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Background: Giant cell arteritis (GCA) is a large-vessel vasculitis affecting elderly people, and most frequently women (sex-ratio of 2.3). Some studies suggest an increased risk of malignancies in GCA.
Objectives: We aimed to describe the clinical, paraclinical characteristics and outcomes of GCA patients with concomitant malignancy and compare them to a control group without malignancy.
Methods: Patients with a diagnosis of GCA and of solid neoplasm or malignant blood disease, within one year before or after the diagnosis of vasculitis, were included. A random group of age-matched (3:1) control patients from our monocentric inception cohort of GCA patients from Caen University Hospital was constituted.
Results: Twenty-four observations were collected (median age 75.5 years). All fulfilled ≥3/5 ACR criteria. Temporal artery biopsy was positive in 17 cases (70.8%). There were 1 active (4.2%) and 9 former (37.5%) smokers. Only 1 patient had a previous prostate cancer. Malignancies were 10 malignant blood diseases (41.7%, 3 chronic lymphoid leukemias, 3 essential thrombocythemias, 1 myeloma, 1 chronic myelomonocytic leukemia, 1 MALT lymphoma, 1 Waldenström's macroglobulinemia) and 14 solid neoplasms (58.3%, 3 lung, 3 breast, 2 prostate, 1 thyroid, 1 colon, 1 pleural cancers, 1 melanoma, 1 Kaposi's sarcoma and 1 Merkel cell carcinoma). Malignancy was diagnosed at a median of 1 month after GCA diagnosis in 21 patients and before in the other 3. Diagnosis of malignancy was made in consultation in 5 patients (3 skin cancers and 2 breast cancers), on lab tests in 13 (thrombocytosis, anemia or increased prostate specific antigen) and on imaging in 6. Treatments of malignancy included chemotherapy alone in 8 patients (33.3%), simple monitoring in 6 patients (25%), surgery alone in 4 patients (16.7%), surgery and radiotherapy and/or chemotherapy in 4 patients (16.7%), decrease of corticosteroids in 1 patient, and 1 patient was lost to follow-up. Two patients (8.3%) died from infectious complications, 8 patients (33.3%) had a GCA relapse, including one with concomitant malignancy relapse. After a median follow-up of 16 months [0–134], 5 patients (20.8%) were weaned from steroids, all considered in malignancy remission. Seven patients (29.1%) were still under chemotherapy, 9 patients (37.5%) were considered to be in malignancy remission. There were more males in patients with concomitant malignancy, compared to the control group (respectively 15/24 and 21/72, p<0.005).
Conclusions: Our study shows an over-representation of male gender in GCA with concomitant malignancy. Vasculitis outcomes were not influenced by the malignancy treatment. The diversity of malignancies encountered in this study raises the issue of an incidental association. Initial clinical and paraclinical follow-up dictated by vasculitis may have led to an early identification of associated malignancy, and thus represent a lead time bias.
Disclosure of Interest: None declared
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AB0589 THE ROLE OF ANCA SPECIFICITY IN THE CLINICAL MANIFESTATIONS AT DISEASE ONSET: COMPARISON BETWEEN PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS AND MICROSCOPIC POLYANGIITIS

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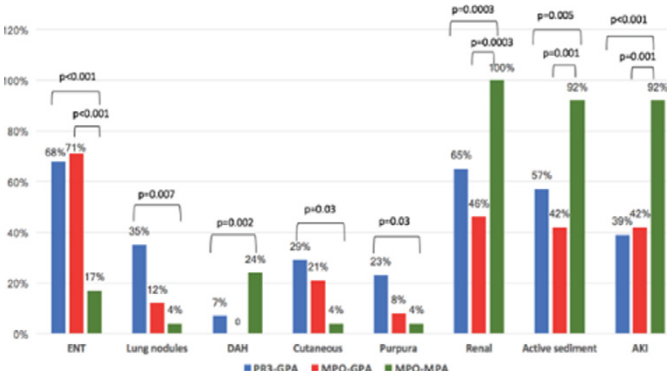
Background: ANCA specificity, rather than clinical diagnosis, has been suggested to influence the phenotype and clinical course of ANCA associated vasculitis (AAV) (1,2).
Objectives: To investigate differences in clinical presentation at disease onset between MPO-ANCA-positive granulomatosis with polyangiitis patients (MPO-GPA), PR3-ANCA-positive-GPA (PR3-GPA), and MPO-ANCA-positive microscopic polyangiitis (MPO-MPA).
Methods: Clinical records of AAV patients from three third level rheumatologic centers in Northern Italy were retrospectively analyzed.
Results: Of the 133 AAV patients included, 84 were PR3-GPA, 24 MPO-GPA, and 25 MPO-MPA. Patients with MPO-MPA were significantly older at diagnosis compared to both PR3-GPA and MPO-GPA (average age 63±10, 49±15, 55±29, respectively) (Table 1).
Patients with MPO-GPA experienced a significant diagnostic delay compared to PR3-GPA (17±30 vs 7±14, p=0.02). ENT involvement was equally frequent in both GPA groups despite ANCA specificity, and significantly more represented than the MPO-MPA group (68%, 71% and 17% respectively; p<0.001). Figure 1. Renal involvement was significantly more frequent in MPO-MPA patients (100%) compared to GPA (p<0.001), without differences between MPO-GPA (46%) and PR3-GPA (65%). Alveolar haemorrhage (DAH) was an onset manifestation mainly in MPO-MPA compared to the other two groups (24% vs 7% in PR3-GPA; p=0.02). Cutenous manifestations, mainly purpura, were significantly more reported in PR3-GPA compared to MPO-MPA (29% vs 4%; p=0.03).
Conclusions: Clinical phenotype of GPA at disease onset did not seem to be influenced by ANCA specificity. Despite ANCA positivity (PR3 or MPO), GPA patients were significantly different from MPA.

