FRI0205

CLINICAL FEATURES OF MONONEURITIS MULTIPLEX ASSOCIATED WITH ANCA-ASSOCIATED VASCULITIS

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Background: ANCA-associated vasculitis sometimes presents mononeuritis multiplex which worsens the prognosis and activity of daily living in patients.

Objectives: This study aimed to determine the clinical feature of mononeuritis multiplex associated with AAV.

Methods: Consecutive patients with AAV who visited Tokyo Medical Center between April 2006 and December 2019 were included in this study. We examined the following clinical features: prevalence of neuropathy, age of onset, sex, the worst blood test values before the initial therapy (white blood cell count (WBC), eosinophil count (Eo), MPO-ANCA, PR3-ANCA, and C-reactive protein (CRP) levels), manual muscle testing (MMT) score of 20 muscles (Max 100 points), and time (days) from the initial symptoms to the initial induction therapies.

Results: A total of 89 patients with AAV were identified. Among them, 19 patients had eosinophilic granulomatosis with polyangiitis (EGPA) (8 males and 11 females, mean age 63.3 ± 2.7), 9 patients had granulomatosis with polyangiitis (GPA) (0 males, 9 females, mean age 75.6 ± 3.9), and 61 patients had microscopic polyangiitis (MPA) (17 males, 44 females, mean age 78.2 ± 1.5). Of the 89 AAV patients, 26 had sensory neuropathy (15/19 EGPA (78.9%), 11/61 MPA (18.0%), and 0/9 GPA (0%)). Motor neuropathy was observed in 19 patients (EGPA 14/19 (73.7%), MPA 5/61 (8.2%), GPA 0/9 (0%)). 15 patients had both sensory and motor neuropathies (EGPA 12/19 (63.2%), MPA 3/61 (4.9%), GPA 0 (0%)). In patients with both sensory and motor neuropathy, sensory impairment preceded in all cases. Among patients with neuropathy, the time from initial symptoms to initial induction therapy in patients with and without motor neuropathy was 34 ± 10.1 days and 30 ± 10.8 days ($p = 0.776$), respectively. Also, when comparing those who were treated within 7 days from the onset of movement disorders with those who were treated later, MMT score two weeks after the start of treatment were 92.15 ± 1.47 vs. 91.25 ± 2.65 ($p = 0.77$).

Between the patients with EGPA with and without sensory neuropathy, there were no significant differences in the following: highest WBC (19620.0 ± 2082.6 vs. 19350.0 ± 4033.5 cells/uL ($p = 0.953$)), highest Eo (10790.6 ± 1774.8 vs. 12440.8 ± 3436.9 cells/uL ($p = 0.6750$)), and highest CRP levels (4.467 ± 0.96 vs. 2.70 ± 1.85 mg/dL ($p = 0.41$)) before the initial therapy. On the other hand, comparing the EGPA patients with and without motor neuron disorder, CRP levels were significantly higher in those with motor impairment than those without ($WBC 20978.6 \pm 2049.8$ vs. 15600.0 ± 3429.9 cells/uL ($p = 0.20$); $Eo 12213.4 \pm 1775.5$ vs. 8127.0 ± 2971.0 cells/uL ($p = 0.25$); $CRP 5.13 \pm 0.89$ vs. 1.20 ± 1.48 mg/dL ($p = 0.04$)). And in patients with motor neuropathy, the decrease in MMT score was significantly correlated with the worst levels of CRP ($p = 0.001$) while the decrease was not correlated with the other blood tests. ANCA levels were not associated with sensory or motor neuropathy. In similar analyses of patients with MPA and GPA, there were no significant findings.

Conclusion: Worst CRP levels before the initial therapy can be a poor prognosis factor for motor neuropathy in patients with EGPA. Therefore, EGPA patients with high CRP levels need to be paid more attention to because of possible development of motor neuropathy.

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FRI0206

THE RELEVANCE OF SERUM IGA LEVEL MEASUREMENT IN ADULT IGA VASCULITIS

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Background:

Objectives: To evaluate the role of serum IgA level as a biomarker in adult IgA vasculitis (IgAV).

Methods: This prospective study included large cohort of histologically proven adult IgAV cases diagnosed between January 2013 and December 2019 at our secondary/tertiary rheumatology centre. All patients underwent a detailed clinical evaluation and laboratory workup. Patients were then stratified based on baseline serum IgA level into two groups (elevated serum IgA vs. normal serum IgA), and clinical features were compared between the two groups. Next we used multivariable logistic regression analysis to determine factors predicting gastrointestinal (GI) or renal involvement in adult IgAV.

Results: During the 84-month observation period, we identified 227 incipient adult IgAV cases (60.6% males, median (interquartile range) age 64 (47–76) years, 44 (19.4%) current smokers). One hundred and eleven (48.9%) patients had elevated serum IgA level at baseline, the rest had normal IgA level. None of the patients had subnormal serum IgA level. Skin involvement, constitutional symptoms, arthritis, GI tract and renal involvement developed in 227 (100%), 32 (14.1%), 30 (13.2%), 62 (27.3%), and 93 (41.0%) patients, respectively. Patients with elevated serum IgA less frequently developed constitutional symptoms ($p = 0.036$) and GI tract involvement ($p = 0.017$), but had more commonly renal involvement ($p < 0.001$), compared to those with normal serum IgA. Results of univariate analysis are shown in Table 1. In the multivariable logistic regression model, elevated serum IgA level persisted as a factor associated with lower risk of GI tract involvement (OR 0.47 (95%CI 0.23–0.95), and a higher risk of renal involvement (OR 2.71 (95%CI 1.48–4.96). The other factors associated with risk of GI and renal involvement are presented in Table 2.

Table 1. Clinical characteristic of IgAV patients with and without elevated serum IgA

Characteristic	All IgAV	Elevated s+++IgA	Normal sIgA	p value
Number of patients (%)	227	111 (48.9)	116 (51.1)	–
Males (%)	60.8	67.6	54.3	0.043
Age (years)*	64 (47-76)	58 (37-74)	66 (55-77)	0.289
Prior infection (%)	32.2	26.1	37.9	0.065
Constitutional symptoms (%)	14.1	9.0	19.0	0.036
Generalized purpura* (%)	52.0	55.0	49.1	0.426
Necrotic purpura (%)	46.3	53.2	39.7	0.046
Arthritis (%)	13.2	9.0	17.2	0.079
GI involvement (%)	27.3	19.8	34.5	0.017
Renal involvement (%)	41.0	53.2	29.3	<0.001

Legend: # median (IQR); GI gastrointestinal; * above the waistline;

Table 2. Risk factors of GI and renal involvement, multiple logistic regression

Characteristic	GI involvement OR (95%CI)	Renal involvement OR (95%CI)
Age	0.98 (0.96-1.0)	1.02 (1.00-1.04)
Current smoking	–	3.32 (1.56-7.07)
Generalized purpura*	5.86 (2.82-12.16)	2.03 (1.13-3.66)
↑ serum IgA	0.47 (0.23-0.95)	2.71 (1.48-4.96)
NLR >3.5	3.37 (1.59-7.12)	2.24 (1.19-4.23)

Legend: * purpura above the waistline; NLR neutrophil to lymphocyte ratio

Conclusion: Serum IgA level might be a useful biomarker in IgA vasculitis, identifying patients at risk for visceral (GI and renal) involvement.

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CANCER INCIDENCE AND FEATURES IN ADULT VASCULITIS PATIENTS

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Background: Vasculitis, like other diseases with chronic inflammation, may be associated with increased cancer risk; possibly due to the inflammation triggering and advancing the neoplastic process. (1-3).