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AB0482 INFLUENCES OF TIME OF INTRODUCTION OF INFLIXIMAB ON THE FUNCTIONAL DISABILITY AND JOB STATUS OF PATIENTS WITH CHRONIC PROGRESSIVE NEURO-BEHCET'S DISEASE

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Background: Chronic progressive neuro-Behcet's disease (CPNBD) is characterized by progressive neurobehaviour changes leading to disability and death. It has been appreciated that methotrexate is effective for CPNBD. Notably, recent studies have demonstrated that infliximab is effective for patients with CPNBD who had inadequate responses to methotrexate. However, the appropriate timing for introduction of infliximab remains unclear.

Objectives: The current studies examined the effects of intervals before introduction of infliximab on the functional disability and job status of patients with CPNBD.

Methods: Eleven patients (8 males, 3 females, ages 35.2±9.3 [mean±SD]), who met the international classification criteria for BD with CPNBD and received infliximab, were retrospectively followed up. The functional disability of the patients was evaluated by Steinbrocker functional classification as is used in rheumatoid arthritis. Correlation between the patients' functional outcome and the intervals before the introduction of infliximab was analyzed by Spearman's rank correlation test.

Results: All the 11 patients had received methotrexate prior to infliximab. The intervals from the onset to the introduction of infliximab and the follow-up periods were 26.6±35.1 months and 65.2±43.6 months [mean±SD], respectively. Among the 11 patients, 9 patients did not show progression after the introduction of infliximab, whereas 2 patients still progressed and lost job. In the latter 2 patients, infliximab had been discontinued before the final follow-up. No patients improved from the functional disability or gained job even after infliximab treatment. The functional disability grades of the patients after the introduction of infliximab were significantly correlated with the intervals from the onset of CPNBD to the introduction of infliximab (r=0.6177, p=0.0476).

Conclusion: The results indicate that the delay of the introduction of infliximab leads to the irreversible functional disability and job loss of the patients with CPNBD. Thus, it is recommended that infliximab should be administered as soon as possible for the patients with CPNBD with inadequate response to methotrexate.

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AB0483 INTERSTITIAL LUNG DISEASE IN PATIENTS WITH ANCA ASSOCIATED VASCULITIS – A PROSPECTIVE SINGLE CENTER STUDY

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Background: Recently, an association between anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV) and interstitial lung disease (ILD) has been uncovered.

Objectives: To determine the rate of ILD in our prospective AAV patient cohort and to compare clinical characteristics of AAV patients with and without associated ILD.

Methods: We retrospectively analysed medical records of prospectively diagnosed and followed AAV patients at our secondary/tertiary rheumatology centre between January 2010 and December 2019. The diagnosis of ILD was based on lung HRCT findings.

Results: During the 10-year observation, we identified 94 incipient AAV patients (46 had granulomatosis with polyangiitis, and 48 microscopic polyangiitis). Thirteen (13.8%) patients had ILD (ILD-AAV group). 12/13 had usual interstitial pneumonia (UIP) pattern and 1/13 non-specific fibrosis on HRCT. ILD was diagnosed in tandem with AAV in 9/13 patients, and 9 months to 5 years prior to AAV in 4/17 patients. Characteristics of ILD-AAV, and non-ILD-AAV groups are presented in Table 1. ILD-AAV patients more commonly reported of weight loss, less frequently had ENT involvement, and were predominantly a-MPO ANCA positive (92.3%). Follow up data were available for 85 AAV patients (90.4%; 13 ILD-AAV and 72 non-ILD-AAV). During the median (IQR) follow up of 22.1 (4.8; 50.0) months, 5/13 (38.5%) ILD-AAV patients died, compared to 6 (8.3%) deaths registered in non-ILD-AAV group during 26.4 (11.6; 70.0) months of follow up. The crude mortality rate evaluated by Cox proportional hazards regression was significantly higher for AAV-ILD group (HR 5.6 (95%CI 1.7-18.7), p=0.005).

Table 1. Clinical characteristics of ILD-AAV and non-ILD-AAV group

Characteristic	ILD-AAV (13)	non-ILD-AAV (81)	p	Characteristic	ILD-AAV (13)	non-ILD-AAV (81)	p
Female	46.2	64.2	0.234	ENT	0	60.5	<0.001
Age*	76 (67;77)	66 (55;77)	0.174	Heart	0	7.4	0.591
Smoking	61.5	39.5	0.226	GI tract	15.4	7.4	0.305
Fever	61.5	53.1	0.766	Kidney	53.8	63.0	0.552
Weight loss	84.6	51.9	0.035	PNS	38.5	29.6	0.531
Arthritis	15.4	14.8	1.0	CNS	0	2.5	1.0
Myalgia	15.4	27.2	0.504	ANCA	100	91.4	0.588
Skin	7.7	19.8	0.451	a-MPO	92.3	44.4	0.002
Eye	0	24.7	0.063	a-PR3	7.7	46.9	0.013

Legend: * median (IQR)); ENT ear-nose-throat; GI gastrointestinal tract; PNS peripheral nervous system; CNS central nervous system;

Conclusion: In our incipient AAV cohort 13% of patients presented with ILD. The AAV patients with ILD had a higher mortality rate than the rest of the cohort.

