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Cutaneous Manifestations of ANCA-Associated Vasculitis

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SESSION INFORMATION

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Session Title: Vasculitis - Poster II: ANCA-Associated Vasculitis

Session Time: 9:00AM-11:00AM

Background/Purpose: The cutaneous manifestations of anti-neutrophil cytoplasmic antibody-associated vasculitis (AAV), including granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA), and eosinophilic granulomatosis with polyangiitis (EGPA), are varied and have not been well characterized. This study aimed to describe the spectrum and extent of dermatologic features of AAV.

Methods: A large, cross-sectional study describing, comparing, and contrasting the cutaneous manifestations of GPA, MPA, and EGPA was performed using data from the Diagnostic and Classification Criteria in Vasculitis (DCVAS) study. The DCVAS study is a large, international, collaborative effort to collect comprehensive clinical data on a large cohort of patients with vasculitis with a goal of developing new classification and diagnostic criteria.

Results: Data from 1274 patients with AAV from 130 centers worldwide were available for this study: 702 (55%) with GPA, 331 (26%) with MPA, and 241 (19%) with EGPA. Cutaneous findings were common in patients with AAV (Table 1), affecting 239 (34%) patients with GPA, 97 (29%) patients with MPA, and 113 (47%) patients with EGPA. The most frequent cutaneous manifestations in each type of AAV were as follows: GPA: petechiae or purpura (N=113; 16%), painful skin lesions of any type (N=66; 9.4%), and maculopapular rash (N=47; 6.7%); MPA: petechiae or purpura (N=33; 10%), livedo reticularis or racemosa (N=25; 7.6%), and maculopapular rash (N=20; 6.0%); and EGPA: petechiae or purpura (N=50; 21%), maculopapular rash (N=36; 15%), pruritus (N=30; 13%), and urticaria (N=19; 7.9%).

Conclusion: This is the largest study of cutaneous manifestations of AAV ever conducted. Utilizing data collected comprehensively via a standard protocol, it demonstrates that skin lesions are quite common and varied in GPA, MPA, and EGPA. Future analyses will focus on examining the association of specific cutaneous manifestations of AAV with other organ system involvement and laboratory findings. Table 1. Cutaneous findings among patients with ANCA-associated vasculitis

GPA N = 702 MPA N = 331 EGPA N = 241 p-value Male, N (%) 352 (50%) 144 (44%) 124 (51%) 0.09 Age, mean (SD) 53 (16) 64 (14) 54 (15) <0.001 PR3-ANCA, N (%) 515 (74%) 22 (6.7%) 10 (4.2%) <0.001 MPO-ANCA, N (%) 66 (9.4%) 286 (86%) 89 (37%) <0.001 Cutaneous Findings, N (%) 239 (34%) 97 (29%) 113 (47%) <0.001 Pruritus 26 (3.7%) 10 (3.0%) 30 (13%) <0.001 Painful skin lesions 66 (9.4%) 16 (4.8%) 24 (10%) 0.03 Cutaneous infarct 12 (1.7%) 1 (0.3%) 5 (2.1%) 0.10 Petechiae / purpura 113 (16%) 33 (10%) 50 (21%) <0.001 Maculopapular rash 47 (6.7%) 20 (6.0%) 36 (15%) <0.001 Livedo reticularis 4 (0.6%) 19 (5.7%) 4 (1.7%) <0.001 Livedo racemosa 2 (0.3%) 6 (1.8%) 1 (0.4%) 0.03 Non-tender nodules 8 (1.1%) 5 (1.5%) 5 (2.1%) 0.56 Tender nodules 21 (3.0%) 6 (1.8%) 4 (1.7%) 0.36 Gangrene 11 (1.6%) 4 (1.2%) 4 (1.7%) 0.91 Splinter hemorrhage 11 (1.6%) 1 (0.3%) 6 (2.5%) 0.06 Ulcer 30 (4.3%) 5 (1.5%) 10 (4.2%) 0.07 Urticaria 5 (0.7%) 2 (0.6%) 19 (7.9%) <0.001 Other 33 (4.7%) 15 (4.5%) 14 (5.8%) 0.75

ANCA: anti-neutrophil cytoplasmic antibody-associated vasculitis; SD: standard deviation; PR3: proteinase 3; MPO: myeloperoxidase; GPA: granulomatosis with polyangiitis; MPA: microscopic polyangiitis; EGPA: eosinophilic granulomatosis with polyangiitis.

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