Background: Giant cell arteritis (GCA) presents two phenotypic patterns: cranial and extracranial. Cranial manifestations are relatively easier to recognize and to diagnose, however since the extracranial are more insidious and less known, it may lead to a delay in diagnosis and in treatment. The main treatment are glucocorticoids (GC) at high doses followed by a descending pattern. The time between the begin of the symptoms and the start of the treatment with GC it's of vital importance in the prognosis of the disease.
Objectives: The present AUDIT describes our cohort's clinical features, the time from symptom onset to rheumatology visit and diagnosis of GCA, as well as the differences in time according to the service originating the referral.
Methods: Audit retrospective, descriptive study of patients diagnosed with GCA by the CGA 1990 criteria. All patients diagnosed with GCA from 2011 to 2021, in a single hospital center and referred to the Rheumatology Service were included. Patients were classified as: cranial phenotype, extracranial or mixed. Demographic and clinical variables and complementary examinations were collected. As well as the time from the first symptom until the patients are referred and diagnosed.
Results: Seventy patients were included in the study. Fifty-one patients (72.9%) were female, their clinical features are shown in Table 1. Twelve of them (17,39%) had a cranial phenotype, sixteen (23,18%) an extracranial phenotype and forty-one (60,42%) a mixed one. Extracranial phenotype patients showed to be younger at the time of the diagnoses than the other groups. Cranial phenotype patients had a lower increase of the acute phase reactants than the other phenotypes (without finding statistically significant differences). In Table 2 are shown the other demographic, clinical and analytic data.

Table 1. Disease Characteristics of the Patients at Baseline according to their phenotype

PHENOTYPE	CRANIAL	EXTRACRANIAL	MIXED	TOTAL
	N=12	N=17	N=41	N=70
Age	75.543 ± 7.48	73.543 ± 8.1	76.356 ±7.46	75.56 ±7.46
Smoker	8.3% (n=1)	11.8% (n=3)	14.6% (n=6)	12.9% (n=9)
Headache	91.7% (n=11)	0%	90.2% (n=37)	68.6% (n= 48)
Ocular involvement	58.3% (n=7)	0%	34.1% (n=14)	30% (n=21)
Fever of unknow origin	0%	29.4% (n=5)	34.1% (n=14)	27.1% (n=19)
Polymyalgia rheumatica	0%	41.2% (n=7)	53.7% (n=22)	41.4% (n=29)
Constitutional syndrome	0%	23.5% (n=4)	36.6% (n=15)	27.1% (n=19)
Cutaneous hyperalgesia	25% (n=3)	0%	31.7% (n=13)	22.9% (n=16)
Anaemia	0%	41.2% (n=7)	46.3% (n=19)	37% (n=26)
C reactive protein	51.33 ± 40.16	77.83 ± 75.30	85.55 ± 81.67	77.58 ± 74.57
Erythrocyte sedimentation rate	60.64 ± 33.13	76.20 ± 36.35	77.93 ± 35	74.65 ± 35.07

Patient referral were made from: Emergency room/ Internal Medicine hospitalization (30%, 21 patients), Primary Care (21.4%, 15 patients), Internal Medicine outpatient clinic (15.7%, 11 patients), Neurology (18.6%, 13 patients) and other specialties outpatient clinic (cardiology, vascular surgery, ophthalmology, hematology and gastroenterology). The median of the time from the first symptom until the patients are diagnosed are 12,9, 13,97 and 7,91 weeks in the cranial, extracranial and mixed respectively. The median of time from the first symptom until the patient referral to rheumatology are 9.34, 12.74 and 5.87 weeks in the cranial, extracranial and mixed respectively. The median of time from patient referral to rheumatology to be assessed are 3.73, 5.59 and 3.43 weeks in the cranial, extracranial and mixed respectively. 75.7% of the patients received treatment with glucocorticoids before being referral to rheumatology.
Conclusion: In our cohort, most of the patients with CGA had a mixed phenotype. The patient referral to rheumatology is made mainly from the emergency room, internal medicine hospitalization and primary care. Patients with mixed phenotype are diagnosed faster than the ones with an exclusively extracranial phenotype. The time between the first symptom and the referral to rheumatology is still long.
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.4888

