

## KSN 2017 Abstract

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### ANCA-ASSOCIATED VASCULITIS WITH BOTH MPO-ANCA AND PR3-ANCA SHARES CHARACTERISTICS OF ANCA-ASSOCIATED VASCULITIS WITH SINGLE ANCA

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**Objectives :** The anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are heterogeneous group of necrotizing inflammation of small vessel and the presence of the ANCA. ANCAs are defined according to the target antigens, leukocyte proteinase 3 (PR3) and myeloperoxidase (MPO). Recently, the ANCA specificity could be better for classification of ANCA-associated vasculitides than the clinical diagnosis. A few patients have both MPO- and PR3 ANCA. However, the clinical characteristics of these patients were not known in detail.

**Methods :** To analyze organ involvement of patients with ANCA-associated vasculitis according to ANCA type focusing both MPO- and PR3-ANCA (both-ANCA) positive vasculitis Methods: The medical records of the patients with positive ANCA and clinical diagnosis or the patients with positive ANCA and vasculitis diagnosis confirmed by biopsy were reviewed at two regional tertiary hospitals. The age at diagnosis, sex, and the organ involvement of kidney, lung, upper airway (nose/sinus/ear), skin, peripheral nervous system, central nervous system, and gastrointestinal tract were collected. The clinical variables were analyzed by ANCA type.

**Results :** Total 82 patients with positive ANCA and clinical diagnosis or histologic diagnosis of vasculitis were searched. MPOANCA positive patients was 63 (76.8%), PR3-ANCA 9 (11.0%), and both MPO- and PR3-ANCA was 10 (12.2%). The age at diagnosis of patients with PR3-ANCA was younger than patients with MPO-ANCA or both-ANCA (PR3-ANCA, 49.6 vs. MPOANCA, 66.1 vs. both-ANCA, 62.1,  $p < 0.05$ ). Moreover, kidney involvement were MPO-ANCA was 77.8%, PR3-ANCA 22.2%, and both-ANCA 80% ( $p < 0.05$ ). Upper airway involvement was also significantly associated with ANCA type (PR3-ANCA, 66.7% vs. MPO-ANCA, 23.8% vs. both-ANCA, 50.0%,  $p < 0.05$ ). The involvement of skin, central or peripheral nervous system, gastrointestinal tract or the presence of lung fibrosis and lung nodule or mass did not differ

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according to ANCA type.

**Conclusions** : ANCA-associated vasculitis with both MPO-ANCA and PR3-ANCA has more kidney involvement than ANCA associated vasculitis with PR3-ANCA and more upper airway involvement than ANCA-associated vasculitis with MPO-ANCA.

**Keywords** : Anti-Neutrophil Cytoplasmic Antibody-Associated Vasculitis; Granulomatosis with Polyangiitis; Microscopic Polyangiitis