

CLINICAL CASE OF CONNECTIVE TISSUE AND NEURODEGENERATIVE DISEASE

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Abstract. We present a patient with 5 years Systemic lupus erythematoses (SLE), antiphospholipid syndrome (APS), dementia the type of Alzheimer and clinical symptoms of neurodegenerative disease (Parkinson's). The history of patient proceeds with progression and overlap of clinical characteristics of systemic and neurodegenerative disease. Combined treatment for systhemic and neurological disease is important for clinical improvement of these patients.

Key words: systemic disease, neurodegenerative disease.

We present a patient with 5 years Systemic lupus erythematoses (SLE), antiphospholipid syndrome (APS), dementia the type of Alzheimer and clinical symptoms of neurodegenerative disease (Parkinson's). From 2016 year patient was complaints from memory disorders, headache, numbness in the fingers, progressive stiffness and rigidity with difficulty in fine movements, discharge of objects, unsteadiness when walking, difficult to serve, pain in the metacarpophalangeal joints. During 1991-1993 sick from severe polymyositis. Patient was conducted Methylprednisolone with dose reduction, after that medication was stoped. No immunological studies. No relapses to date. Clinical examination of internal organs was normal. She was rigidly increased mus-

cle tone of left main, spasmodically-rigidly increased muscle tone of right main with dystonic phenomena. Tendonoperiosteal reflexes are alive bilateral. Gait is unsteady with dysdiadochokinesia. Tests for evaluation of cognitive functions for memory and attention show medium pronounced cognitive disfunction. Laboratory examination show hight activity SLE and antiphospholipid syndrome dsDNA 126,3 ME/ml – normal leves is up to 20 ME/ml, β 2GP1 more than 90 ME/ml – normal leves is up to 10 ME/ml.

Magnetic resonance imaging (MRI) of the brain present vascular induced leukopathic foci, ischemic zone and atrophy of brain bark (Fig. 1).

Changes are progressing under control MRI after 2 years (Fig. 2).





Fig. 1. MRI of brain of 2016 year

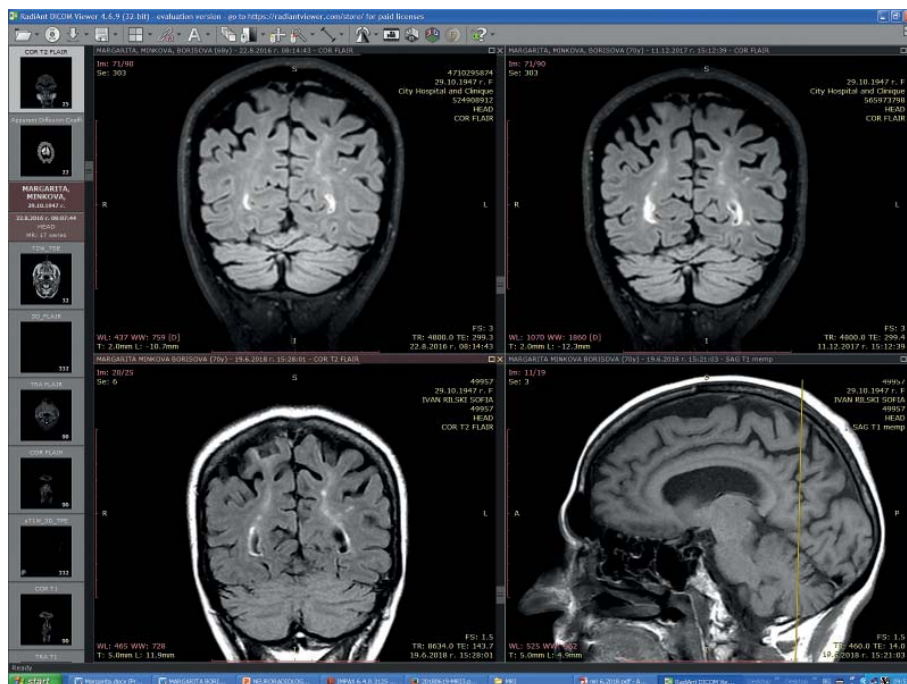


Fig. 2. MRI of brain of 2018 year

One-year cyclophosphamide pulse therapies and Immunovenin 2 cours for 90 ampules were administered in no discernible effect. This treatment was followed by chronic administration of Methylprednisolone 2 tabl daily x 4 mg, Imuran 2 ucne daily x 50 mg, Madopar 3 time daily x 250 mg, Clopidogrel 1 tabl daily.

