THE ROLE OF ANCA AND ANTI-GBM ANTIBODIES TITERS IN PULMONARY-RENAL SYNDROME DUE TO WEGENER'S GRANULOMATOSIS

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Pulmonary-renal syndrome (PRS) is defined as a diffuse alveolar hemorrhage (DAH) and rapidly progressive glomerulonephritis (RPGN) occurring as the presenting manifestation of underlying, multisystem autoimmune disease. It represents a medical emergency with a high mortality which necessitates rapid diagnosis and institution of therapy. We present a retrospective study of 22 consecutive patients with Wegener's Granulomatosis from 2004-2007. The mean age was 44.5 (32-67) years and 6 patient were man. Logistic regression analysis and the Wilcoxon test were included in the statistics. Survival time and death risk were assessed using the Kaplan-Meier estimator and Cox's proportional hazard model. At recognition median BVAS-WG was 30 (23.0-32.5), PO_2 on air 5.8 ±0.5 KPa, creatinine level was 7.2 ±1.4 mg/dl. 15 patients were PR3 positive, 3 were MPO positive, among them 4 patients were also positive for anti-GBM anybodies. Renal biopsy was performed in 16 patients. Histological examination reviled segmental necrotizing crescentic GN in 15 patients. 13 patients were initially dialysis dependent, and 7 required ventilatory support. All patients were treated with methylprednisolone (pulses) and cyclophospamide (pulses), 8 patients underwent plasma exchange. Patients were followed up for 24 ±8 month. Six patients died in the first month. Of the survivors, 55% and 31% were alive after 1 and 2 years of completed follow-up: 73% and 55% of these were dialyzed, respectively. High activity of illness (ANCA titers), neutropenia and infections were frequent contributors to death. Early recognition and proper, less toxic treatment may improve outcome in PRS.