Idiopathic Focal Segmental Glomerulosclerosis Resistant to Ciclosporin-A Therapy

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Dear Sir,

There are conflicting reports about the efficacy of ciclosporin-A (CiA) in steroid-resistant idiopathic focal segmental glomerulosclerosis (FSGS) [1–3]. Therefore we tried CiA in 2 children who had steroid-resistant FSGS. CiA was used at a dosage of 5 mg/kg body weight/day in two divided doses for a period of 3 months.

Case Reports

Case 1. Master S, a 9-year-old boy, presented in 1985 with nephrotic syndrome and normal renal function. There was no evidence of systemic disease. Serum complement profile was normal. Renal biopsy showed FSGS. Immunofluorescence study was negative for IgG, IgA, IgM and C3. He was diagnosed as a case of idiopathic FSGS and was initially treated from 1985 with an alternate day steroid schedule [4] with no response. Prednisolone was withdrawn and he was started on CiA in February 1987 at the above-mentioned dosage. In March 1987, he developed spontaneous pneumococcal peritonitis which was treated successfully. His serum albumin and 24-hour urinary protein excretion at the beginning of therapy were 1.9 g/dl and 2.0 g, respectively, and at the end of therapy they were 1.7 g/dl and 2.1 g, respectively. As there was no response to CiA therapy, it was discontinued at the end of 3 months. CiA toxicity was not encountered.

Case 2. Miss S., an 8-year-old girl, presented with nephrotic syndrome in December 1986 and had normal renal function. There was no evidence of systemic disease and serum complement profile was normal. Renal biopsy showed FSGS. Immunofluorescence study showed no IgG, IgA, IgM and C3 deposits. She was diagnosed to have idiopathic FSGS and was resistant to steroid therapy. Prednisolone was stopped and she was started on CiA at the above-mentioned dosage in February 1987. Her serum albumin and 24-hour urinary protein excretion at the beginning of therapy were 2 g/dl and 1.5 g, respectively, and at the end of therapy they were 1.9 g/dl and 6.4 g, respectively. CiA toxicity was not seen. Due to absence of response, CiA was discontinued at the end of 3 months.

In both these cases of steroid-resistant idiopathic FSGS in children, there was no response to CiA therapy. This is in contrast to the partial remission seen in idiopathic FSGS treated with CiA as

reported [1]. Our findings of persistent nephrotic state with CiA therapy are consistent with other reported series [2, 3].

References

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