The Interesting Case

ANCA-positive Churg–Strauss syndrome with renal failure

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Churg–Strauss syndrome was described in 1951 as a granulomatous variant of polyarteritis nodosa with eosinophilic tissue infiltration [1]. The clinical presentation corresponds to that of a systemic vasculitis involving one or more extrapulmonary organs with asthma and peripheral blood eosinophilia in excess of 1.5 × 10⁹/l [2].

Our 56-year-old patient was primarily treated for spastic bronchitis, and for suspected but unproven tuberculous lymphadenopathy. He was referred because of dialysis-dependent renal failure. He suffered from intermittent fever, progressive spastic dyspnoea and malnutrition. Laboratory examination disclosed marked eosinophilia (1.9 × 10⁹/l), high ESR (90 mm/h), profound anaemia (haemoglobin 83 g/l), and positive p-ANCA (1:160 by immunofluorescence).

Churg–Strauss syndrome was suspected and confirmed by renal biopsy. Of the 18 glomeruli in the specimen 16 were encircled by large crescents in different stages. Granulomatous periglomerulitis was also noted. One preglomerular arteriole was affected by florid necrotizing vasculitis with a mild granulomatous reaction (Figures 1, 2). The tubules were largely destroyed. Interstitial oedema was noted with diffuse cellular infiltration, consisting mainly of eosinophils and plasma cells (Figure 3). Immunofluorescence showed only mild segmental positivity for IgG in two glomeruli with C3 deposition in Bowman’s capsule; the remaining eight glomeruli were completely sclerotic.

The patient was treated with a combination of high-dose methylprednisolone (with subsequent oral prednisone) and continuous cyclophosphamide (2 mg/kg per os) together with five plasma exchanges. After almost 2 months of treatment renal function recovered partially (PCr below 450 μmol/l). His FEV1 remained low (45%), and repeated bouts of asthma occurred.
gastric ulcer which had developed despite prophylactic treatment with H2 blockers.

Glomerulosclerosis, progressive interstitial fibrosis, and vascular sclerosis with considerable narrowing of the vascular lumen were found at postmortem. There were no signs of active disease in the kidney (Figure 4). Patchy, small vascular necroses were occasionally found in the paratracheal lymph nodes (Figure 5). Renal failure is thought to be rare in the Churg–Strauss syndrome, but this case illustrates that it may occasionally occur in cases with late referral and particularly high disease activity.

References


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