

Figure 2. Correlation coefficient and regression line of the number of Th17 cells, the number of Treg cells, Th17/Treg with the number of neutrophil, the number of lymphocyte, the NLR were represented as scatter plots.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.1411

FRI0209

EFFECTS OF ADD-ON METHOTREXATE IN POLYMYALGIA RHEUMATICA PATIENTS: A RETROSPECTIVE STUDY

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Background: Guidelines on polymyalgia rheumatica (PMR) recommend early introduction of methotrexate (MTX), especially in patients with worse prognosis such as flare or glucocorticoid (GC)-related adverse events (AE).¹ GC-AE are reported in up to 65% of PMR patients,² and 50% of secondary care patients are unable to discontinue GCs, emphasizing the need for GC-sparing agents.³ However, evidence regarding MTX efficacy in PMR remains limited.²

Objectives: To assess the efficacy of add-on MTX in preventing subsequent flares and GC-sparing in PMR patients.

Methods: In a retrospective cohort of newly diagnosed PMR patients visiting our hospital from April 2008 - January 2018, patients starting methotrexate (index event) were compared to first-time flaring PMR patients in whom MTX was not started (control group). Concomitant inflammatory rheumatic diseases were excluded. Data on patient, disease and treatment characteristics were compared. Main outcomes were difference in number of subsequent flares per year between groups (multivariable Poisson regression) and mean GC-use (total GC-dose/total follow-up; multivariable linear regression). In the MTX group only, also incidence rate ratio of flare before vs. after starting MTX was assessed.

Results: Of 454 PMR patients, 262 were selected; 42 receiving MTX and 220 in the control group. Reasons for prescribing MTX were GC ineffectiveness and/or GC-related AE and MTX starting dose was 10, 15 and 25 mg/week in 11%, 82% and 2% respectively. Adjusted for covariates, mean GC-use was 1.21 higher in the MTX group compared to the control group ($p = 0.155$). The yearly incidence rate of flares in the MTX group did not differ from the control group: incidence rate ratio (IRR) 0.93, (95% CI 0.53-1.63). The yearly flare rate was 1.19 before and 0.42 after MTX initiation, resulting in an IRR of 0.36 (95% CI 0.24-0.53).

Conclusion: MTX is infrequently prescribed in daily clinical practice, despite guideline recommendations. No difference in GC use or flare incidence was seen between MTX treated patients and controls, although within MTX treated patients, flare rates were lower after MTX start. Confounding by indication may explain the lack of difference in the outcomes between groups. The optimal timing and dosage of MTX in PMR remains unclear, justifying a clinical trial.

References:

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Table 1. Patients characteristics methotrexate versus controls

	MTX N = 42	Controls N = 220	P-value	Difference (95% CI)
Time of PMR diagnosis				
Female(%)	22(52)	127(58)	0.611	-5.3 (-11.0; 21.7)
Age, years*(SD)	62(7)	67(10)	0.002	-5.0 (-8.2; -1.8)
Previous history PMR*(%)	7(17)	13(6)	0.025	10.8 (2.0; 19.5)
PMR symptoms, weeks(IQR)	9(6-16)	5(5-16)	0.639	
Bilateral shoulderpain(%)	42(100)	209(95)	0.221	5.0 (-1.6; 11.6)
Morning stiffness>45 minutes(%;n=33 versus 159)	32(76)	177(80)	0.533	-4.3 (-17.5; 9.0)
Elevated ESR/CRP*, (%;n=41 versus 218)	40(96)	185(84)	0.023	12.7 (1.4; 24.0)
At index event				
PMR duration, weeks*** (IQR)	87(41-116)	53(33-80)	0.001	
GC-dose index event	39(93)			
Oral	10(5-15)	210(95)		
Mg(IQR)*	3(7)	5(0-8)	0.000	
Intramuscular(%)	120(100-120)	10(5)		
Mg(IQR)	120	100(80-120)	0.355	
Mean GC-dose until index event, mg, (IQR; n=33 versus 170)*	7.1 (5.9-9.0)	5.7 (3.8-7.4)	0.000	
During follow-up				
Flares, n(%)	21 (50)	100 (45)	0.616	4.5 (-21.0; 11.9)
Weeks to first flare (IQR)	36 (24-51)	39 (22-66)	0.517	
Mean GC-dose, mg (IQR)	6.2 (4.6-9.7)	4.7 (2.9-6.9)	0.004	
Daily GC-dose year1, mg (IQR; n=32 versus 153)*	5 (2.5-7.9)	2.5 (0-5)	0.03	

* Significant alpha level < 0.05

**Before diagnosis until index event

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.2986

FRI0210

ORBITAL PSEUDOTUMOR AMONG PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS – DATA FROM THE POLISH REGISTRY POLVAS

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Background: Orbital inflammatory masses have been described as the common manifestation of granulomatosis with polyangiitis (GPA) occurring in 7-45% of patients.