References:

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AB0484 PROTEINASE 3–ANTINEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)–POSITIVE AND ANCA–NEGATIVE OR MYELOPEROXIDASE–ANCA–POSITIVE PATIENTS WITH GRANULOMATOSIS WITH POLYANGIITIS: DISTINCT PATIENT SUBSETS

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Background: Most patients with clinical diagnoses of Granulomatosis with polyangiitis (GPA) are proteinase 3 (PR3)-ANCA positive, but a significant minority are myeloperoxidase (MPO)-ANCA positive or are negative for ANCA [1]. Several clinical and genome-wide association studies have suggested that classification based on ANCA type, i.e., PR3-ANCA positivity as opposed to MPO-ANCA positivity, may be more relevant clinically than the traditional classification based on specific diagnosis [2].

Objectives: To analyze demographic feature, disease manifestations and laboratory findings of patients with PR3-ANCA positive GPA in comparison with patients ANCA-negative or MPO- ANCA positive GPA.
Methods: This is a retrospective analysis of 37 patients with GPA from a single center in Ukraine observed from 2010 till the end of 2019. The clinical and demographic data, initial Birmingham vasculitis activity score (BVAS/WG), erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were compared between patients with PR3-ANCA positive GPA and ANCA-negative or MPO-ANCA positive GPA.
Results: Of the 37 patients analyzed, 24 (64.9%) had PR3-ANCA–positive GPA, 6 (16.2%) had MPO-ANCA–positive GPA and 7 (18.9%) had ANCA-negative GPA. ANCA–negative GPA patients were younger at diagnosis compared to PR3-ANCA–positive and MPO-ANCA-positive patients (36 versus 47 and 49 years; p = 0.04). The gender ratio was similar in patients with PR3-ANCA–positive GPA and patients with MPO-ANCA–positive GPA or ANCA-negative GPA (33% vs 38% male, p= 0.61). The ocular manifestations - conjunctivitis/episcleritis (15% vs 50%) and ear involvement - otitis, mastoiditis (0% vs 33%) occurred more often in patients with PR3-ANCA–positive GPA (p<0.05), whereas sensory peripheral neuropathy (54 % vs 21%) and Raynaud's syndrome (31 % vs 0%) were more frequent in compared group (p<0.05). ANCA-negative patients with GPA had lower, but no significant, initial BVAS/WG score than PR3-ANCA–positive or MPO-ANCA-positive patients with GPA (17.9 versus 23.5 and 24.8; p=0.20). There were no significant differences between groups in ESR or CRP levels and in the frequency of involvement of other organs and systems.
Conclusion: We demonstrate clinical differences between PR3-ANCA–positive patients with GPA and MPO-ANCA–positive or ANCA-negative patients with GPA. The eye and ear involvement are common for patients with PR3-ANCA–positive GPA. The MPO-ANCA–positive GPA or ANCA-negative GPA is characterized by higher frequency of sensory peripheral neuropathy and Raynaud's syndrome.
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AB0485 INVESTIGATION OF PERMANENT ORGAN DAMAGE IN GIANT CELL ARTERITIS: DISEASE FLARES ARE ASSOCIATED WITH INCREASED DAMAGE SCORES

