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AB0593 ANCA-ASSOCIATED VASCULITIS WITH BOTH MPO-ANCA AND PR3-ANCA SHARES CHARACTERISTICS OF ANCA-ASSOCIATED VASCULITIS WITH SINGLE ANCA

S.M. Kim[1], J. Kim[2], S.W. Kang[2], S.-C. Shim[2], S.-J. Yoo[3]. *[1]Internal Medicine, Chungbuk National University Hospital, Cheong-ju; [2]Internal Medicine, School of Medicine Chungnam National University; [3]Internal Medicine, Chungnam National University Hospital, Daejeon, Korea, Republic Of*

Background: The anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides are heterogeneous group of necrotizing inflammation of small vessel and the presence of the ANCA. ANCAs are defined according to the target antigens, leukocyte proteinase 3 (PR3) and myeloperoxidase (MPO). Recently, the ANCA specificity could be better for classification of ANCA-associated vasculitides than the clinical diagnosis. A few patients have both MPO- and PR3 ANCA. However, the clinical characteristics of these patients were not known in detail.
Objectives: To analyze organ involvement of patients with ANCA-associated vasculitis according to ANCA type focusing both MPO- and PR3-ANCA (both-ANCA) positive vasculitis
Methods: The medical records of the patients with positive ANCA and clinical diagnosis or the patients with positive ANCA and vasculitis diagnosis confirmed by biopsy were reviewed at two regional tertiary hospitals. The age at diagnosis, sex, and the organ involvement of kidney, lung, upper airway (nose/sinus/ear), skin, peripheral nervous system, central nervous system, and gastrointestinal tract were collected. The clinical variables were analyzed by ANCA type.
Results: Total 82 patients with positive ANCA and clinical diagnosis or histologic diagnosis of vasculitis were searched. MPO-ANCA positive patients was 63 (76.8%), PR3-ANCA 9 (11.0%), and both MPO- and PR3-ANCA was 10 (12.2%). The age at diagnosis of patients with PR3-ANCA was younger than patients with MPO-ANCA or both-ANCA (PR3-ANCA, 49.6 vs. MPO-ANCA, 66.1 vs. both-ANCA, 62.1, $p<0.05$). Moreover, kidney involvement were MPO-ANCA was 77.8%, PR3-ANCA 22.2%, and both-ANCA 80% ($p<0.05$). Upper airway involvement was also significantly associated with ANCA type (PR3-ANCA, 66.7% vs. MPO-ANCA, 23.8% vs. both-ANCA, 50.0%, $p<0.05$). The involvement of skin, central or peripheral nervous system, gastrointestinal tract or the presence of lung fibrosis and lung nodule or mass did not differ according to ANCA type.
Conclusions: ANCA-associated vasculitis with both MPO-ANCA and PR3-ANCA has more kidney involvement than ANCA-associated vasculitis with PR3-ANCA and more upper airway involvement than ANCA-associated vasculitis with MPO-ANCA.
Disclosure of Interest: None declared
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AB0594 RELATION OF CERTAIN BLOOD PICTURE PARAMETERS AND VASCULAR ENDOTHELIAL GROWTH FACTOR TO CLINICAL MANIFESTATIONS AND DISEASE ACTIVITY IN BEHÇET'S DISEASE PATIENTS

T.A. Gheita[1], R.E. Rabea[1], B.R. Sakr[1], S.M. Abd ElHamid[2]. *[1]Rheumatology and Clinical Immunology; [2]Clinical Pathology, Faculty of Medicine - Kasr Al-Ainy School of Medicine - Cairo University, Cairo, Egypt*

