Summary

Title: Comparison of ANCA-associated systemic vasculitis clinical features in patients requiring more and less intense immunosuppressive treatment.

Objective: ANCA-associated systemic vasculitis (AAV) is a group of rare diseases characterized by unpredictable course, which patogenesis was not entirely explained so far. The main objective of this study was the comparison of clinical picture between AASV patients requiring more and less intense immunosupressive treatment.

Materials and methods: Analysis involves 62 AASV patients diagnosed on the basis of actual diagnostic criteria inpatient in Department of Internal Diaseases and Rheumatology and Department of Internal Diseases and Nephorology of Central Clinical Hospital of the Ministry of the Interior and Administration in Warsaw. Case history was taken, patients were examined, BMI was calculated, data such as: blood count, CRP, ESR, AST, ALT lipids profile, glucose, uric acid, creatinine, eGFR, urine, proteinuria, serological tests (p-ANCA, c-ANCA); ECG, blood pressure, imaging studies (chest X-ray, CT of chest and paranasal sinuses, USG of abdominal cavity), histopathologic examination, assessment of disease activity using BVAS and applied treatment were analysed.

Results: 44 patients (71,0%) were diagnosed with GPA, 16 (25,8%) with EGPA and 2 (3,2%) with MPA. Examined group consisted of 39 (62,9%) women and 23 (37,1%) mem. Patients age was between 29 to 88 years old and mean age was 62,4 years old. More than half of the patients suffered from generalised form of disease (54,8%), 14 (22,6%) patients were diagnosed with severe or resistant form, early systemic (14,5%) and localized (8,1%) form were the most uncommon. Kidneys (41 patients, 61,1%) and lungs (32 patients, 54,8%) were most often involved. Lung involvement was diagnosed more often in men than women; it was also connected with higher diasease activity using BVAS score. Median od c-ANCA antibodies was twice higher in group treated more intense with immunosupressive drugs than among patients treated less intense; p-ANCA median was statistically significant higher (336 CU) among patients treated more intense than in the other group (66,7CU, p-0,011). Patients receiving more intense immunosupressive treatment were given more often clucocorticosteroids (100% vs 75%, p=0,003), metyloprednisolone i.v. (46,7% vs 8,3%, p=0,019), and cyclophosphamide (84,8% vs 0%, p<0001) in comparison to less intense treated group, which more often received MTX and azathioprine. Disease relapses were more often among patients diagnosed with severe and resistant form of disease (41,7% and 16,3% respectively, p=0,108).

Conclusions:

1. Intense immunosupressive treatment is statistically more often used in patients with kidney involvement.
2. There was no relationship between lung involvement and c-ANCA and p-ANCA average level.
3. There was higher c-ANCA level among GPA patients with kidney involvement. There was no difference between p-ANCA level among EGPA patients with kidney involvement.
4. There was no correlation between CRP, ESR, ANCA level and disease activity index using BVAS score among EGPA and MPA patients.

There is a positive correlation of c-ANCA level and BVAS score in GPA patients.
5. Patients with disease relapses were more often diagnosed with severe and resistant form of disease than other patients.