Table 1. Clinical characteristics of patients with GPA and MPA at disease onset according to ANCA specificity

	GPA-PR3 (A) (n=84)	GPA-MPO (B) (n=24)	MPA-MPO (C) (n=25)	p
Male/female n (%)	46 (55%)/38 (45%)	10 (42%)/14 (58%)	10 (40%)/15 (60%)	0.29
Age at disease onset (average ± SD)	49±15	55±19	63±10	**p<0.001** A vs C p<0.001 B vs C p=0.03
Diagnostic delay (months ± SD)	7±14	17±30	10±19	0.07
Systemic symptoms	54 (65%)	16 (67%)	19 (76%)	0.59
ENT	55 (68%)	17 (71%)	4 (17%)	**<0.001**
Pulmonary	57 (69%)	16 (67%)	16 (64%)	0.86
Cutaneous	24 (29%)	5 (21%)	1 (4%)	**0.03**
Ocular	16 (20%)	3 (12%)	2 (8%)	0.34
Cardiovascular	4 (5%)	2 (9%)	1 (4%)	0.77
Gastrointestinal	2 (3%)	0	0	0.56
Renal	53 (65%)	11 (46%)	25 (100%)	**<0.001**
Nervous system	26 (31%)	8 (33%)	8 (35%)	0.94

Figure 1. Differences in onset clinical manifestations among PR3-GPA, MPO-GPA and MPO-MPA



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AB0590 PERFORMANCE OF 2017 ACR/EULAR PROVISIONAL CLASSIFICATION CRITERIA FOR GRANULOMATOSIS WITH POLYANGIITIS IN CHILEAN POPULATION

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Background: Anca Associated Vasculitis (AAV), are a group of necrotizing primary vascultitis, whith multisistemic manifestation, of unknown etiology. The variants are: Microscopic Polyangiitis (MPA), Granulomatosis with polyangiitis (GPA), Granulomatosis whith Polyangiitis and Eosinophilia (GPE) and AAV limited to one organ. Until now, there are no diagnostic criteria for AAV. Therefore definitions, as Chapell Hill consensus Conference Nomenclature, classification criteria and the physician judgement are used for diagnosis. Currently the DCVAS (Diagnosis and Classification Criteria in Vasculitis) proyect is developing diagnostic criteria for AAV, using data-driven methods. The preliminary DCVAS classification criteria for granulomatosis whith polyangiitis has been recently realesead.
Objectives: To evaluate and compare the acuraccy of ACR/EULAR 2017 provisional Classification Criteria for GPA whith the ACR 1990 Classification Criteria In Chilean patients with AAV.
Methods: All adult patients (>18 yo) with diagnoses of AAV according to their rheumatologist judgment, from 2000–2016 at the University of Chile, Clinical Hospital (UCCH), were included. Clinical variables of interest were extracted form medical chart and AAV database, wich is kept for these patients at the Rheumatology Section of UCCH. Based on that data, the Classification criteria ACR 1990 and 2017preliminary ACR/EULAR (DCVAS) classification criteria for GPA were applied to each individual. Sensibility, especifity, Likehood ratio (LR +/-), predictive values (PPV/NPV) and accuracy were calculated for both sets of Criteria as compared to Clinical diagnosis
Results: 93 patient were included in the study. 59 patients with GPA, 33 with