Short Communication

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Mitochondrial Complex I Deficiency among Egyptian Pediatric Patients with Steroid-Resistant Nephrotic Syndrome

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Abstract

Background and objectives: Nephrotic Syndrome (NS) is one of the most common glomerular diseases among children. Up to 20% of patients are steroid resistant (SRNS) representing a challenging subset at high risk of developing end-stage renal disease. Renal manifestations of mitochondrial diseases (MIDs) include nephrotic syndrome, renal insufficiency, nephrolithiasis, Bartter-like syndrome, focal segmental glomerulosclerosis, and nephrocalcinosis. The objective of the current study is to measure the activity of mitochondrial complex I in renal biopsies obtained from pediatric patients diagnosed with SRNS compared to steroid-sensitive nephrotic syndrome (SSNS) patients in order to elucidate its role in pathogenesis and the prognosis for further genetic work.

Subjects and Method: Renal biopsies of 120 patients diagnosed with nephrotic syndrome based on clinical and laboratory findings, divided into two groups, SRNS (60 patients) and SSNS (60 patients). Pathological study and spectrophotometric measurement of mitochondrial complex I in renal biopsy and muscle homogenates were performed for both groups.

Results: Positive consanguinity was a remarkable finding in 44 patients among the SRNS group (73%), compared with 33 patients among the SSNS group (55%). Complex I activity was significantly lower in the SRNS group (0.2657 ± 0.1831 nmol/ml/min), than in the SSNS group (0.4773 ± 0.1290 nmol/ml/min) (). There was a significant positive correlation between complex I activity and the heaviness of proteinuria among the SRNS group (r 0.344,). There were statistically significant differences in serum C3 and C4 levels between both groups (, 0.053, respectively).

Conclusion: Mitochondrial complex I deficiency in patients who have a Nephrotic syndrome complaint may play a role in their responsiveness to steroid therapy and the development of SRNS and even the prognosis of their illness.

Biography

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