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Treatment of Wegener's granulomatosis with cyclosporin

SIR, Cyclosporin has been used successfully as an immunosuppressive drug in allografted patients. In addition, data have been published on the beneficial influence of cyclosporin in autoimmune diseases such as uveitis,¹ myasthenia gravis,² and polymyositis.³ Furthermore, there is evidence for the effectiveness of cyclosporin in patients with diabetes mellitus type I.⁴

In this report we present a patient with Wegener's granulomatosis in whom conventional therapy with cyclophosphamide and prednisone failed. With the administration of cyclosporin, however, remission of the disease was achieved.

In January 1986 a 61 year old man was admitted to our hospital for evaluation of progressive respiratory distress, cough, and severe sinusitis with rhinorrhoea. Eleven months earlier the diagnosis of Wegener's granulomatosis had been made elsewhere on the basis of clinical symptoms and characteristic histopathological findings of an extensive destructive granulomatous reaction with necrotising vasculitis of the upper and lower respiratory tracts. Consequently, treatment with cyclophosphamide (150 mg/day) and prednisone (40 mg/day) was started. At that time no renal involvement was found. Although initially this treatment appeared to be successful, his complaints about dyspnoea and pain behind his right eye were progressive in November 1985. A computed tomographic (CT) scan showed severe destruction of the right concha inferior and sinus maxillaris and part of the right orbita. For this reason the doses of cyclophosphamide and prednisone were increased to 200 mg and 100 mg a day respectively. Nevertheless, his condition deteriorated further, which made admission to hospital necessary in January 1986. On admission he was very dyspnoeic at rest,

with severe rhinorrhoea and nasal discomfort. Laboratory studies, including erythrocyte sedimentation rate, C reactive protein, liver and kidney function, were normal. There were no signs of infection. An x ray examination of the chest showed the presence of bilateral fibrosis without hilar adenopathy. Examination of pulmonary function showed obstructive abnormalities. Review of the nasopharynx and lung biopsy specimens taken earlier confirmed the diagnosis of Wegener's granulomatosis. Since the disease appeared to be progressive despite optimal therapy we decided—after informed consent of the patient—to start treatment with cyclosporin (10 mg/kg body weight/day). At the same time cyclophosphamide was stopped and prednisone tapered off slowly. He has now been receiving cyclosporin and a low dose of prednisone for nine months and is doing very well. Rhinorrhoea and nasal discomfort have disappeared completely and dyspnoea becomes manifest only on major physical exercise; he is even able to cycle without difficulty. A control CT scan showed that the process of destruction of the upper airway organs and orbita has been stopped. Trough levels of cyclosporin (using the high performance liquid chromatography method) range from 140 ng/ml to 180 ng/ml, measured in whole blood. Serum creatinine has been increased slightly to 130–150 µmol/l (normal <120 µmol/l) during treatment.

In this case cyclosporin appears to have been a very effective drug in the treatment of Wegener's granulomatosis. We suggest, therefore, that this drug be considered as an alternative to cyclophosphamide in patients who are resistant to conventional therapy or suffer severe side effects.

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