Background: Behçet's disease (BD) is a systemic inflammatory condition sharing the clinical features of both auto- inflammatory diseases and variable vessel vasculitis. Endothelial dysfunction (ED) plays an important role in the pathogenesis of BD. Several markers can be used to evaluate ED as mean platelet volume (MPV), red cell distribution width (RDW), neutrophil-lymphocyte ratio (NLR) and vascular endothelial growth factor (VEGF).
Objectives: The aim of the present study was to measure the MPV, RDW, NLR and serum level of VEGF in BD patients and to study their relation with disease manifestations and activity.
Methods: Ninety six BD patients and 60 matched controls were enrolled in this study. The MPV, NLR, RDW and serum VEGF level were measured. Disease activity was assessed by the BD Current Activity Form (BDCAF). The influence of an associated metabolic syndrome (MetS) was also considered.
Results: The mean age of the 96 patients was 34.9±10.1 years (18–73 years), male:female 4.7:1 and the disease duration was 9±7 years (0.6–40 years). MetS was present in 13.7%. Two patients were siblings and 5 had juvenile-onset BD. The RDW and NLR were significantly higher in patients (15.5±2% and 2.7±2.9) than controls (14.3±1.03% and 1.5±0.8) ($p<0.001$ each), while the MPV and VEGF were comparable. The MPV was significantly decreased in patients with vascular involvement (p=0.04) and increased in those with psychiatric disorders (p=0.02). The RDW was significantly higher in patients with vascular involvement (p=0.04) especially those with venous thrombosis and in those with neurological manifestations (p=0.03). The NLR was higher in males (p=0.01) and in those with retinal vasculitis (p=0.03) and vein occlusion (p=0.02). None of the parameters significantly correlated with the BDCAF. However, the NLR was the most valuable parameter to predict disease activity at a cut-off level of 1.69 ng/L (sensitivity 75%, specificity 55.6%). The MPV significantly correlated with the body mass index (BMI) (p=0.008), cholesterol (p=0.01) and low density lipoprotein (LDL) ($p<0.001$); the RDW correlated with the erythrocyte sedimentation rate (ESR) (p=0.003) and total leucocytic count (TLC) (p=0.04); the NLR with TLC (p=0.001) and blood urea (p=0.001) and VEGF with the TLC (p=0.048) and high density lipoprotein (HDL) (p=0.02). None of the parameters was significantly different according to the presence of MetS. Regarding the medications received, the RDW was significantly higher in patients who received cyclophosphamide and warfarin than those who did not (p=0.003 and $p<0.001$ respectively) and the level of VEGF tended to be lower in patients who received colchicine (p=0.06).
Conclusions: In BD, only the RDW and NLR were significantly increased raising the possibility of a potential role in the disease susceptibility and pathogenesis with no obvious relation to the disease activity or to an associated MetS. Together with the serum VEGF, they may all serve as useful markers to reveal the pattern of organ involvement while the NLR was the most valuable cost effective parameter to signify the disease activity. The influence of medications warrants further studies.
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AB0595 NOVEL APPROACHS BASED IN CURRENT EVIDENCE IN ANCA-ASSOCIATED VASCULITIS WITH RENAL INVOLVEMENT TREATMENT

V. Vazquez[1], G. Gonzalez[2], J. Robaina Sindin[2]. *[1]Nephrology, Hospital Simplemente Evita, Gonzalez Catan, Pcia Buenos Aires; [2]Nephrology, Hospital de Clínicas José de San Martín, Universidad de Buenos Aires, Ciudad Autonoma de Buenos Aires, Argentina*

Background: ANCA-associated vasculits (AAV) are a group of multi-system autoinmune diseases characterized by inflamation and necrosis in small and medium vessels.AAV could respond to diferent therapeutics protocols depends on diverse levels of clinical severity and early treatment could improve the outcome of the disease.In spite of recognized efficay of regimens consisting of cyclophosphamide and high-dose corticosteroids to control the AAV,efforts to minimize drugs-related toxicity led to consider targeted therapies.
Objectives: Considering the currently quality evidence in therapies novely proposed to AAV and the severity of renal disease presentation,we suggest new rational approaches emphasizing targeting B-cells therapy and preventing disease relapse
Methods: We identified the latest quality evidence using methodological search filters,assessed the evidence quality with Cochrane Renal Goup check list and determinated the strenght of recommendations by Levels of Evidence (Oxford Centre for Evidence-based Medicine)
Results: Rituximab (RTX), a monoclonal anti-CD 20antibody, has emerged as the biologic agent more using in AAV patients in current publications and unlike latest Guides and Recommendations published,RTX would be recommended in induction and maintenance AAV with renal involvement treatment (Table 1).

