**268. Central nervous system involvement in ANCA-associated vasculitis – a retrospective cohort study.**

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**Background:** Both peripheral and central nervous system (CNS) may be involved in patients with ANCA-associated vasculitis (AAV), with peripheral symptoms predominating. CNS involvement is rare symptom of AAV with reported prevalence rate of 7% to 11%.

**Methods:** The purpose of this study was to describe the clinical presentation of patients with ANCA-associated vasculitis (GPA - granulomatosis with polyangiitis and MPA - microscopic polyangiitis) presenting with CNS involvement treated in one academic center between 1988 and 2021. Medical database of 255 GPA and MPA vasculitis patients were retrospectively reviewed, and demographics, serological, and clinical features of the patients presenting CNS involvement throughout the disease course were recorded. Comparisons of disease characteristics and long-term outcomes were performed between patients with and without CNS involvement.

**Results:** 27/255 patients (10.5%) had symptoms of CNS involvement associated with the disease. All but one patient had abnormalities confirmed in MRI or CT of the head. Headache (16/28) was the main clinical symptom, followed by vestibular syndrome (11/24), deafness (7/10), motor impairment (9/24) and seizures (4/10). Three patients had psychiatric symptoms and dementia. CNS involvement was characterized by cerebral ischaemic lesions in 19, pachymeningitis in 8, and haemorrhagic lesions in 1 patient. Three patients had thrombosis of brain sinuses and 1 patient had Posterior Reversible Encephalopathy Syndrome (PRES). In 5 of them CNS changes were secondary to infiltrations from skull base. None of our patients had pituitary gland involvement. Two patients had both cerebral and spinal cord pachymeningitis. Lumbar puncture was performed only in one patient. None of the patients had brain biopsy performed. When compared with group without CNS involvement we found only two statistically important differences: patients with CNS involvement were more often cANCA positive and they had more often eye involvement.

**Conclusions**: CNS involvement is severe complication of AAV. Headaches, cANCA positivity and eye involvement are associated with this complication.

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