AB0609 GAPS IN THE DIAGNOSIS OF TAKAYASU ARTERITIS: A ROMANIAN COHORT

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Background: The diagnosis of Takayasu arteritis still represents a big challenge, even though the disease was first described more than 100 years ago. Proof of that is the continuing effort to develop criteria with high specificity and sensitivity.[1]
Objectives: The aim of the study was to describe the clinical and paraclinical aspects of the disease at the time of diagnosis. Additionally, we wanted to evaluate the damage score in view of the diagnosis delay.
Methods: We retrospectively studied the clinical chart of 17 consecutive patients admitted to the Rheumatology Clinic of Emergency County Hospital from Cluj-Napoca, Romania between 2003 and 2021.
Results: There was just one man in the cohort (0.05%). The average age at the time of diagnosis was 37.5 years old. The average delay in the diagnosis was 33 months, while the biggest delay was of 240 months. The diagnosis was made by rheumatologists in 58% of the cases, while 29% by cardiologists and 11.7% by internal medicine specialists. The most frequent symptoms were limb claudication (64.7%), fatigue (52.9%) and headache (52.9%). Vascular bruits were found in most of the patients at the time of diagnosis (70.5%), as well as blood pressure and pulse difference between arms (52.9%). Unobtainable blood pressure was seen in 5 patients (29.4%). There was a discrepancy in 3 patients (17.6%) between inflammatory markers and inflammation in the vessels' walls, as confirmed by imaging techniques (angioMRI or angioCT). Most of the patients were classified as type V arteritis (35%), followed by type IV (18%), type I (17%), type IIA, respectively type IIB (12% each) and type III (6%). Stenosis of the involved arteries occurred 4.3 times more than aneurysm formation. Eight patients required vascular interventions at the time of diagnosis – four were treated by balloon angioplasty and six by stent placing. The average TADS (Takayasu Arteritis Damage Score) was 1.5 and it was not associated with delay in diagnosis (p>0.05).
Conclusion: Takayasu arteritis is a disease with heterogenous and nonspecific symptoms. For those reasons the delay in diagnosis is usually long, the treatment is started late and damage appears early in the course of the disease. This is reflected by the high amounts of vascular procedures needed at the time of diagnosis. We need better diagnostic criteria and specific monitoring scores.
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Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2022-eular.4953

Vasculitis - small vessel vasculitis

AB0610 DEVELOPMENT OF MALIGNANCY IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

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Background: Patients (pts) with antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) reported to have an increased risk of malignancy compared to general population [1]. However, studies on risk factors for the development of malignancy in AAV pts are limited.
Objectives: We aimed to evaluate the frequency, clinical features and associated factors of malignancy in pts with AAV.
Methods: In this study, we retrospectively evaluated 287 pts with AAV. Thirty-three pts with EGPA and 14 pts with missing data were excluded. ANCA test was analysed based on immunofluorescence and/or Elisa results and divided into two serological groups; c-ANCA/PR3$^+$ (positive) and p-ANCA/MPO$^+$.
Results: Data of 240 pts (54.6 % female) were analysed. Mean age of diagnosis was 55.6±14 (range; 17-88) years and median disease duration was 67.5 (range; 3-255) months. Of those pts, 175 (73 %) had GPA and 65 (27 %) had MPA. ANCA results were available in 230 pts; 123 were c-ANCA/PR3$^+$ (53.5 %), 85 were p-ANCA/MPO$^+$ (37 %) and 22 were ANCA negative (ANCA$^-$) (9.5 %). Kidney (75.8 %) and lower respiratory tract (74.4 %) were the most common organs involved but they did not differ according to presence of malignancy (9.4 % vs 5.1 %; p=0.4 and 9.4 % vs 5 % p=0.4 respectively.) Twenty-two malignancies were observed in 20 pts (8.3 %; eleven c-ANCA/PR3$^+$, three p-ANCA/MPO$^+$ and two ANCA negative pts). Lung and thyroid papillary cancer in three; bladder, prostate, breast and kidney in two; adrenal gland, oral squamous cavity and sarcoma of retroperitoneum in one patient, hematological (three myelodysplastic syndrome, one chronic lymphocytic leukemia and one lymphoma) in five pts. Six pts (30 %) had prior and/or concomitant AAV diagnosis. Development of malignancy did not differ according to age, gender, diagnosis, seropositivity (ANCA$^+$ vs ANCA$^-$) and ANCA subgroups (Table 1). There was no association between malignancy and cumulative dose of cyclophosphamide (CYC) or history of smoking (p=0.96 and p=0.2, respectively). In univariate analysis, malignancy was associated with presence of cardiovascular disease (CVD) (p=0.003 OR 11.7) and mortality (p=0.04 OR 4.6), higher BVAS score at baseline (p=0.049) and higher VDI score (p=0.02). Significant association was observed between malignancy with CVD (95 % CI 2.2-83 OR 13.4 p=0.005) and mortality (p=0.044 95 % CI 1.03-8.5 OR:2.95) in multivariate analysis.