Table 1

AAV Induction Therapy	Early systemic (GF>60 ml'):MTX + GC (Ib,B) Severe generalised (Cr>5.68 mg/dl):CFM IV/PO + MPS (Ia,A) Generalised with contraindication to CFM:RTX + GC (Ib,B) Prophylaxis against *Pneumocystis jirovecii* (in CFM or RTX therapy):Cotrimoxazol PO (IIb,B) Severe with RPGN: Plasma Exchange-adjuvant therapy (Ia, B)
AAV Maintenance Therapy	Low-dose GC + AZA up 18 months (IIb,B) Low-dose GC + LF (less safer) (Ib,B) Low-dose GC + MTX with GF >60 ml/'(Ib,B) Avoid use CFM long-term (higher relapse risk) (Ia,A) In GPA:RTX + GC low-dose each 6 months up18 months (Ib, B)
AAV Relapse	Minor Relapse:increase GC dose (IIb,C) Major Relapse:RTX + GC (Ia,A) Major Relapse with CFM cumulative dose <36 gr:CFM + GC (Ib,B) Plasma Exchange and/or MPS (IIb,C)
VAA Refractaria	RTX + GC,specially patients whose never recived RTX (II,B) Plasma Exchange in RPGN and/or dialysis-dependen (Ia,B)

GF: glomerular filtrate, MTX:metotrexate, GV: glucocorticoids, Cr: creatinine, CFM: ciclophosphamide, IV: intravenous, PO: oral, MPS: metilprednisolone, RTX: rituximab, RPGN: rapidly progressive glomerulonephritis, AZA: azathioprine, LF: leflunomide, GPA: granulomatosis with polyangiitis.

Conclusions: Current therapeutical protocols for AVV with renal involvement show that emerging therapies like RTX could improve rates of relapses and treatment-related toxicity. Further studies would provide target-therapeutical options
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AB0596 INTERFERON-Γ RELEASE ASSAY (T-SPOT.TB) IN THE DIAGNOSIS OF TUBERCULOSIS INFECTION IN PATIENTS WITH BEHCET'S DISEASE: A SINGLE CENTER EXPERIENCE IN CHINA

W. Zheng[1], X. Wu[2], P. Chen[3], L. Zhao[1], J. Liu[1], L. Zhang[4], X. Liu[4], Y. Zhao[1], X. Zeng[1]. *[1]Rheumatology, Peking Union Medical College Hospital, BEIJING; [2]Rheumatology, General Hospital of Tianjin Medical University, Tianjing; [3]Rheumatology, Affiliated Mindong Hospital of Fujian Medical University, Fujian; [4]Infectious Diseases, Peking Union Medical College Hospital, BEIJING, China*