Objectives: Identification and characterization of patients with orbital pseudotumor among Polish patients based on the national vasculitis registry, POLVAS.

Methods: Clinical presentation and management of all GPA patients fulfilling ACR criteria or Chapel Hill Consensus Conference definition included to the Polish registry POLVAS who developed orbital masses in the course of GPA were evaluated.

Results: Ocular involvement was found in 114 (27%) of 417 GPA patients registered in POLVAS, 34 (8%) developed orbital masses. Mean patients' age was 47.8 (range from 19-75) yrs., 23 (67%) were women. Forty four per cent of the patients developed tumor at the beginning of the disease, 56% during relapse. Patients' characteristics on diagnosis of orbital mass: 24 cANCA, 2 pANCA, and 8 ANCA negative, 9% active smokers and 31% past smokers, 29% had localized disease, 21% early systemic and 50% systemic with organ involvement, 29% had other type of ophthalmological involvement before pseudotumor occurred, 88% had active paranasal sinus involvement, 41% lungs, 15% CNS, 15% skin and 6% heart manifestations. Thirty seven per cent of patients had positive nasal swabs cultures, 50% of which were positive for *Staphylococcus aureus*. In 65%, tumor occurred during steroid therapy (46% had prednisone more than 5mg/d) and 45% on immunosuppressive treatment (19% when treated with AZA, 16% MTX, 6.5% MMF and 3.5% CYC). Due to orbital mass 86.5% were treated with CYC and 13.5% with RTX. Twenty one per cent had complete remission of the pseudotumor, 76% partial remission and in 3% patients there was no response to the treatment; 43% developed visual impairment, 20% suffered from blindness.

Conclusion: Orbital inflammatory mass was not common manifestation of GPA among our patients. The mass developed at the beginning or in the course of the disease, even during immunosuppressive treatment. Orbital masses have been resistant to therapeutic interventions and were accompanied by high risk of visual impairment.

Disclosure of Interests: Anna Masiak: None declared, Marcin Ziętkiewicz: None declared, Krzysztof Wójcik: None declared, Katarzyna Wawrzycka-Adamczyk: None declared, Radosław Jeleniewicz: None declared, Marta Madej: None declared, Joanna Kur-Zalawska: None declared, Katarzyna Jakuszko: None declared, Małgorzata Wiśłowska: None declared, Hanna Storoniak: None declared, Michał Korniczak: None declared, Barbara Bułto-Piontecka: None declared, Iwona Brzosko: None declared, Małgorzata Stasiak: None declared, Eugeniusz Kucharz: None declared, Alicja Dębska-Ślizień: None declared, Maria Majdan Consultant of: Roche, Amgen, Speakers bureau: Roche, Amgen, Jacek Musiał: None declared, Zbigniew Zdrojewski: None declared

DOI: 10.1136/annrheumdis-2020-eular.2881

FRI0211 **VASCULITIS ASSOCIATED WITH MYELODYSPLASTIC SYNDROME AND CHRONIC MYELOMONOCYTIC LEUKEMIA: FRENCH MULTICENTER CASE CONTROL STUDY**

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Background: Myelodysplastic syndromes (MDS) and MDS/myeloproliferative neoplasms (MDS/MPN) can be associated with vasculitis.

Objectives: In this nationwide study by the "French Network of dysimmune disorders associated with hemopathies" (MINHEMON) the objective was to evaluate characteristics, treatment and outcome of vasculitis MDS-MDS/MPN.

Methods: Retrospective analysis of patients that presented a MDS/MPN associated with vasculitis and compared the overall survival and acute leukemia with MDS without vasculitis.

