When this treatment fails, dapsone and retinoids might be effective. Immunosuppressive agents and corticosteroids may be required to manage severe (systemic) disease activity. Prednisolone or pulse methylprednisolone are useful in treating relapses. Often, continued treatment with oral prednisolone is necessary to control cutaneous symptoms. Azathioprine, methotrexate, cyclophosphamide, mycophenolate mofetil and thalidomide can be effective in treating chronic cutaneous lupus erythematosus [2–4]. However, randomized controlled trials are hardly available to prove or compare the effectiveness of different treatment modalities for lupus skin disease.

Various combinations of current therapeutic options had been used in the two patients described. They were either unsuccessful in ameliorating disease, or induced adverse events. Therefore, rituximab was administered.

Rituximab is a chimeric monoclonal anti-CD20 antibody that depletes mature B-cells for a median 6 months, sparing progenitor B-cells and plasma cells. Initially used in treating lymphoma, rituximab has been successfully applied in several autoimmune diseases [5], among which pemphigus vulgaris resistant to conventional immunosuppressives [6, 7]. It induced long-term B-cell depletion (6–12 months) with an even longer period of remission in the few patients studied. It has also shown to be effective and safe in treating severe therapy-resistant SLE [8, 9], with the possibility of maintenance therapy [10]. Treated SLE patients mostly suffered from severe renal, haematological or central nervous system disease. Here, we described successful treatment of two patients with predominant lupus skin disease, refractory to other therapy, suggesting that rituximab might be an alternative for the treatment of therapy-resistant lupus skin disease.

Key messages

- Rituximab is an alternative treatment for refractory lupus skin disease.

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Fig. 1. SLE skin lesions before starting rituximab, on 60 mg prednisolone daily (left). Recovery from skin lesions 6 weeks after rituximab, on 12½ mg of prednisolone daily (right).

Wegener’s granulomatosis presenting with an acute ST-elevation myocardial infarct (STEMI)

SIR, A 35-year-old man of Indian origin presented to the casualty department with a 3 week history of progressively worsening left-sided chest pain. Cardiac risk factors included a 12 yr history of
type 1 diabetes mellitus, high cholesterol, a family history of cerebrovascular disease and a 9 pack-year smoking history. Four months prior to admission he had suffered a similar episode of chest pain. Exercise tolerance test at that time was reported as normal, and routine blood tests showed only a mild neutrophilia.

On admission, an ECG showed ST elevation of over 2 mm in the inferior leads and reciprocal changes laterally, consistent with acute myocardial infarction (MI). Urgent percutaneous coronary angiography revealed a distal occlusion of the left anterior descending (LAD) coronary artery, which wrapped around the cardiac apex, and multiple small vessel occlusions, suggestive of a thromboembolic event or vasculitis (Fig. 1A and B). There was also minimal non-flow limiting atheroma. The patient underwent primary stent insertion to the distal LAD artery (Fig. 1C) with immediate resolution of his pain and ECG changes.

Following the procedure he gave a history of sore throat, neck pain, back pain and 10 kg weight loss over the preceding 4 weeks. Examination was unremarkable apart from mild hypertension of 157/74. Blood tests showed haemoglobin 10.7 g/dl, MCV 79.6 fl, platelets 503 x 10^9/l, white cell count 13.0 x 10^9/l (neutrophilia), creatinine 89 μmol/l, CRP 141 mg/l and ESR 111 mm/h.

Over the next 11 days he developed a spiking temperature of >38°C, widespread joint and muscle pains, wrist synovitis, a cough productive of blood-stained sputum and aphthous ulceration of the tongue. An echocardiogram showed apical akinesia and diastolic dysfunction but normal valves and pericardium. Urinanalysis showed 3+ protein, 3+ blood and normal microscopy. Further blood tests showed a positive cANCA with anti-proteinase 3 antibodies of 83 (normal range <20). RF was present with a titre of 103 IU/ml (normal range <40) and ANA was present at a titre of 1:360 (homogeneous pattern). Antibodies to extractable nuclear antigen, double-stranded DNA and cardioplin were not detected and lupus inhibitor was negative. Serum complement C3 fraction level was raised and C4 was normal. Immunoglobulins G and A levels were raised. His haemoglobin level fell to 8.8 g/dl and his creatinine level climbed to 144 μmol/l. Blood and urine cultures did not grow any organisms. Initial throat swabs were sterile but throat swabs and sputum cultured a week later grew both *Staphylococcus aureus* and β-haemolytic streptococcus. Anti-streptolysin titre was positive. A chest X-ray showed no significant abnormalities.

