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A case of SLE and ANCA associated vasculitis overlap syndrome

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Background: Renal involvement of ANCA associated vasculitis (AAV) is typically a pauci-immune crescentic glomerulonephritis(GN) which is a rare condition in children and clinically characterized by features of rapidly progressive GN with rapid loss of renal function. ANCA are found in up to 20% of Systemic lupus erythematosus (SLE) and some patients fulfilling both SLE and AAV classification criteria were recently defined as having SLE/AAV overlap syndrome. Here we present a case of SLE/AAV overlap syndrome.

Case: A 16 yr-old girl with no past history was referred to our hospital for the evaluation of proteinuria and hematuria which were found on school screening. Her initial laboratory findings were as follows : WBC 3,930/uL, Hb 11.4g/dL, plt 149k/uL, BUN 12.3mg/dL, creatinine 0.8 mg/dL, C3 94mg/dL, C4 41.2mg/dL , albumin 3.8g/d L, Urine protein(1+) RBC >30, Urine protein/cre ratio 2.1. Initially, we suspected Ig A nephropathy because her renal biopsy demonstrated a few immune deposit on EM despite of inadequate IF specimen and some crescents on LM. We started enalapril treatment. After 2 months of treatment, She came into ER because of hemoptysis occurred suddenly. Her chest x-ray and CT scan showed pulmonary hemorrhage and her renal function deteriorated(serum creatinine 1.34 mg/dL). She began to recover with methylprednisolon pulse therapy and 7 sessions of plasmapheresis. And subsequent evaluation demonstrated positive MPO ANCA and crescentic GN with chronic change on repeated renal biopsy. Then, we began induction therapy of pauci-immune crescentic glomerulonephritis with monthly intravenous cytoxan and corticosteroid. Her renal function slowly improved, but not completely. After 2 yrs of maintenance therapy, immunosuppression was stopped and soon after RPGN relapsed without extrarenal symptoms. We suspected concomittant SLE by some autoantibodies (ANA, ds DNA Ig G), low complement and leukopenia and lymphopenia. Now she is in ESRD state in spite of re-induction therapy with rituximab.

Conclusion: SLE/AAV overlap syndrome is rare in children and presents with a severe clinical symptoms (rapidly progressive GN and frequent pulmonary involvement).

Keywords : AAV, SLE, overlap, RPGN, child