The review of the scientific literature represents single clinical cases. Combined treatment for systemic and neurological disease is important for clinical improvement of these patients.

DISCUSSION

Systemic lupus erythematoses (SLE) is a chronic, autoimmune, multisystem connective tissue disorder. SLE is most prevalent in females, with a female:male ratio of approximately 9:1 and highest incidence between 20 and 40 years of age [1].

Clinical manifestations of SLE are likely the consequence of a multifaceted, patient-unique autoimmune process, which evolves over the course of

disease and is characterized by production of auto-antibodies, immune complex/complement-mediated tissue injury, and vasculopathy [2].

The pathophysiology of movement disorders in APS and SLE is not completely understood, and multiple mechanisms may play a role. Specific antigenic targets of APL include phospholipids, phospholipid-protein complexes and phospholipid-binding proteins to result in an intricate cascade of reactions that may contribute to clinical manifestations [3]. Classically, APL antibody – antigen binding affects coagulation to result in thrombosis. Many, although not all, people who develop movement disorders in the context of APS and SLE have ischemic-appearing lesions on neuroimaging [3-10].

Hypokinetic Movement Disorders. There are 8 cases associated with parkinsonism reported to date, 7 of which were associated with ischemic-appearing changes on MRI [5, 7-9]. Of these, 5 individuals had associated cognitive dysfunction [5, 6] and 2 had other movement disorders (dystonia and ataxia). Only one person improved with levodopa treatment, and notably, his parkinsonism occurred in the context of transient basal ganglia hyperintensities after pulmonary endarterectomy under total cardiopulmonary bypass for chronic thromboembolic pulmonary hypertension attributed to his history of APS [9].

Ataxia, myoclonus, dystonia and tremors have been described in the context of SLE. Cerebellar findings have been reported in less than 2% of people with SLE overall but in up to 25% of those who have infarcts [10].

Parkinsonism and other hypokinetic movement disorders have also been reported in the context of SLE. Parkinsonism has been described in over 30 cases and is highly associated with other neuropsychiatric symptoms [10]. The majority of people with SLE and parkinsonism have had thalamic and/or basal ganglia T2 hyperintensities on MRI. Improvement has been reported with immunosuppressive agents (ie, corticosteroids, azathioprine, and cyclophosphamide) and dopaminergic therapy (ie, levodopa and dopamine agonists). Clinical response to treatment is variable, although improvement has occurred in most cases [10].

Dementia is a clinical syndrome that encompasses a set of neurologic symptoms involving difficulties in memory, speaking, problem solv-

ing, and thinking abilities, leading to the impairments of personal and social life. Among several types of dementia, Alzheimer's disease (AD) and vascular dementia (VD) are most commonly observed. Dementia has been observed in up to 56% APS patients and a study on non-SLE patients with neurological symptoms showed that over 50% of the patients with high levels of APS developed dementia.

The management of movement disorders associated with APS and SLE is complex. Most therapeutic decisions are made based on the suspected pathophysiologic mechanism causing neurologic damage. A variety of empiric therapeutic options have been tried including immunotherapy, anticoagulation, antiplatelets, and symptomatic treatments based on movement disorder phenomenology.

Most individuals see improvement in movement symptoms with a combination of symptomatic therapies, antiplatelets, anticoagulants, and immunosuppressants. More research is needed to better understand the underlying pathophysiology of these disorders and guide the development of effective treatments.

Conflict of interest. The author declares that there is no conflict of interest related to the publication.

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