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A 50-year-old female patient with a 15-year history of epilepsy was admitted to our hospital due to generalized convulsive status epilepticus. The patient was initially hospitalized in the Intensive Care Unit and then in the Department of Neurology. The repeated magnetic resonance imaging of the head was normal, as well as the results of extensive diagnostics for infectious causes. Cerebrospinal fluid (CSF) examination showed an increased protein level and slightly elevated cytosis with lymphocyte predominance. Antinuclear antibodies ANA Hep-2 were detected in serum in titer > 1:2560 (speckled type of immunofluorescence) without specificity in the immunoblot testing. Due to the appearance of psychotic symptoms, the patient was referred for further observation to the Department of Psychiatry. During observation confusion, depressed mood, anxiety and visual and auditory hallucinations were present. Due to persistent epileptiform activity in electroencephalography (Fig.1A), the anti-epileptic treatment was modified - lamotrigine and valproic acid were started and carbamazepine was gradually discontinued. Due to persistent psychotic symptoms, antipsychotic drug (risperidone) was administered. In laboratory tests decreased levels of hemoglobin (11.1 g/dl), white blood cells (2.52 G/l), neutrocytes (0.81 G/l) and platelets (109 G/l) were observed. Considering the dynamics of the changes, the drug-induced toxicity seemed unlikely. During internist’s consultation patient’s history revealed periodic arthralgias and muscle weakness, periodic enlargement and tenderness of the cervical and submandibular lymph nodes (not related to infection), sicca symptoms, flaking skin on hands and feet. The Schirmer's Test was performed revealing impaired tear secretion (right eye - 8mm, left eye - 9mm, N>15 mm) and unstimulated whole saliva flow was significantly lowered (0.1 ml/5 min, N>0.5ml/5min). Based on the medical history and examination systemic connective tissue disease was suspected. Differential diagnosis included systemic lupus erythematosus (SLE) and primary Sjögren's syndrome (pSS). Both are chronic inflammatory autoimmune diseases, and central nervous system (CNS) involvement may occur in the disease course [1,
After histopathological evaluation of labial salivary glands, where typical lymphocytic infiltrates were present (Fig. 1B), the patient was diagnosed with pSS (ACR/EULAR criteria from 2016 were fulfilled) [3]. Lumbar puncture was performed to assess whether neuropsychiatric symptoms may result from CNS involvement in the course of pSS. In the meantime patient's condition has deteriorated - tremor of the limbs, anxiety, speech articulation problems and disorientation were observed. The CSF results showed the presence of few lymphocytes in the field of view, increased concentration of protein and albumin, presence of oligoclonal IgG bands (type IV according to Charcot classification) and presence of antinuclear antibodies with immunofluorescence pattern similar to that found in serum (Fig. 1C). Based on the overall clinical picture the patient was diagnosed with CNS involvement in the course of pSS. Improvement of general condition and complete resolution of neuropsychiatric symptoms have been observed after immunosuppressive treatment with intravenous metyprednisolone pulses (5x500mg), followed by oral prednisone 1 mg/kg and intravenous cyclophosphamide infusions (15mg/kg once a month for six months). This case, along with other reports [4], points the need for an interdisciplinary approach to rheumatic diseases.


Fig. 1A. Electroencephalography (EEG) - Incorrect recording due to generalized background slowing, theta wave series over the left-sided leads and single focal spike-wave paroxysms over the frontal region during hyperventilation.
Fig. 1B. Histopathological evaluation of minor salivary glands.

Labial salivary glands with lymphocytic infiltrates which fulfill the criteria for Sjogren's syndrome (focus score ≥1; refers to a cluster of 50 or more lymphocytes per 4 mm²).
Fig. 1C. Evaluation of the cerebrospinal fluid (CSF) for the presence of autoantibodies.

Speckled staining pattern by indirect immunofluorescence on monkey liver cells.