

CYTOKINES AND PROTEINS INVOLVED IN IMMUNOPATHOGENESIS OF GRANULOMATOSIS WITH POLYANGIITIS AND THEIR ASSOCIATION WITH CLINICAL FEATURES OF THE DISEASE

Dobroslav Kyurkchiev¹, Tsvetelina Yoneva², Ekaterina Ivanova-Todorova¹, Adelina Yordanova¹,
Georgi Vasilev¹, Ekaterina Kurteva¹, Rasho Rashkov²

¹*Laboratory of Clinical immunology, University hospital "St. Ivan Rilski";
Department of Clinical immunology, MF, Medical University of Sofia, Bulgaria*

²*Clinic of Rheumatology, University hospital "St. Ivan Rilski";
Department of Internal medicine, MF, Medical University of Sofia, Bulgaria*

Aim: Granulomatosis with polyangiitis (GPA) or Wegener's disease is autoimmune disorder associated with anti-neutrophil cytoplasmic antibodies (ANCA) in most cases directed against proteinase 3 (Pr3). Our aim was to investigate the serum levels of some cytokines (TNF α and IDO) and proteins (LAMP-2 and plasminogen) engaged in immunopathogenesis of GPA and to detect associations of these factors with some clinical and laboratory features of GPA.

Materials and Methods: 34 patients with GPA and 21 healthy controls were enrolled in our study. Indirect immunofluorescence for detection of ANCA, flowcytometry for detection of B cells in Rituximab treated patients and ELISA for determination of plasma levels of TNF α , IDO, LAMP-2 and plasminogen were immunological methods used in our study.

Results: Our results demonstrate elevated levels of TNF α and IDO in stage of activity of GPA detected by Birmingham vasculitis activity scale (BVAS). Only TNF α was up regulated in patients with more severe forms of the disorder according EUVAS. In patients with pulmonary engagement with granulomas increased TNF α was demonstrated as well. We found out that decreased level of serum plasminogen correlates with renal involvement and proteinuria. Our results couldn't find any association with LAMP-2 plasma levels and clinical course of the disorder.

Conclusion: The results obtained unveil part of protein secretory factors involved in immunopathogenesis of GPA as well as their potential role as diagnostic tools of some clinical features of the disease.