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

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Background: Chronic progressive neuro-Bechet'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].