Idiopathic Rapidly Progressive (Crescentic) Glomerulonephritis with Myeloperoxidase Antibodies – A New Entity Linked to Female Sex?

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Dear Sir,

Rapidly progressive (crescentic) glomerulonephritis (RPGN) is characterized by an acute nephritic syndrome with development of renal failure within a short period of time and pathohistologically by crescentic glomerulonephritis (GN). Crescentic GN may occur as idiopathic GN but is frequently associated with multisystem disorders such as systemic vasculitis, collagen disease, Goodpasture's syndrome and bacterial infections. Immunohistologically, three categories of RPGN can be recognized: (1) anti-GBM antibody RPGN, (2) RPGN with deposition of immune complexes and (3) RPGN without relevant immune deposits. More than 80% of cases with RPGN belong to groups 2 and 3 [1]. Since the introduction of the antineutrophil cytoplasm antibody (ANCA) test for screening of Wegener's granulomatosis (WG) and related vasculitic disorders, the serological identification of idiopathic RPGN without immune deposits has been considerably improved [2, 3]. As referral center for WG we have identified a subgroup of patients with crescentic glomerulonephritis that differs from WG on clinical, serological and pathohistological grounds. This subgroup (MP) of patients (n = 23) presented to our unit with features of systemic vasculitis. Five patients had only renal disease, 10% disclosed a pulmonary-renal syndrome, and 33% a multiorgan involvement resembling WG or microscopic polyarteritis. This subgroup of patients (n = 23) could be distinguished from our cases with biopsy-confirmed WG and renal involvement (n ≈ 31) by:

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Fig. 1 a (Peri)-nuclear staining of neutrophil cytoplasm, b Fine, diffuse granular staining of neutrophil cytoplasm.

(1) Predominant affection of females

MP: 21 females, 2 males (median age 54 years, range 25–78)

WG: 17 females, 14 males (median age 55 years, range 16–78)

(2) Presence of different antibodies against neutrophils (ANCAs)

Immunofluorescence of neutrophils

MP: perinuclear (granulocyte-specific antinuclear

antibodies) staining Q^{3/4}i) (see fig. 1a)

WG: fine diffuse cytoplasmic pattern (3^{1/2}i) (see fig. 1b)

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Elisa with the 29-kD protein or myeloperoxidase MP: 2^{3/8} patients had myeloperoxidase antibodies (median 13.2, range 2.1–66.7; normal value < 2); no patient showed 29-kD-protein antibodies WG: 3^{1/4} patients had antibodies against the 29-kD protein (median 70, range 20–200; normal value < 10); only 1 patient had additional myeloperoxidase antibodies (3) Renal

pathology

MP: 5⁰ patients with renal biopsy had crescentic GN, 2 patients mesangial proliferative GN; on immunohistology 3⁴0 had no glomerular immune deposits, 3⁴0 discrete granular deposits of IgA and 3⁴0 IgG; in addition to arteriolar involvement (arterio-litis) renal arteritis (vasculitis of interlobular and/or of larger arteries) was detected in 1²0 cases WG: 16¹7 patients showed crescentic glomerulo-nephritis, 1 patient chronic sclerosing GN; no immune deposits were detected; no patient disclosed intrarenal arteritis (affection of small- and medium-sized arteries) as observed in MP, although arterio-litis was frequently present

The incidence of MP: WG amounts to 1:6 in our unit

References

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