Stiff person syndrome as initial manifestation of systemic lupus erythematosus: a case report

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Background: Stiff person syndrome (SPS) is a rare, but disabling neurological syndrome, associated with various autoimmune pathologies. Aims: This is a case-report of possible SPS, manifesting a year before the full-clinic of systemic lupus (SLE). Results: 67-year-old Caucasian female patient admitted to the clinic in February 2016 with acute gaze apraxia and unilateral blepharospasm. She had a history of hypertension and no autoimmune pathology. EMG showed periorbicular dystonia. MRI revealed vascular foci in the right frontal lobe. Symptoms regressed after a few weeks. After 6 months the condition suddenly deteriorated. Patient had painful stiffness of her legs, and she was restricted to the wheelchair. EMG revealed dystonia of lower extremities. There was CRP elevation to 22 mg/l, CSF protein 0.56 g/l, positive ANA 1:100. Paraneoplastic antibodies were negative. There was no opportunity to test GAD-antibodies, but due to the suspicion of SPS an immunotherapy was performed - methylprednisolone 500 mg IV, then per os intake from 32 mg. There was a dramatically improvement - the functions of lower extremities recovered completely. Her condition remained normal for a year. The exacerbation developed acutely in autumn 2017 after hyperinsolation with photodermatitis, legs rash, arthritis, heart failure, and painful legs stiffness. Carditis, atrial fibrillation, pneumonitis, nephritis, anemia, ESR 60 mm/h, CRP 56 mg/L, ANA 1:320 were revealed. SLE was diagnosed based on SLICC criteria. Patient received methylprednisolone 24 mg, hydroxychloroquine 200 mg, gabapentin with positive somatic and neurological dynamics. Conclusion: This case supports that SPS can outpace other symptoms of systemic autoimmune diseases.