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Table 1. Factors associated with malignancy in pts with AAV

	Univariate analysis		
Variables	**Malignancy + (n=20)**	**Malignancy – (n=220)**	**p value (OR)**
Age (years) ǂ	60±13.4	55.3±14	0.15
Gender (female)	10 (50)	121 (55)	0.7
Diagnosis *			
GPA	15 (8.6)	160 (91.4)	0.8
MPA	5 (7.7)	60 (92)	
ANCA status*			
c-ANCA/PR3	11 (9)	112 (91)	0.16
p-ANCA/MPO	3 (3.5)	82 (97)	
Cardiovascular disease*	8/38 (21)	10/199 (5)	***0.003 (11.7)***
Cerebrovascular accident*	2/15 (13)	16/222 (7)	0.3
Coronary heart disease*	4/18 (22)	19/218 (9)	0.08
Avascular necrosis*	4/35 (11)	14/199 (7)	0.3
Venous thrombosis*	3/21 (14)	15/217 (7)	0.2
BVAS score at admission ǂ	21.3±6	16.6±6.7	***0.049***
Smoking history (ever)*	6/53 (11)	8/156 (5)	0.2
Cumulative CYC dose (g) ᴷ	5 (7.4)	4.5 (7.8)	0.96
VDI scoreᴷ	3.5 (4)	2 (2)	***0.02***
Relapse (n=230, %)	5/80 (6.3)	11/150 (7.3)	0.8
Remission at six months (n=160, %)	8/104 (7.7)	5/56 (9)	0.8
Mortality*	7 (35)	35 (16)	***0.04 (4.6)***

OR: Odds ratio *(n, %) ǂ mean± Std dev. ᴷmedian; IQR

In survival analysis, mortality rate was higher in pts had malignancy compared to those without (Figure 1, p=0.035).

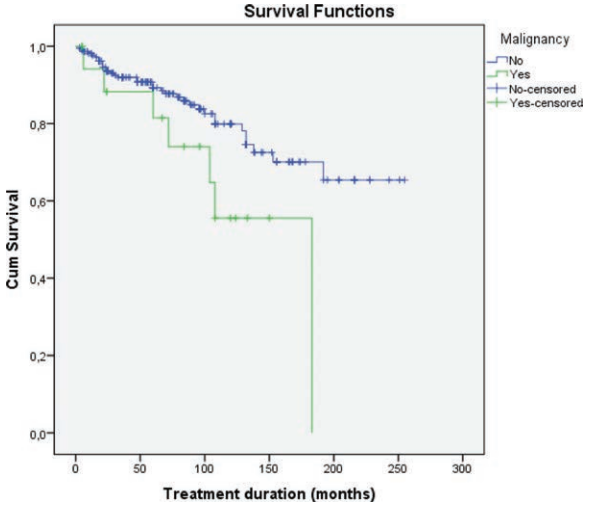


Figure 1. Survival analysis of mortality according to presence of malignancy in pts with AAV. Log-Rank: p=0.035

Conclusion: Significant proportion of pts with AAV developed malignancy in our cohort. Development of malignancy was associated with CVD, higher baseline BVAS and VDI scores. Our study also revealed lower survival rate in patients who developed malignancy. Further studies are needed to clarify risk factors for malignancy in patients with AAV.

