AB0391

LOW SERUM COMPLEMENT C3 LEVEL AS A RISK FACTOR FOR RELAPSE OF ANTINEUTROPHIL CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS: A RETROSPECTIVE COHORT STUDY

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Background: The alternative pathway of complement activation has recently been recognized as a key pathogenic event in antineutrophil cytoplasmic antibody (ANCA)-associated vasculitis (AAV). Some previous studies have reported that low serum complement C3 level in AAV patients is associated with more severe renal disease, worse renal prognosis, or higher mortality. However, the correlation between low serum C3 level and AAV relapse remains unclear.

Objectives: To analyze the clinical characteristics and outcomes of AAV patients with low serum C3 levels at the time of diagnosis.

Methods: We conducted a retrospective observational cohort study including 83 consecutive patients diagnosed with AAV in our hospital from January 1999 to December 2020. Serum C3 levels were measured at diagnosis. AAV included microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA), and eosinophilic granulomatosis with polyangiitis (EGPA); patients with ANCA-negative AAV were excluded. Patients were divided into low- and high-C3 groups (C3 < 100 and $\geq$ 100 mg/dL, respectively). We compared the clinical characteristics, and relapse-free and overall survival rates, of the two groups, and identified predictors of AAV relapse.

Results: Of the 83 patients (MPA, n = 61; GPA, n = 18; EGPA, n = 4), 20 (24%) were in the low-C3 group. We found no significant group difference in sex, body mass index, disease type, ANCA subtype, Birmingham Vasculitis Activity Score (BVAS), or treatment. The low-C3 group patients were older (p=0.01), and had a higher Five Factor Score (FFS) (p=0.01) and a lower remission rate (p=0.02), than the high-C3 group. The generalized Wilcoxon test revealed that the relapse-free survival time was significantly shorter in the low-C3 group (29 months; 95% confidence interval [CI]: 15–49) than in the high-C3 group (82 months; 95% CI: 61–NA; p=0.01) (Figure 1A). The overall survival was also shorter in the low-C3 group (83 months; 95% CI: 8–121) than in the high-C3 group (112 months; 95% CI: 77–NA; p=0.03) (Figure 1B). In the Cox proportional hazards model, a low C3 level (< 100 mg/dL) (hazard ratio [HR], 3.01; 95% CI: 1.29–7.04, p=0.01) and GPA (HR, 3.04; 95% CI: 1.32–7.01; p=0.01) were independent predictors of AAV relapse.

Figure 1. Kaplan-Meier estimates of the relapse-free (A) and overall (B) survival rates of AAV patients by baseline serum C3 levels. Eight patients who did not show remission were excluded in the relapse-free survival analysis. Black line: high-C3 group ($\geq$ 100 mg/dL); red line: low-C3 group (< 100 mg/dL).

Conclusion: AAV patients with low C3 levels at diagnosis were at higher risk of relapse. Larger prospective studies are required to confirm these findings.

Disclosure of Interests: None declared

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AB0392

PRELIMINARY VALIDATION OF THE ITALIAN VERSION OF ANCA-ASSOCIATED VASCULITIS PATIENT-REPORTED OUTCOME (AAV-PRO_ita) QUESTIONNAIRE: FATIGUE AND CHRONIC PAIN AS UNMET NEEDS BY CURRENT TREATMENTS

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Background: The ANCA-associated vasculitis patient-reported outcome (AAV-PRO) questionnaire is a 29-item disease-specific PRO measure for AAV (1), proving to be valid, reliable, feasible, and able to discriminate among disease states.

Objectives: The purpose of this study is to validate the Italian version of AAV-PRO questionnaire (AAV-PRO_ita).

Methods: AAV-PRO has been translated in collaboration with Oxford and Bristol University (UK). Then, consecutive Italian-speaking AAV-patients were collected in order to validate the questionnaire. Inclusion criteria are AAV diagnosis (ANCA positivity at least one time and/or biopsy-proven AAV), and age $\geq$ 18 years old. Participants completed the AAV-PRO_ita three different times: at baseline, after 5–7 days, and at month 3. Friedman test was applied to verify internal consistency of the questionnaire over time.

Results: 27 AAV-patients (16 females, 11 males) were recruited and completed the questionnaire at baseline, after 5–7 days, and at month 3. The main diagnosis was granulomatosis with polyangiitis (GPA) (19/27, 70.4%), followed by eosinophilic granulomatosis with polyangiitis (EGPA) (8/27, 29.6%). Mean (SD) BVASv3 was 1 (2) at baseline, and it was unchanged at month 3 in 25/27 (92.6%). The mean age at onset of disease was 54.7 $\pm$ 17.3 years and the median disease duration was 51 months (1–3 interquartile range 34–76). Their history was characterized by at least one relapse in 10/27 (37%) patients, at least one hospitalization due to AAV in 19/27 (70.4%), and ESRD in 1/27 (3.7%). At the last follow up, 24/27 (88.9%) of patients were taking an immunosuppressive drug [rituximab (8/24), azathioprine (8/24), methotrexate (7/24) or mycophenolate mofetil (1/24)] and more than two-thirds were also taking glucocorticoids (19/27, 70.3%). Fatigue (item 9) and arthralgia (item 6) were the most important symptoms, and half of the patients (14/27, 51.9%) classified them as “moderate” or “severe”. Nearly one-third of patients (8/27, 29.6%) admitted to being “usually” or “always” concerned about their future (item 19) and long-term therapy (item 29) (Figure 1). Friedman test was not significant (p>0.05), indicating internal consistency of the questionnaire. A large multicentre Italian study has been organized based on this preliminary data coming from the coordinating centre, and it is now recruiting. The results will be soon available by the Italian Vasculitis Study Group.

Conclusion: The AAV-PRO_ita is a new disease-specific PRO measure for AAV. It is a self-administered questionnaire and could become an important tool to evaluate health-related quality of life in AAV (2). As in other systemic autoimmune diseases, fatigue and chronic pain represent an unmet need to be addressed by future treatment strategies.

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Figure 1.

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AB0393

PRE-EXISTING MORBIDITY AS A RISK FACTORS FOR MORTALITY IN IgA VASCULITIS

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Background: IgA vasculitis is generally considered to be a self-limiting condition, but this is at odds with the increased mortality observed in adult patients with IgA vasculitis (1).