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.

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FRI0211

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

A. Roupie¹, B. Terrier², P. Fenaux¹, O. Fain¹, A. Mekinian¹ on behalf of Minhemon. ¹Sorbonne University, Internal Medicine Department, Saint Antoine Hospital, Paris, France; ²Paris Descartes V University, Internal Medicine Department, Saint Antoine Hospital, Paris, France

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.

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FRI0212

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

S. Monti¹, L. Dagna², C. Campochiaro², A. Tomelleri², G. Zanframundo¹, C. Klersy³, F. Muratore⁴, L. Boiardi⁴, R. Padoan⁵, M. Felicetti⁵, F. Schiavon⁶, M. Bond⁷, A. Berti⁷, R. Bortolotti⁷, C. Nannini⁸, F. Cantini⁸, A. Giollo⁹, E. Conticini¹⁰, A. Gattamelata¹¹, R. Priori¹¹, L. Quartuccio¹², E. Treppo¹², G. Emmi¹³, M. Finocchi¹³, G. Cassone¹⁴, A. Hoxha¹⁵, R. Foti¹⁶, M. Colaci¹⁶, R. Caporali¹⁷, C. Salvarani¹⁸, C. Montecucco¹ on behalf of Italian Society of Rheumatology Vasculitis Study Group. ¹Pavia, Rheumatology, IRCCS Fondazione Policlinico S. Matteo, Pavia, Italy; ²Milan, Immunology, Vita-Salute San Raffaele University, Milan, Italy; ³Pavia, Biometry and Clinical Epidemiology, IRCCS Fondazione Policlinico S. Matteo, Pavia, Italy; ⁴Reggio Emilia, Rheumatology Unit, AUSL-IRCCS di Reggio Emilia, Reggio Emilia, Italy; ⁵Padova, Rheumatology, Padova, Italy; ⁶Padua, Rheumatology, Modena, Italy; ⁷Trento, Rheumatology, Trento, Italy; ⁸Prato, Rheumatology, Prato, Italy; ⁹Verona, Rheumatology, Verona, Italy; ¹⁰Siena, Rheumatology, Siena, Italy; ¹¹Rome, Policlinico Umberto I, Rome, Italy; ¹²Udine, Rheumatology, Udine, Italy; ¹³Florence, Immunology, Florence, Italy; ¹⁴Modena, Rheumatology, Modena, Italy; ¹⁵Vicenza, Ospedale San Bortolo, Vicenza, Italy; ¹⁶Catania, Rheumatology, Catania, Italy; ¹⁷Milan, Rheumatology, Ist. Pini, Milan, Italy; ¹⁸Reggio Emilia, Rheumatology Unit, AUSL-IRCCS di Reggio Emilia, University of Modena and Reggio Emilia, Reggio Emilia, Italy

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 ± 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