On the basis of the oropharyngeal symptoms, microscopic haematuria, renal failure and a positive cANCA, a diagnosis of...
Wegener’s granulomatosis (WG) was made. The patient was started on antibiotics and transferred to a renal unit where he was treated with plasma exchange followed by prednisolone and cyclophosphamide. He made a good recovery and returned to work 3 months later.

Cardiac involvement in WG was first described by Wegener in 1936 [1]. Abnormalities reported in this and subsequent case studies include myocarditis, pericarditis, valvular defects, vegetations, conduction defects and coronary arteritis, caused by either granulomatous infiltration or vasculitis [1–3]. The incidence of cardiac disease in WG is unclear. In one of the largest studies, where 158 patients with WG were followed up for an average of 8 years, 6% had disease affecting the heart, with only a third of these having involvement of the coronary arteries [3]. However, there are several reports of patients developing ‘silent’ MIs, often only identified following post mortem or during routine investigations [2, 4–6]. This calculation of the incidence of coronary arteritis could, therefore, be an underestimate.

Surprisingly, there are only three reports of patients with WG developing MIs with symptoms of chest pain [7–9]. In all three cases, the MI occurred early in the course of the disease and approximately a week after the diagnosis of WG being made. One patient died from the MI before immunosuppressive treatment could be started [7]. Post mortem examination revealed all three arteries to be thrombosed at the distal segments. With the other two cases, the MI occurred after commencing immunosuppressive treatment [8, 9]. Both patients underwent coronary angiography and were both found to have significant stenotic lesions in the LAD artery requiring stenting. They both made a good recovery. There has also been a case report of a patient with WG developing pulmonary oedema with ST changes consistent with an MI, but who did not experience chest pain [10]. The MI also occurred early in the course of the disease, soon after diagnosis and once steroid therapy had been started. He died shortly after. Post mortem examination reported diffuse coronary arteritis with arterial occlusion, although did not specify which artery.

Our case is unique in that this is the first reported case of a patient presenting with a symptomatic MI as the initial presenting symptom of their WG. Wegener’s granulomatosis classically presents with granulomatous disease of the respiratory tract (localized WG) followed by inflammation of the small- to medium-sized blood vessels (generalized WG) [11]. Interestingly, in our case, the coronary angiogram showed that our patient had developed a coronary vasculitis, whilst having only mild upper respiratory tract symptoms and few of the constitutional symptoms typical of generalized WG. It is possible that the pre-existing atherosclerosis in our patient led to symptoms of myocardial ischaemia in the early stages of the vasculitis. However, it is noted that of the eight previous reports of MIs in patients with WG, several also appear to have occurred during an early stage of the disease, suggesting that this is the period in which patients with WG may be most susceptible to coronary arteritis [4–10].

This is only the third coronary angiogram reported in a patient with active WG. Like the other two angiograms [8, 9], this showed no abnormalities diagnostic of vasculitis, as is often seen in other systemic vasculitides such as polyarteritis nodosa, but did show an unusual distribution of occluded small- and medium-sized vessels, particularly at the cardiac apex. Involvement of the distal segments was also seen in one of the other previously reported angiograms [9] and has also been seen during post-mortem examination [7]. These observations would seem to reflect the fact that WG is a vasculitis that predominantly affects the smaller blood vessels first [11], with the distal branches possibly being more likely to be affected in WG because of their smaller diameter. It is also noted that all three angiograms identified stenoses in the LAD artery rather than the other main branches but we cannot provide an explanation for this.

In conclusion, we report a unique case of a young man with multiple cardiac risk factors, who presented with an apparently straightforward acute inferior MI who subsequently developed symptoms, signs and serological evidence of WG. We have observed that coronary arteritis may occur early in WG and suggest careful cardiac screening of all patients with newly diagnosed WG.

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Severe infections following leflunomide therapy for Rheumatoid Arthritis

SIR, We read with great interest the report by Hocevar et al. [1] about the association of pulmonary tuberculosis with leflunomide therapy in rheumatoid arthritis (RA). We face a similar problem in our rheumatology practice. The average prevalence of all forms of tuberculosis in India is estimated to be 5.05 per thousand [2], so it is not surprising that the commonest infectious complication of disease-modifying anti-rheumatic drug (DMARD) therapy that we see is tuberculosis (TB). We find most cases of TB in patients