening of the choroidal cerebral plexus, and analysis of the cerebrospinal fluid revealed eosinophilic meningitis. Another 3 months later, the patient developed a persistent fever and a pulmonary infiltrate that did not respond to antibiotics. Broncho-alveolar lavage revealed profound eosinophilia (30%). During admission, eosinophilic skin infiltrates and generalized edema developed. Now, we considered the diagnosis of Churg–Strauss syndrome, and therapy with prednisone at a dose of 1 mg/kg was initiated. Subsequently, the patient developed a painful injected pharynx and chest pain, with diffuse ST segment elevation on electrocardiography. Echocardiography revealed a mild pericardial effusion, without wall motion abnormalities. Intravenous gamma-globulin (400 mg/kg per day for 5 days) was now administered. As the chest pain recurred and there was a rise in the plasma troponin-I concentration, a coronary

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**Fig. 1** Left coronary angiogram in right inferior oblique view before (a) and after (b) 1 year of immunosuppressive therapy with prednisone and cyclophosphamide. Before the treatment, there were two prominent aneurysms in the intermediate branch and one large

aneurysm proximally in the obtuse marginal branch (a, indicated by white arrows) in addition to diffuse wall irregularities in all branches. After the treatment, all these lesions have disappeared completely (b)

angiography was performed. This showed multiple aneurysms and stenotic lesions in all coronary arteries (Fig. 1a). MRI scanning excluded involvement of the aorta and its major branches. In view of the potentially fatal coronary lesions, a more powerful immune-suppressive regimen consisting of methylprednisolone and cyclophosphamide (750 mg and 1,250 mg i.v. per day during 3 days, followed by oral doses of 1 mg/kg/day prednisone, and 2 mg/kg/day of cyclophosphamide) was chosen. The prednisone was tapered gradually and stopped after 1 year. The cyclophosphamide was tapered and stopped after 16 months. A control coronary angiography 1 year after initiation of therapy showed a complete regression of all lesions (Fig. 1b).

Our patient fulfilled the classification criteria of the Churg–Strauss syndrome as established by the American College of Rheumatology (asthma, eosinophilia, pulmonary infiltrates, and a biopsy containing accumulation of eosinophils), although he also had signs and symptoms compatible with other vasculitis syndromes, such as Kawasaki disease (bilateral conjunctival injection, injected pharynx).

Endocardial thickening due to local infiltration of eosinophils, which can trigger intracardial thrombus formation, is the most common presentation of heart involvement in patients with the Churg–Strauss syndrome [1, 2]. Pericardial involvement, presenting either as an acute pericarditis with pericardial effusion, as in our patient, or as chronic constrictive pericarditis, has been reported in 8–32% of patients [1]. In contrast, clinically significant

vasculitis of the coronary arteries has been reported only anecdotally [3–5]. In general, symptomatic cardiac involvement is associated with poor prognosis in patients with medium or small vessel vasculitis and justifies a strong and immediate immunosuppressive treatment. Indeed, the present case report shows that therapy with prednisone and cyclophosphamide may lead to a complete and uneventful recovery of potentially fatal coronary artery lesions.

**Disclosures** None

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