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Background: Development of organ damage is a major concern in patients with systemic vasculitis. Treatment may also contribute to this important outcome. Scoring systems has been developed to evaluate organ damage in systemic vasculitis and specifically for large vessel vasculitis (1).
Objectives: We aimed to investigate permanent organ damage and determining factors in our giant cell arteritis GCA cohort.
Methods: Organ damage detected at the time of diagnosis and / or follow-up and irreversible for at least 3 months in GCA patients followed up between 1998-2018 were recorded by using Vasculitis Damage Index (VDI) and Vascular Vasculitis Damage Index (LVVID) fom patient records of our vasculitis clinic. In the statistical evaluation, chi-square, students t-test and logistic regression analysis were used.
Results: Eighty-nine patients (64% women, mean age 67.9 ± 9.1) included in the study, the mean follow-up duration was 61.6 ± 58.6 months. All organ damage findings according to both VDI and LVVID are shown in table-1. In this cohort, cardiovascular damage items and diabetes mellitus were prevalent at baseline. At least one damage item was present in 53 (59,5%) according to VDI; 54 (%60,7) according to LVVID and agreement was high between two damage indices (kappa=0.97). Forty-seven of patients (52%) had a damage item presumably with contribution of GC treatment e.g. locomotor system findings, hypertension, diabetes and cataract; 12 (13,5%) had damage items related to disease (total or partial vision loss, ischemic optical neuropathy). Mean time to diagnosis after initial symptoms was longer in patients with permanent vision loss (10,2±4,3 vs. 5,2±1,2 months p=0.006). The presence of damage was associated with flares in univariate and multivariate analysis (29/54 vs. 2/35 p<0,001 OR=19 %95 GA 4,2– 87,9). All patients who had a flare during the first year (n = 15) developed signs of damage at follow-up. No association was found between the development of organ damage and the age of diagnosis, the time between first complaint and diagnosis, presence of cranial, ophthalmologic findings, PET-CT positivity, cumulative steroid dose, and DMARD use.
Conclusion: In our study, permanent organ damage was analysed by using different indices. In this patient population baseline cardiovascular damage and diabetes mellitus were frequent as expected but information for osteoporosis was lacking. More than half of the patients had damage and significant part of the present items was considered due to corticosteroid treatment. The most common damage item developed was osteoporosis. There was a very good agreement between the two indices, despite few specific items in LVVID. The striking relationship of disease flare with damage and frequency of visual problems despite treatment indicate the necessity of new treatment strategies.
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AB0486 ANALYSIS OF 89 PATIENTS WITH GIANT CELL ARTERITIS FROM TURKEY: PET-CT AS AN EMERGING METHOD FOR DIAGNOSIS AND HIGH FLARE RATE WITH STANDARD CARE

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Background: The prevalence of giant cell arteritis (GCA) in Turkey has been reported lower than other European countries and the information on clinical patterns, diagnostic modalities, treatment and prognosis of GCA are limited (1).
Objectives: We aimed to analyse our GCA cohort from a large outpatient clinic for the last 20 years.
Methods: Data of the GCA patients followed up at least for 6 months in our vasculitis clinic between 1998 and 2018 evaluated retrospectively according to EULAR 2018 GCA clinical research recommendations (2). Chi-square, students t-test, logistic regression analysis and Kaplan-Meier test were used for statistical analysis.
Results: Eighty-nine patients with adequate follow-up data (64% female, mean age 67.9 ± 9.1) were analysed. Median follow up duration was 46 months (3-256) and mean time to diagnosis after presenting symptom (TTD) was 5,9±1,2 months (0-60). Polymyalgia rheumatica was found in 36 (40.4%) patients. The clinical findings of the patients are shown in Table-1. Mean TTD was longer in patients with acute vision loss (AVL) (11±4 vs. 4,8±1,1 months p=0.002). Mean CRP was 90,7±82 (8-343) mg/L and ESR was 103,7±25 (52-138) mm/h at the time of diagnosis. Mean age was lower (63±2 vs 69±1 p=0.01); mean CRP (141,8±107,3 vs. 76,6±67,9 mg/dL p=0.023) and ESR (120,8±25,1 vs. 99,3±24,3 mm/h p=0.004) was higher in patients without cranial symptoms (extracranial GCA group). PET-CT findings compatible with large vessel vasculitis were present in 64% (34/53). Sixteen of 19 (%84,2) patients in the extracranial GCA group had positive PET-CT. Temporal artery (TA) biopsy positivity was 64% (34/53). Sensitivity of ACR 1990 Criteria was 77,5% and GIACTA study inclusion criteria was 58,4% in this cohort at diagnosis. Fullfilment of GIACTA criteria was still present in 12 (13,5%) patients after six months of follow up. Treatment data was shown in table-2. Total flare rate was 34,8% and flare risk was lower in the extracranial GCA group (3/20 vs. 28/69 p=0.035 OR=0.78 %95 CI 0.64 – 0.96). Reduced survival was observed in cases diagnosed older than 65 years (168,8±23,9 vs 209±17,3 months p=0,015).
Conclusion: The analysis of the largest single center cohort from Turkey confirmed that delayed diagnosis is associated with vision loss. A subgroup of patients without apparent cranial symptoms but positive PET-CT findings is delineated. These patients are younger, present with higher inflammatory response and fewer relapses. The sensitivity of ACR criteria in our cohort is less than 80%. High flare rate especially in GCA patients with cranial symptoms and GIACTA criteria fullfilment after 6 months of treatment in more than 10% of the patients show a need for for new treatment options.
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