**Case Report**

**Simultaneous IgA nephropathy and Wegener’s granulomatosis – overlap or coincidence (the role of renal biopsy)**

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**Introduction**

Extracapillary and pauci-immune necrotizing glomerulonephritis is a prominent feature of renal involvement in patients with Wegener’s granulomatosis (WG). However, typical pulmonary, ocular or ear–nose–throat (ENT) lesions may precede renal involvement and establish the diagnosis of the disease. The role of renal biopsy in the management of such patients is still debated.

**Case**

A 32-year-old man presented with episcleritis and scleritis associated with the subsequent development of diffuse arthralgia, maxillary sinusitis and laryngotracheitis. The previous medical history was unremarkable except for diagnosis of relapsing otitis media 7 years earlier. He was a non-smoking, non drinking white-collar worker. Blood pressure was 125/80 mmHg; he was afebrile. Oropharynx, tracheal laryngeal and bronchial endoscopy showed severe subglottic and tracheal inflammation with histological evidence of necrotizing angiitis. White-cell count and C-reactive protein were increased at 14 800/mm³ and 18 mg/dl, respectively and plasma antineutrophil cytoplasmic antibodies (ANCA) positivity (1/50) with anti-proteinase 3 specificity (32 U/l) (anti-myeloperoxidase negative) was seen. Antinuclear antibodies, anti-type II collagen antibodies and anti-cardiolipine antibodies were negative. Complement was normal. Plasma creatinine and urea were 70 µM and 6 mM, respectively. Urine analysis revealed discrete proteinuria (0.8 g/d) and microscopic haematuria (80 000/mm³). Thoracic tomodensitometric study revealed two small excavated nodular lesions of the right upper lobe. Renal sonography and intravenous urography showed no urinary lithiasis and no ureteral involvement. Light-microscopy study of renal biopsy showed discrete mesangial hyperplasia and no necrotizing angiitis, extracapillary proliferation or tubulointerstitial lesions. Immunofluorescence analysis revealed diffuse mesangial IgA deposits. He was treated with oral corticotherapy and monthly pulsed cyclophosphamide. Nine months later, he still had active pulmonary lesions but remained free of renal disease.

**Discussion**

The association of IgA nephropathy and Wegener’s granulomatosis has rarely been documented in the literature. This association may be fortuitous and reflect the high incidence of IgA nephropathy in the general population. Alternatively, it may suggest common pathophysiological mechanisms in these two diseases. Indeed, Andrassy et al. [1] reported the occurrence of de novo IgA glomerulonephritis in three patients with previously documented WG and biopsy-proven pauci-immune crescentic glomerulonephritis. In all cases de novo mesangial IgA deposits occurred while patients were in remission, with no signs of systemic disease or positive ANCA titres, and in two cases they were associated with the reappearance of a nephritic sediment. In a fourth patient with rapidly progressive glomerulonephritis, positive c-ANCA titres and a 3-month history of afebrile sinusitis with no signs of vasculitis, histological examination of renal tissue revealed mesangial enlargement and diffuse interstitial fibrosis with mesangial and peripherical deposits of IgA, C3c and C3d [1]. Cases of IgA nephropathy with rapidly progressive glomerulonephritis and positive anti-myeloperoxidase antibodies and/or p-ANCA have also been reported in the literature [2]. Whether this represents a true continuum between IgA nephropathy and small-vessel vasculitis remains to be elucidated.

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Abnormal urinary sediment is one of the four cardinal American College of Rheumatology criteria for WG [3]. Ureteral vasculitis is an uncommon cause of haematuria in the course of WG, and normal ureterography made it an unlikely cause in our patient [4]. However, this observation clearly emphasizes the need for a thorough evaluation of a patient with suspected WG and haematuria, and namely for the histopathological examination of kidney tissue. A recent prospective study demonstrated that renal biopsy altered the management in 32% of patients with isolated haematuria and proteinuria [5]. Cyclophosphamide is the cornerstone therapy of systemic WG; however, limited forms of WG with preferential involvement of ENT have been individualized [6]. The lack of progression of the disease in some of these patients has raised the question of a less aggressive therapy [7]. Exclusion of angiitis-related haematuria relies essentially on renal biopsy and may be critical in the care of these patients.

Recent studies have highlighted the high specificity of c-ANCA and anti-proteinase 3 antibodies in the diagnosis of WG in patients with proteinuria and haematuria [8]. However, the specificity of c-ANCA is not absolute. Davenport [9] reported two patients in whom positivity of c-ANCA in the context of systemic disease and microscopic haematuria and/or proteinuria led to misdiagnosis of WG and was eventually associated with tuberculosis and non-Hodgkin’s lymphoma. Finally, in a prospective study of c-ANCA positivity and clinical criteria in diagnosing WG in patients with suspected vasculitis, up to 50% of patients with positive c-ANCA did not have WG [10]. Our observation further demonstrates that the diagnosis of non-angiitis-related haematuria should not be overlooked in patients with WG.

References


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