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POS0250 SIGNIFICANT DAMAGE OCCURS EARLY IN THE COURSE OF EOSINOPHILIC GRANULOMATOSIS WITH POLYANGIITIS AND IS MAINLY DUE TO DISEASE-RELATED SEQUELAE

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Background: Following the introduction of effective immunosuppressive treatments, ANCA-associated vasculitides (AAV) have become chronic diseases with a remitting-relapsing course. Therefore, preventing chronic damage accrual during follow-up is critical, as relapses, treatment-related side effects, and comorbidities may significantly affect the long-term outcomes of AAV patients. At present, no study specifically evaluated the burden of damage in patients with eosinophilic granulomatosis with polyangiitis (EGPA).
Objectives: To describe short-term (6 months) and long-term (5 years) damage accrual in patients with newly diagnosed EGPA.
Methods: Patients diagnosed with EGPA, according to ACR criteria and/or Chapel Hill definitions and regularly followed-up in our vasculitis center for ≥5 years were included. Damage accrual was assessed with the Vasculitis Damage Index (VDI). Short-term and long-term damage accrual was defined by VDI at 6 months and at 5 years, respectively, and categorized as related to vasculitis or its treatment.
Results: VDI data at 6 months were available for 45 EGPA patients: 24 (53.3%) female, mean age at diagnosis 51.6±13.0 years. ANCA were positive in 17 patients (37.8%), with MPO being the only detected enzyme immunoassay (EIA)-specificity. At 6 months mean VDI was 2.8±1.3; 25/45 (55.6%) and 6/45 patients (13.3%) presented ≥3 and ≥5 items, respectively, whilst only 1 patient (2.2%) showed no items of damage. VDI data at 5 years were available for 32/45 EGPA patients (71.1%): 16 (50%) female, mean age at diagnosis 51.5±13.1 years. MPO-ANCA were positive in 13 patients (40.6%). At 5 years mean VDI was 3.5±1.3, with 26/32 (81.3%) and 7/32 patients (21.9%) presenting ≥3 and ≥5 items, respectively; notably, no patients presented a VDI=0 at 5 years.
The most frequent disease-related VDI items at 6 months and at 5 years were asthma, chronic sinusitis, peripheral neuropathy, cardiomyopathy, pulmonary function tests abnormalities and nasal blockage (Figure 1). Osteoporotic fractures, diabetes and systemic hypertension were the most commonly reported treatment-related items at 6 months and at 5 years (Figure 1). Damage accrual progressively rose during the 5-year follow-up (P=0.023), mainly due to disease-related items rather than treatment-related items both at 6 months (disease related VDI 2.6±1.2, treatment-related VDI 0.3±0.6) and at 5 years (disease related VDI 2.9±1.2, treatment-related VDI 0.6±0,7). No significant difference in terms of damage accrual was observed between ANCA-positive and ANCA-negative patients (P >0.5).
Conclusion: In our cohort of EGPA patients damage accrual occurs early, with more than half of the patients displaying ≥3 VDI items already at 6 months. Poor control of previous disease activity, particularly ENT and respiratory manifestations, contributes to progressive damage accrual more than treatment side effects.

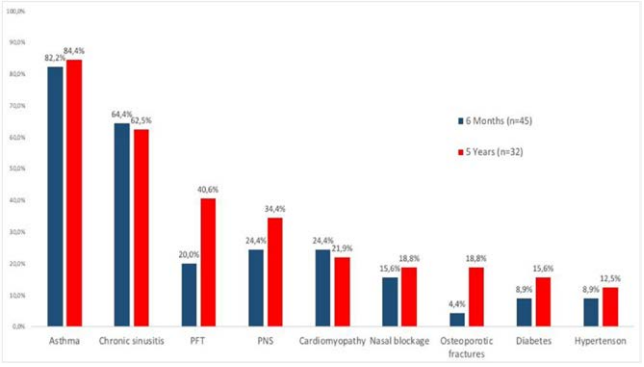

Figure 1. Disease-related and treatment-related VDI items at 6 months and at 5 years in patients with EGPA.