Results: Seventy patients with vasculitis and MDS/MPN were included, with a median age of 71.5 [21-90] years and male/female ratio of 2.3. Vasculitis was diagnosed prior to MDS/MPN in 31 patients (44.3%), with a median time of 27 months [1-120] between two diagnosis, and after in 20 patients (6 months [1-59]). In comparison to 183 MDS/MPN without dysimmune features showed no difference in MDS/MPN subtypes distribution nor median IPSS/CPSS scores in patients with and without vasculitis. The vasculitis subtypes was giant-cell arteritis (GCA) in 24 patients (34%). Eleven patients (20%) had Behçet's-like syndrome and 6 patients (9%) presented with polyarteritis nodosa. Steroids (60 mg/day [0-500] of prednisone equivalent) were used as first-line therapy for MDS/MPN vasculitis in 64/70 patients (91%) and 41 (59%) received combined immunosuppressive therapies during the follow-up. After the follow-up of 33.2 months [1-162], 31 patients (44%) finally experienced sustained remission. At least one relapse during the 33.2 months [1-162] follow-up occurred in 43 patients (61%). Relapse rates were higher in patients treated by DMARDs (odds ratio at 4.86 [95% CI 1.38 - 17.10]), but did not differ from biologics (odds ratio 0.59 [95% CI 0.11-3.20]) and azacytidine (odds ratio 1.44 [95% CI 0.21-9.76]) (steroids considered as reference). Overall survival and progression to acute myeloid leukemia in MDS/MPN vasculitis were not significantly different from MDS/MPN patients without any dysimmune features (p=0.5).

Conclusion: This first largest study of MDS/MPN vasculitis show no correlation of vasculitis subtypes with various subtypes and severity of MDS/MPN, and no significant impact of vasculitis on overall survival and progression to acute myeloid leukemia. The high relapse rates and steroid dependence raise the question of combined therapies to steroids. Whereas DMARDs use seem to be avoid specific azacytidine therapy could be considered for even low-risk MDS/MPN vasculitis.

Acknowledgments: minhemon gfm gfev
Disclosure of Interests: None declared
DOI: 10.1136/annrheumdis-2020-eular.4485

FRI0212 **THE ROLE OF AGE ON THE CLINICAL PRESENTATION AND RELAPSE RATES IN A LARGE COHORT OF 720 PATIENTS WITH GIANT CELL ARTERITIS**

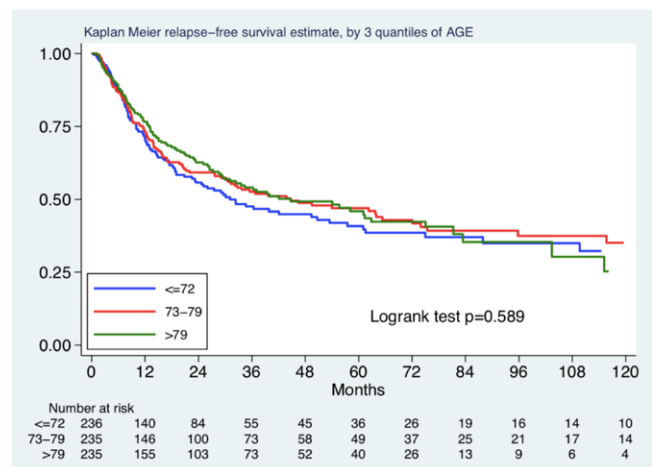
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Background: Giant cell arteritis (GCA) is the most frequent systemic vasculitis after the age of 50 years old. Recent interest in the processes of immune and vascular aging have been proposed as a disease risk factor. Data on the impact of age at diagnosis of GCA on the clinical course of the disease are scarce

Objectives: To assess the role of age at diagnosis of GCA on the risk and time to relapse

Methods: Centres participating in the Italian Society of Rheumatology Vasculitis Study Group retrospectively enrolled patients with a diagnosis of GCA until December 2019. The cohort was divided in tertiles according to age at diagnosis (≤ 72 ; 73-79; > 79 years old). Negative binomial regression was used to assess the relapse rate according to age groups, and Cox regression for time to first relapse.

Results: Of 720 patients enrolled in 14 Italian reference centres, 711 had complete follow-up data (female 50%; mean age 75 \pm 7). Median follow-up duration was 34 months (IQR 16;70). Patients in the older group at diagnosis (> 79 years) had more frequent visual loss compared to the 73-79 and ≤ 72 age groups (31% vs 20% vs 7%; p<0.001), but lower rates of general symptoms (56% vs 70% vs 77%; p<0.001). Large-vessel (LV)-GCA was less frequent in the older group (18% vs 22% vs 43%; p<0.001). At least one relapse occurred in 47% of patients. Median time to relapse was 12 months (IQR 6;23). Age did not influence the rate of relapses [18 per 100 persons/years (95%CI 15;21) vs 19 (95%CI 17;22) vs 19 (95%CI 17;22)], nor the time to first relapse (Figure 1). LV-GCA, presentation with significantly elevated c-reactive protein (> 50 mg/L) and general symptoms were independent predictors of relapse.



Conclusion: Age at diagnosis of GCA influenced the clinical presentation and risk of ischaemic complications, but did not affect the relapse rate during follow-up. LV-GCA occurred more frequently in younger patients and was an