CLINICAL NEPHROLOGY, PRIMARY AND SECONDARY GLOMERULONEPHRITIS - 1

RENAL AND EXTRA RENAL OUTCOMES IN ANCA NEGATIVE AND ANCA POSITIVE SMALL VESSEL VASCULITIS

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Introduction and Aims: About 10-30% patients with small vessel vasculitis (SVV) are ANCA negative. Literature about comparing their outcomes with ANCA positive patients is limited to one study from China. In this study we compared similar outcomes in a British cohort of patients.

Methods: A retrospective review of all the patients with renal involvement due to pauci-immune SVV diagnosed between 1998-2011 in a tertiary renal centre was done. Data regarding demographics, renal and patient survival, extra renal organ involvement, treatment and relapse rates were collected.

Results: 221 patients were diagnosed between 1998-2011 with pauci-immune SVV, of which 165 were ANCA positive (74.7%) and 56 were ANCA negative (25.3%). The average age at diagnosis of the ANCA positive and ANCA negative patients was 62 and 52 years respectively (p<0.001). Overall ANCA negative patients had significantly less life threatening extra renal organ involvement (p=0.004) including pulmonary (p=0.022) and eyes- ENT (p=0.001) compared with ANCA positive patients. Renal survival was significantly worse in the ANCA negative patients (p=0.003, Hazard ratio=2.00, 95% confidence interval = 1.28 to 3.14) (figure-1). Age adjusted patient survival was also significantly worse in this group (p=0.004, Hazard ratio=1.97, 95% confidence interval=1.24 to 3.12).

Data regarding treatment and relapse rates were not available for a sizeable proportion of patients but for those that it was, there does not appear to be a significant difference in plasma exchange and cumulative cyclophosphamide dose between the 2 groups.

There was a trend towards increased tendency for relapse for the period of follow up in ANCA positive patients compared with ANCA negative patients (p=0.061).

Conclusions: In this retrospective study we found that, ANCA negative patients compared to ANCA positive patients-
1) Are younger.
2) Have less frequent life threatening extra renal organ involvement.
3) Have poorer renal survival
4) Have poorer patient survival
5) Are less likely to have relapse.

These differences in the presentations and outcomes could help in managing these patients.

Our study had several limitations including:
1) Retrospective study
2) Significant data regarding treatment was not available.