LETTER

Child onset steroid-dependent nephrotic syndrome with a combination of minimal glomerular changes and IgA deposits: long-term follow-up

Dear Editor,

Child onset minimal change nephrotic syndrome (NS) associated with mesangial IgA deposition is reported as a variant of minimal change disease[1], or an overlap of minimal change disease and IgA nephropathy[2]. The long-term outcome of patients with such histological findings remains unclear.

We report the case of a boy who presented with NS at the age of 1.8 years. During 10-year monitoring, he experienced nine relapses, on high alternate doses of prednisolone. After the 3rd relapse, cyclophosphamide was administered at a dose of 2 mg/Kg/d for eight weeks. Fourth and 5th relapses occurred three and six months after cyclophosphamide treatment, and thus, cyclosporine was initiated at a dose of 5 mg/Kg/d. A 6th relapse occurred after 1.3 years on cyclosporine due to low levels. After three years in remission, a gradual decrease of cyclosporine was attempted resulting in a 7th relapse again due to low levels of cyclosporine. During the following two years, he had two further relapses (8th and 9th), with therapeutic cyclosporine levels and with no evidence of infection. Renal biopsy at 11.5 years of age, after long-term treatment with cyclosporine, showed minimal glomerular changes and mild IgA deposition. The patient never developed hematuria and hypertension, and maintained normal renal function throughout the monitoring period.

Similar to our case, the coexistence of minimal change NS with mesangial IgA deposition has been considered as a marker of an easily relapsing course[1,2] .

The corticosteroid-sparing agents used in our patient were in agreement with those suggested by the recent KDIGO guidelines[3] for children with steroid dependent NS, where cyclophosphamide or cyclosporine are recommended to maintain remission. Although randomized control trials have demonstrated no significant differences in the risk of relapse between cyclosporine during treatment and cyclophosphamide; cyclophosphamide is considered less effective in steroid dependent NS[3], as was the case with our patient. Despite the fact that immunosuppressive agents other than steroids are recommended in steroid dependent NS, their benefit in IgA nephropathy is limited, unless there is crescentic IgA nephropathy with deteriorating renal function[4].

A limitation in the present case is that renal biopsy was performed long after the onset of NS and thus it is not known whether the coexistence of minimal glomerular changes and IgA deposition was present from the onset or if it appeared during the course of the disease.

In conclusion, based on the overall clinical course and the long-term monitoring of this patient, it can be speculated that the coexistence of minimal glomerular changes with IgA deposition could be a risk factor for steroid dependent NS, without manifestations of IgA nephropathy. Further case studies with long-term monitoring are necessary to reach definitive conclusions.

References

1. Gallego N, Gonzalo A, Mampaso F, Ortuño J. Steroid-dependent nephrotic syndrome with minimal glomerular changes and mesangial IgA deposits. Child Nephrol Urol. 1988-1989; 9: 98-100.
2. Barbiano di Belgiojoso G, Mazzucco G, Casanova S, Radaelli L, Monga G, Minetti L. Steroid-sensitive nephrotic syndrome with mesangial IgA deposits: a separate entity? Observation of two cases. Am J Nephrol. 1986; 6: 141-145.
3. Chapter 3: Steroid-sensitive nephrotic syndrome in children. Kidney Int Suppl (2011). 2012; 2: 163-171.
4. Chapter 10: Immunoglobulin A nephropathy. Kidney Int Suppl (2011). 2012; 2: 209-217.

Conflict of interest

None declared.

Keywords: Minimal change disease, IgA deposition, child

Siomou E[1], Pavlou M[1], Kalaitzidis R[2]

[1]Department of Pediatrics
[2]Department of Nephrology
University Hospital of Ioannina, Ioannina, Greece

Corresponding author: Siomou E. MD, University Hospital of Ioannina, Stavros Niarchos avenue, 45500 Ioannina, Greece, tel: +302651099793, fax: +302651097080, e-mail: eksiomou@yahoo.gr