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POS0829 SPECIFICITY OF PANCA AUTOANTIBODIES IN AUTOIMMUNE DISEASES

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Background: The clinical significance of pANCA by indirect immunofluorescence is well-established. However, their clinical utility is sometimes hindered by the fact that pANCA are also detected in various autoimmune diseases. Myeloperoxidase (MPO) is considered as the major autoantigen recognized by pANCA in ANCA-associated vasculitides (AAV) and predominantly in microscopic polyangiitis (MPA). However, information regarding the targets of pANCA in other autoimmune diseases is still elusive.

Objectives: To investigate the specific autoantigens recognized by pANCA in autoimmune diseases.

Methods: Sera from all patients that were found positive for pANCA in the diagnostic laboratories of the Department of Pathophysiology, School of Medicine, National and Kapodistrian University of Athens and the Department of Immunology and Histocompatibility, Evangelismos General Hospital, Athens, Greece during the last two years were studied. The pANCA+ sera were evaluated for reactivity with the major antigens that are recognized by pANCA [MPO, lactoferrin, neutrophil elastase, cathepsin G and bactericidal/permeability increasing protein (BPI)] by a commercially available multiplex ELISA (ANCA profile ELISA, Euroimmun, Lubeck, Germany).

Results: A total of 82 patients were included in the study. All patients had positive pANCA by indirect immunofluorescence with a titre ranging from 1/160 to 1/640. According to respective classification criteria, 21 patients had systemic vasculitides (15 MPA, 1 granulomatosis with polyangiitis; GPA, 1 Behcet's disease; BD, 1 aortitis, 2 Henoch-Schönlein purpura; HSP and 1 cryoglobulinemic vasculitis; CV), 29 had systemic lupus erythematosus (SLE), 6 antiphospholipid syndrome (APS), 8 Sjögren's syndrome (SS), 2 rheumatoid arthritis (RA), 1 systemic scleroderma (SSc), 14 Hashimoto thyroiditis and 1 sarcoidosis. The specificities of pANCA in each entity are shown in the following table.

Autoimmune Diseases		Antigens recognized by pANCA+ sera				
		MPO	Elastase	Cathepsin G	BPI	Lactoferrin
Vasculitides	MPA	66.7 (10/15)	0 (0/15)	0 (0/15)	0 (0/15)	0 (0/15)
	GPA	100 (1/1)	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)
	BD	100 (1/1)	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)
	Aortitis	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)
	HSP	0 (0/2)	0 (0/2)	0 (0/2)	0 (0/2)	0 (0/2)
SLE	CV	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)	100 (1/1)
		6.9 (2/29)	0 (0/29)	0 (0/29)	0 (0/29)	6.9 (2/29)
APS		16.6 (1/6)	16.6 (1/6)	0 (0/6)	0 (0/6)	0 (0/6)
SS		0 (0/8)	12.5 (1/8)	0 (0/8)	12.5 (1/8)	0 (0/8)
RA		50 (1/2)	0 (0/2)	0 (0/2)	0 (0/2)	0 (0/2)
SSc		100 (1/1)	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)
Hashimoto		0 (0/14)	0 (0/14)	0 (0/14)	0 (0/14)	0 (0/14)
Sarcoidosis		0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)	0 (0/1)

Conclusion: pANCA positive staining in AAVs is highly restricted to MPO specificity. On the contrary, pANCA staining pattern in other autoimmune diseases, involves unknown autoantigens that are under investigation in our laboratory.

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POS0830 FACTORS INFLUENCING PATIENT-REPORTED OUTCOMES IN ANCA-ASSOCIATED VASCULITIS

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Background: Patient-reported outcomes (PROs) are currently poorly integrated in the clinical evaluation of disease activity in patients with ANCA-associated vasculitis (AAV).

Objectives: To assess the distribution of the Patient Global Assessment (PtGA) in patients with AAV in stable remission, and to identify correlates of PtGA; to assess the discordance between PtGA score and PhGA.

Methods: Patients with a diagnosis of AAV [eosinophilic granulomatosis with polyangiitis, granulomatosis with polyangiitis, microscopic polyangiitis] in stable, complete remission (defined by a BVAS=0) and with a Physician Global Assessment (PhGA)=0 were included. A questionnaire including several aspects of disease captured by PROs was collected. PtGA on a 0-100 mm visual analogue scale (VAS) was assessed, with higher scores representing higher/worse levels of disease activity. Similarly, VAS for pain, chronic damage according to the patient's opinion, general health (GH), fatigue, and sleep quality were collected. The worst symptom in the patient's opinion affecting the overall assessment of disease activity was recorded. The Cragg Hurdle model was used to assess the predictors of PtGA.

Results: 65 patients were included, female 57%, mean age 61±12 years. Mean disease duration at enrollment was 8±6 years. Mean vasculitis damage index (VDI) was 4.4 ±2.3, with 45% of patients having a VDI ≥ 5. Despite having been classified as being in remission, PtGA was elevated in 37% of patients. We explored several correlates of PtGA. Higher degree of damage accrual (VDI) did not influence the patient's evaluation of current disease activity. Similarly, we did not identify a correlation between older age, educational level, number of organ-systems involved, number of comorbidities, the number of previous major or minor relapses, higher disease duration, nor the type of AAV diagnosis (figure 1, panel A). Only sex significantly correlated with PtGA scores: 19 (51%) of female patients reported an elevated PtGA compared to only 5 (18%) of male (p=0.009). PtGA resulted to be significantly correlated with other (mostly modifiable) PROs including VAS pain, perception of the level of chronic damage accrual, GH, and fatigue (figure 1, panel B). The agreement between patients' and physicians' assessments of disease activity was 63%. Patients reported pain, followed by chronic respiratory symptoms to be the worst-experienced ongoing manifestations affecting their evaluation of disease activity.

Conclusion: A significant proportion of patients with AAV considered to be in remission by the physician still declares to have persistent aspects of uncontrolled disease. PtGA is significantly influenced by persistent pain and fatigue, which warrant better assessment in the future.

Figure 1. Forrest plots of correlates of the Patient Global Assessment (PtGA): patient- and disease-related items (Panel A); influence of other patient-reported outcomes (Panel B). Cut-off values were identified based on the median value of the population.