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

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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.
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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

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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

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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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