Background: In many aspects Behcet's disease (BD) is correlated with tuberculosis (TB). They may mimic each other on clinical manifestations. For example erythema nodosa, arthritis, gut lesions could be occurred in both disease and it is difficult for differential diagnosis. Some patients with newly-diagnosed BD were found to have active or latent or old TB and anti-TB therapy could relieve some of the symptoms. In etiology, it has been postulated that tubercle bacilli may act as a trigger of BD through the mechanism of molecular mimicking. Vice versa, defective cell-mediated immunity in BD patients may increase individual susceptibility of TB.It is extremely important to clarify TB existence or not in BD patients. Since BD patients commonly need glucocorticoid and immunosuppressant or even biologic agent such as anti-TNFa. And these medications may increase the risk of occurrence or flare of TB. But positive result of PPD shows little value in this special population due to acupuncture reaction in BD patients.In this pilot study, the values of interferon-γ release assay (T-SPOT-TB) in diagnosing active TB in BD patients were explored.
Objectives: To investigate the diagnostic value of the T-SPOT.TB in BD patients complicated with tuberculosis infection.
Methods: The clinical, radiology and laboratory data were collected and analyzed in 175 hospitalized BD patients from the Peking Union Medical College Hospital between January 2010 and March 2015. Statistical analysis was carried out using IBM SPSS version 20.
Results: Of the 175 BD patients, the positive rate of tuberculosis infection in BD patients was 34.3%. The sensitivity, specificity, positive predictive value (PPV) and negative predictive value (NPV) of the T-SPOT.TB test for the diagnosis of ATB were 87.5%, 73%, 36.8%, 98.3%, respectively. Positive likelihood ratio (PLR) and negative likelihood ratio (NLR) were 3.24 and 0.17. The median number of SFCs in the BD-ATB group was higher than that in the BD-LTB group and BD-OTB ($p<0.001$ and p=0.012). By ROC method, it was suggested that 70 SFCs act as a cutoff for diagnosing BD-ATB with the sensitivity, specificity, PPV, NPV, PLR and NLR were 87.5%, 85%, 24.6%, 98.5%, 5.79 and 0.15, respectively. The rates of LTB infection had no significant difference between BD patients and healthy controls. Agreement between T-SPOT.TB and TST in BD patients measured by the kappa coefficient was poor (kappa =0.37).Multiple logistic regression analysis revealed that BD patients with positive T-SPOT.TB had the highest likelihood of ATB (OR 11.93, 95% CI 2.108–67.508, p=0.005).
Conclusions: T-SPOT.TB is a promising test in the diagnosis of BD complicated with tuberculosis infection, and higher number of SFCs may has a higher risk of ATB.
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AB0597 LOW-DOSE RITUXIMAB AS INDUCTION THERAPY FOR JAPANESE PATIENTS WITH ANCA-ASSOCIATED VASCULITIS

Y. Takakuwa, H. Hanaoka, T. Kiyokawa, K. Ishimori, H. Iida, H. Nagafuchi, T. Okazaki, S. Ozaki. *Division of Rheumatology and Allergy, Department of Internal Medicine, St. Marianna University School of Medicine, Kawasaki, Japan*

Background: Four times of once-weekly doses of 375 mg/m^2 rituximab (RTX) are frequently used as remission induction therapy for ANCA-associated vasculitis (AAV). Since this regimen has been basically generated from experience of B-cell non-Hodgkins' lymphoma in the Europe and North America, appropriate dose and interval for patients with AAV in other population have been poorly investigated. Here we comprehensively analyzed the efficacy and tolerability of high or low dose regimen of RTX in Japanese patients with AAV.
Objectives: We investigated the efficacy and safety issue of low dose RTX for Japanese patients with AAV.
Methods: We retrospectively examined AAV patients who met the 2012 Chapel Hill classification from 2006 to 2016. We divided them into 2 groups, those treated with high-dose (HD) and low-dose (LD) RTX. HD RTX was the original regimen and LD RTX consisted of twice of one-weekly dose of 375 mg/m^2. We evaluated cumulative complete remission (CR) rate and relapse-free rate for 1.5 years. CR was defined as BVAS=0 and relapse was defined as BVAS$\geqq$1.
Results: We evaluated 17 patients with HD and 11 patients with LD RTX. Higher percentage of elderly patients was observed in LD group ($p<0.01$). No significant difference was found in BVAS (p=0.49) and VDI (p=0.15) before treatment. No significant difference was found in cumulative CR rate (p=0.90) (Fig. 1A), relapse-free rate (p=0.48) (Fig. 1B), B cells counts and serious adverse events. We found patients with nasal involvement and pulmonary nodule/cavity formation had higher relapse rate in LD group than those with HD group (p=0.05,and p=0.09 respectively).

