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SAT0224

PRESENTATION, MANAGEMENT AND OUTCOME OF ANCA ASSOCIATED VASCULITIDES IN ITALY: A 20-YEAR FOLLOW-UP STUDY

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Background: ANCA associated vasculitis (AAV) are systemic diseases with a wide spectrum of clinical presentation and organ involvement.

Objectives: To analyze presentation, management and 5 years-outcomes of AAV patients diagnosed between 2000 and 2018 in the monocentric cohort of Padova Vasculitis Center.

Methods: We retrospectively collected all AAV patients diagnosed between 2000 and 2018 and followed in the Vasculitis Center of Padova University. We focused on demographic and clinical features at baseline, first line immunosuppressive (IS) treatment, 6 months mortality, 5 years-relapse rate and survival rate and. In the analysis, we kept separate eosinophilic granulomatosis with polyangiitis (EGPA) from granulomatosis with polyangiitis (GPA) and microscopic polyangiitis (MPA).

Results: We identified 171 patients (F/M 93/78) with AAV diagnosed since 2000 (61 between 2000-2010 and 110 between 2011-2018, % of increased in diagnosis rate +183%): 82 (48%) were GPA, 28 (16%) MPA and 61 (36%) EGPA. The patients were mostly Caucasian (98%), with a mean age at diagnosis of 58.4±16.1 years, slightly younger if diagnosed in the second decade (2010-2018) than in first decade (2000-2010) (59.5 ±16.6 years vs 61.6±14.7 years, p=0.049). ANCA were tested in 163 patients: 23% ANCA negative, 37% cANCA/PR3, 39% pANCA/MPO and 1% double positive.

GPA/MPA patients were mostly ANCA positive at diagnosis (96/110 patients), in particular we counted 12 (15%) GPA-ANCA negative patients, 55 (68%) GPA-PR3, 13 (16%) GPA-MPO, 1 GPA with double positivity while MPA resulted only ANCA positive, 22 (82%) MPA-MPO and 5 (19%) MPA-PR3. In EGPA group, only 28/54 presented with ANCA positivity, all p-ANCA/MPO specificity.

Disease severity at diagnosis was assessed only in GPA/MPA patients: 12/101 limited disease, 34 early systemic, 29 generalized and 26 severe. In particular, GPA/MPA more frequently presented with systemic symptoms (76%), ENT and lung involvement (respectively 62% and 66%). Renal vasculitis was reported in 65% of patients with a mean eGFR of 50.2±39.8 ml/min. Moreover, we registered 17 alveolar haemorrhages, 4 cardiac involvement and 3 gastrointestinal (GI) disease.

EGPA patients, instead, presented more frequently with ENT (85%) and lung involvement (96%) (mostly uncontrolled asthma and pulmonary infiltrates). 31 (54%) EGPA presented nerve involvement (4 with CNS involvement) and 9 (16%) with cardiac involvement. Interestingly, no renal or GI vasculitis was reported.

The first line IS treatment administered was: cyclophosphamide (CYC) in 66 (38%), rituximab (RTX) in 14 (8%), azathioprine (AZA) in 27 (16%), methotrexate (MTX) in 25 (14%) and mycophenolate (MMF) in only 5 (3%).

Mortality rate at 6 months in all cohort resulted of 1.2% (2 event), while the 5-years survival calculated with Kaplan Meier method was 94.3% (8 events). All deaths occurred in the GPA/MPA group. A relapse occurred in 36% of patients, with a significant higher frequency in GPA/MPA than in EGPA (46.3% vs 17%, p=0.017).

Conclusion: Our cohort is characterized by significant higher rate of AAV diagnosis in the last decade and this could reflect the increasing incidence and prevalence reported in literature¹. Interestingly, GPA diagnosis, especially with cANCA/PR3 specificity, was prevalent in our cohort despite some authors reported an higher prevalence of MPA in Mediterranean area¹⁻². Finally, EGPA confirmed a better outcome at last follow up than GPA and MPA, but we noted that no renal and GI involvement was observed in our EGPA patients.

REFERENCE

- [1] Berti A, Dejaco C. Update on the epidemiology, risk factors, and outcomes of systemic vasculitides. *Best Pract Res Clin Rheumatol.* 2018;32(2):271-294.
- [2] Romero-Gómez C, et al. Epidemiological study of primary systemic vasculitides among adults in southern Spain and review of the main epidemiological studies. *Clin Exp Rheumatol.* 2015;33(2 Suppl 89):S-11-8

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SAT0225

TARGETED SERUM METABOLIC PROFILE IN PATIENTS WITH TAKAYASU ARTERITIS

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Background: Takayasu arteritis (TAK) is a chronic inflammatory arteritis that mainly affects aorta and, its major branches. Often the course is unpredictable and it remains a challenge to assess disease activity despite the availability of acute phase reactants and imaging modalities [1]. Management heavily relies on physician global assessment. Analysis of serum metabolites using nuclear magnetic resonance spectroscopy is a promising exploratory approach towards identifying new biomarkers in TAK which may cast some light on disease pathogenesis and disease activity.

Objectives: Aim of the study was to evaluate the performance of serum amino acids as determinants of disease activity in TAK.

Methods: Patients with TAK fulfilling ACR 1990 criteria were enrolled and disease activity assessed using ITAS2010 and ITAS-A[2]. Samples were analysed using 1D 1H 800 MHz Nuclear magnetic resonance spectrometer equipped with a Cryoprobe (at 300 K). The spectra were processed and the concentration of amino acids were measured with respect to internal reference metabolite formate assuming its concentration 10 µM in commercial software program CHENOMX (www.chenomx.com) and the resulted normalized values were compared using Tukey's multiple comparison test and diagnostic potential was evaluated using receiver operating characteristic (ROC) curve analysis.

Results: 45 active TAK patients with ITAS-A ≥ 4 and 53 inactive TA patients with median age 27 years [IQR, 22-35 and IQR, 23-37 years respectively] were enrolled. Female to male (F:M) ratio was 3.5:1 and 4.9:1 and median duration of illness 5 [IQR, 2-9 years] and 3 [IQR, 1-6 years] years in active and inactive group respectively. Majority had class V disease. 43 individuals served as health control (HC) with mean age 30 years and F:M ratio of 4.3:1. Serum levels of Phenylalanine (Phe), Tyrosine (Tyr), Histidine (His), Valine (Val) and Alanine (Ala) were found to be decreased in TAK patients suggesting augmented utilization in the immune-metabolic pathways mainly to replenish the energy demand and self-repair mechanism under conditions of inflammation and oxidative stress. An analysis of amino acids ratio revealed significantly higher His/Tyr, Ala/Tyr and Val/Tyr ratio in HC when compared to active TAK patients. Ratio were lower in active TAK when compared to inactive TAK patients but only Val/Tyr ratio was significant (AUROC, 0.64 and p, 0.01). Ratio served as a gradient with lowest value in active disease and gradually increasing and moving towards HC as disease was getting controlled

Figure 1: Comparison of various amino acid ratio in active and inactive TAK patient with respect to healthy control. Ratio served as a gradient with lowest in active disease and gradually moving towards HC as disease was getting controlled.

*significant p value; Ala, alanine; His, Histidine; NC, Normal healthy control; Tyr, tyrosine; Val, Valine