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POS0251 ARE CLINICAL FEATURES AND OUTCOMES OF ANCA (+) EGPA PATIENTS DIFFERENT FROM ANCA NEGATIVE ONES?

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Background: Recent reports describe EGPA as having two subgroups as ANCA positive and ANCA negative. Furthermore, there could be differences in terms of clinical features and outcomes (1).
Objectives: The aim of this study was to compare the clinical characteristics and outcomes of ANCA positive EGPA patients with those of ANCA negative ones.
Methods: This retrospective, descriptive study included 50 EGPA patients (male/female:13/27) from our prospective vasculitis database from October 2014 (2). The patients had fulfilled the ACR 1990 or DCVAS criteria for EGPA. In addition to clinical features (activity index) and treatment regimens, outcomes of patients (relapse and damage) were also reviewed. For relapse-free survival analysis, time to first relapse was compared according to ANCA phenotype by Kaplan-Meier survival analysis.
Results: Of the patients, 17 (34%) were in ANCA (+) group, 33 (66%) were in ANCA (-) group. Renal involvement and peripheral neuropathy were more frequent in ANCA (+) patients whereas ANCA (-) patients were significantly younger at the time of diagnosis and they had more nasal polyposis. The median BVAS at the EGPA diagnosis was significantly high in ANCA (+) group. All 3 of patients with cardiac involvement were in the ANCA (-) group but difference was found in terms of FFS and VDI between ANA (+) and (-) groups. However, pulse steroid and cyclophosphamide treatments were more commonly used in ANCA (+) group while mepolizumab were used in ¼ of ANCA (-) patients (Table 1). During median 47 (IQR 69.9) months follow up, about 40% of patients had at least one relapse but there was no difference for relapse-free survival rate according to the ANCA status (Figure 1).
Conclusion: Not only clinical features and disease activity but also treatments received were significantly different between ANCA (+) and (-) patients. These results could partially define two distinct subgroups of EGPA. However, these groups were similar regarding damage and relapse.

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Table 1. Demographics and clinical characteristics of the patients with EGPA during disease course

Characteristics*	ANCA (+) (n=17)	ANCA (-) (n=33)	P
Age at the diagnosis (years)	53.8 ± 16.3	37.9 ± 14.3	0.001
Sex, female	10 (58.8)	17 (51.5)	0.62
Asthma	12 (70.6)	29 (87.9)	0.13
Nazal polyp	2 (11.8)	15 (45.5)	0.017
Eosinophilia (>1000/μL)	11 (64.7)	27 (81.8)	0.18
Disease duration (years)	4.1 (5.9)	3.7 (5.9)	0.69
Organ specific involvement, %			
- Pulmonary	76.5	78.8	0.85
- ENT	82.4	87.9	0.59
- Renal	52.9	3	<0.001
- PNS	47.1	9.1	0.002
- Cardiac	0	9.1	N/A
BVAS at the diagnosis	17 (13)	9 (4)	0.002
Revized FFS			
- 0	11 (64.7)	26 (78.8)	0.28
- ≥ 1	6 (35.3)	7 (78.8)	
VDI	1 (1)	1 (1)	0.41
Treatment regimens for induction, %			
- Pulse steroid	64.7	11.2	0.002
- Cyclophosphamide	58.8	18.2	0.004
- Rituximab	11.8	3	0.21
- Mepolizumab	0	12.1	N/A
Treatment regimens for maintenance, %			
- Rituximab	11.8	3	0.21
- Mycophenolate mofetil	35.3	3	0.002
- Mepolizumab	0	24.2	N/A
- Azathioprine or methotrexate	47.1	42.4	0.75
Relapse, n (%)	7 (41.2)	12 (42.9)	0.66
Exitus, n (%)	2 (11.8)	1 (3)	0.26

* Med (IQR) for numerical data excluding age; mean ± SD for ageANCA: Antineutrophil cytoplasmic antibody, BVAS: Birmingham Vasculitis Activity Score, ENT: Ear, nose, and throat, IQR: Interquartile range, med: median, N/A: Not applicable, PNS: Peripheral nervous system, VDI: Vasculit damage index