Membranoproliferative Glomerulonephritis Associated with Buckley's Syndrome, Treated with Cyclosporin

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Dear Sir,

In 1982, we published in Nephron a case of Buckley's syndrome associated with membranoproliferative glomerulonephritis [3]. Now, we report the results of cyclosporin treatment in this patient.

Buckley's syndrome (hyper-IgE syndrome) is a rare every 2 weeks, congenital immune defect associated with recurrent infections and severe atopic eczema. It is characterized by a permanent increase of serum IgE levels and abnormal granulocyte chemotaxis; there is no treatment other than symptomatic [1].

Because cyclosporin can reduce hyper-IgE levels in Brown-Norway rats with experimental chloride mercury nephritis [2], we used it in our patient. Classical features of Buckley's syndrome were present since childhood, with long-lasting generalized atopic eczema, recurrent focal infections especially sinusitis and IgE levels permanently above 10,000 KIU/l . Proteinuria and microscopic hematuria appeared at age 17, in 1964; in 1966 renal biopsy revealed MPGN type I. During follow-up bouts of infected sinusitis, generalized eczema and enhancement of proteinuria occurred simultaneously. This patient was treated with nonsteroidal anti-inflammatory drugs (NSAID) from 1967 to 1984, with a reduction of proteinuria; nevertheless hypertension and a slow increase of creatinine serum levels were noted in 1982 leading to a stop of NSAIDs. Then he received ketotifen with weak improvement of the eczematous lesions. In April 1985, creatinine levels were $250 \mu\text{mol/l}$ and proteinuria 5 g/24 h. Blood pressure was controlled by a beta-

blocking drug (atenolol 200 mg/day). Without modifying other treatments, oral cyclosporin was given 400 mg/day, once in the morning (5 mg/kg/day). A dramatical improvement was noted within 3 days: itching, which had been present night and day for more than 20 years, totally disappeared and eczematous lesions cleared progressively within 2 weeks. Cyclosporin plasma levels (measured every 2 weeks, 24 h after the precedent intake), fluctuated from 30 to 100 ng/ml and total blood levels from 130 to 270 ng/ml. After 3 months of this treatment we tried to reduce the dose to 300 mg/day; within 24 h, itching recurred and cutaneous lesions progressively reappeared. With the initial dosage, itching and eczema disappeared again. We are now trying to find the minimal effective dose (March 1986).

During the administration of cyclosporin, the following observations were made: (1) a sensation of well-being was spontaneously noted by the patient; (2) a decrease of serum IgE levels from more than 10,000 KIU/l. This effect was also dose-dependent, for an IgE increase was observed during the period of dosage reduction; (3) creatinine levels fluctuated from 250 to 300 $\mu\text{mol/l}$, and proteinuria slowly decreased to less than 0.50 g/day; (4) although this patient already had definite renal failure, no sign of nephrotoxicity was observed: urinary beta-2-microglobulin and enzymuria remained normal. No other side effect was observed after 10 months of treatment. Thus, during cyclosporin therapy we observed simultaneously the reduction of IgE levels, the cure of eczematous lesions and a decrease in proteinuria, with stabilization of renal function.

Membranoproliferative Glomerulonephritis Associated with Buckley's Syndrome, Treated with Cyclosporin

383

tion of the renal function. But it appears probable that this treatment must be carried on on a long-term basis with all the risks of the side effects of this drug.

The mechanism of action of cyclosporin in such a case remains hypothetical, as are the links between Buckley's syndrome and the MPGN.

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