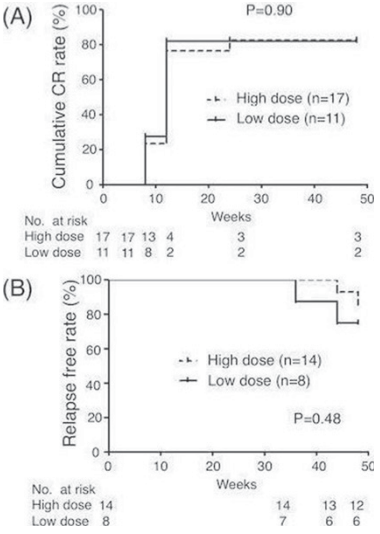

Figure 1

Conclusions: LD regimen of RTX, especially in elderly patients, might be effective in remission induction therapy in AAV.
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Scleroderma, myositis and related syndromes

AB0598 SERUM DEFENSIN LEVEL IN SYSTEMIC SCLEROSIS PATIENTS

T. Emiroglu[1], H. Küçük[2], B. Goker[2], S. Haznedaroglu[2], H. Pasaoglu[3], Ö. Varan[2], M.A. Öztürk[2], O.T. Pasaoglu[3], A. Tufan[2]. *[1]Department of Internal Medicine; [2]Department of Internal Medicine, Division of Rheumatology; [3]Department of Biochemistry, Gazi University Faculty of Medicine, Ankara, Turkey*

Background: Skleroderma is an autoimmune disease characterized by fibrosis of skin and lung as well as involvement of kidney, gastrointestinal system and heart (1). Etiology and exact mechanism of disease is poorly understood. A small number of studies have examined the role of AMPs on autoimmune diseases. It has been demonstrated that the amount of alfa- and beta-2 defensin serum levels are increased in systemic lupus erythematosus patients (2.3). Likewise, the association between AMPs and other diseases such as idiopathic pulmonary fibrosis, diffuse panbronchiolitis, pulmoner alveolar proteinosis and psoriasis has been reported (4).
Objectives: No study investigated the role of AMPs on scleroderma patients. Hence, we aimed to investigate AMP serum levels and their possible association in these patients.
Methods: There were 42 patients (40 female, mean age 42 years) and 38 healthy subjects (32 female, mean age 38 years) in the study. For SSc patients, the following data were recorded at enrollment: disease subset (limited/diffuse), autoantibodies (antinuclear, anti-centromere (ACA), and anti-scl), blood tests, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP), modified Rodnan skin score, presence and history of digital ulcers, presence and history of involvement kidney and gastrointestinal system, interstitial lung disease detected by chest HRCT and pulmonary function tests, estimated pulmonary arterial systolic pressure at echocardiography.
Results: There were 42 patients (40 female, mean age 42 years) and 38 healthy subjects (32 female, mean age 38 years) in the study. Twenty-nine of the patients had diffuse systemic sclerosis and thirteen of the patients had limited systemic sclerosis. Average disease duration is 5.5 years. Pulmonary involvement was detected in twenty patients. The levels of beta 1 and beta 2 defensin that are epitelyal defensins were higher than control group but it has not reached statistical significance. (beta-1 defensin 235±178 vs 185±24 pg/ml, p=0.08 and beta2 defensin 253±453 vs 152±101 pg/ml, p=0,1). Alpha defensin levels in scleroderma patients were significantly higher than control group (563±415 vs 377±269 ng/mL,p=0.02). In sub-group analysis patients with interstitial lung disease had a higher level of alpha defensive than those without involvement (684±473 vs430±299 ng/ml, p=0.04). There was an negative correlation between alfa defensin and Rodnan score (r=0.30) and CRP (r=0.34).
Conclusions: Alpha defensin levels in scleroderma patients were significantly higher than control group, There may be an increase in the level of alpha defensin in a cause of vasulopathy in scleroderma patients