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Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.249

AB0611

NEUTROPHIL EXTRACELLULAR TRAPS AND INTERSTITIAL LUNG DISEASE IN ANCA-ASSOCIATED VASCULITIS: A SCOPING REVIEW

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Background: Deregulated neutrophil extracellular traps (NET) formation is implicated in various diseases, including ANCA-associated vasculitis and pulmonary fibrosis (PF). Lung involvement is frequent in AAV, and interstitial lung diseases (ILD) are strongly related to MPO-ANCA positivity and mainly reported in microscopic polyangiitis. The association between AAV and ILD is a strong indicator of poor prognosis and limited survival. Neutrophils, ANCA and NET interplay in PF development in AAV.

Objectives: This study aimed to review the literature concerning the implications of NET in lung fibrogenesis focused specifically on AAV associated with ILD, and the potential of NET as a theranostic marker.

Methods: Through scoping review methodology, we used a descriptive thematic analysis to understand the pathogenic role of NETs in patients with AAV and pulmonary fibrosis and their further role as a theranostic marker of this disease.

Results: Quality assessment of articles was evaluated through SANRA methodology (Table 1). The implications of NET in the pathogenesis of AAV and ILD (Figure 1), as well as an association between these two diseases, have been identified, but the underlying pathophysiological mechanisms are still unknown. Pharmacological or genetic inhibition of NET release reduces disease severity in multiple inflammatory disease models, indicating that NETs are potential therapeutic targets. In this regard, despite the lack of clinical data, we may hypothesize that an optimal management of AAV-ILD patients would require not only B-cells targeted therapy, but also NETs inhibition.

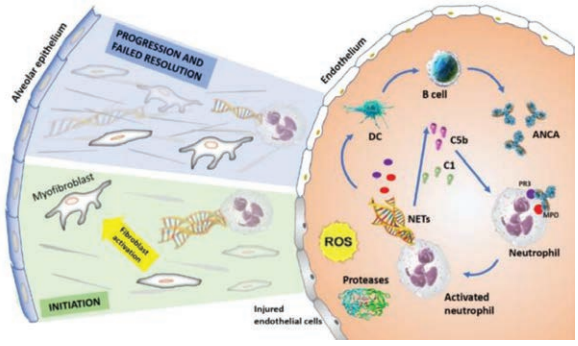


Figure 1. The role of NET in interstitial lung disease associated with ANCA-associated vasculitis

Conclusion: Preliminary findings seem to display a lack of efficacy of traditional immunosuppressants, such as Rituximab, in this subset of patients, while to date no patients suffering from a definite ILD was enrolled in clinical trials. Further insights would be provided by their employment, as a combination treatment, in the common clinical practice. Although we can imagine that inhibition of NETs in patients with AAV-ILD could reduce severity and mortality, we still lack a scientific basis that could improve our understanding of the disease from a molecular point of view.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2022-eular.479

Table 1. SANRA Score for quality assessment

N°.	Authors	Justification of the article's importance for the readership	Statement of concrete aims or formulation of questions	Description of the literature search	Referencing	Scientific Reasoning	Appropriate presentation of data	Total score
1	Pandolfi L et al. 2021	2	2	0	1	2	2	9
2	Radermecker C et al. 2020	1	2	0	2	2	1	8
3	Zhang S et al. 2019	2	1	0	2	2	1	8
4	Skopelja-Gardner S et al. 2019	1	1	0	1	2	2	7
5	Khawaja AA et al. 2020	2	2	0	1	2	1	8
6	Chrysanthopoulou A et al. 2014	2	1	0	2	2	1	8
7	Kraaij T et al. 2018	2	1	0	2	1	2	8
8	Tang S et al. 2015	2	1	0	1	2	1	7
9	Yoshida M et al. 2016	1	2	0	1	2	1	7
10	Nakazawa D et al. 2014	2	1	0	2	2	2	9
11	Hattanda F et al. 2019	2	2	0	1	2	1	8
12	Schreiber A et al. 2017	2	2	0	1	1	2	8
13	van Dam LS et al. 2019	2	2	0	1	1	1	7
14	Roitsch S et al. 2018	1	2	0	1	2	2	8
15	Takeuchi H et al. 2017	2	2	0	2	2	2	10
16	Su B et al. 2021	2	1	0	1	2	2	8
17	Söderberg D et al. 2015	2	1	0	1	2	1	7
18	Surmiak M et al. 2016	1	2	0	1	2	2	8

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