ANCA-associated vasculitis - is it still a diagnostic and therapeutic challenge?

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Background: The aim of the study was to describe the rare case of vasculitis with the presence of ANCA antibodies in a 36-year-old woman. ANCA associated vasculitis is a rare and heterogeneous group of diseases. It can occur at any age, but more often it affects adults and the elderly. In the course of the disease, the involvement of organs such as the lungs and kidneys is typical. It is usually manifested by progressive deterioration of renal function, which may lead to terminal renal failure. The clinical presentation is characterized by the occurrence of general, muscular-joint, neurological and skin symptoms.

The study analyzes the case of patient who has developed the above disease entity.

Case report: The disease was manifested in the form of arthritis with tenderness of the muscles of the lower legs, increasing dizziness, disturbance of gait balance, lower limb parasthesia and skin lesions with morphology of vasculitis. Acute kidney damage occurred in the course of systemic disease. A kidney biopsy confirmed the features of scantimmune glomerulonephritis with the presence of crescents. Serological tests showed the presence of c-ANCA antibodies. Plasmapheresis with simultaneous administration of Solu-Medrol was used in the treatment. Then, intravenous immunoglobulins were started in parallel, initiating Endoxan therapy according to the EUVAS regimen. After completion of remission induction treatment, maintenance therapy with Azathioprine and then Mycophenolate Mofetil was introduced. Due to the ineffectiveness of the treatment, Rituximab was used. The therapy did not bring the expected results, therefore the patient is qualified for kidney transplantation.

Conclusions: The rarity and varied clinical course of ANCA-associated vasculitis mean that they are often overlooked in differential diagnosis in patients with symptoms from multiple organs and systems. Therefore, they constitute a significant diagnostic problem. It is important that the disease is detected as soon as possible and that appropriate treatment is started to prevent complications of the described disease entity.

Key words: ANCA-associated vasculitis, acute kidney injury, immunosuppressive therapy, rituximab