







## Coexistence of juvenile systemic lupus erythematosus and Sjögren's disease in a 13-year-old girl

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**Introduction:** Confirming systemic autoimmune rheumatic diseases in children is a challenge. Many diseases lack validated classification criteria, so we use criteria developed for adults.

**Case description:** A 13-year-old girl with an episode of parotitis, persistent fatigue, cervical lymphadenopathy, splenomegaly, malar rash, erythematous papules, photosensitivity, intermittent fever ( $\leq 38.5^\circ\text{C}$ ) without infection, prior arthralgia, and then arthritis of many joints. No clinical or subjective sicca symptoms. Bacterial, viral infections (human immunodeficiency virus, Epstein-Barr virus, cytomegalovirus, parvovirus B19), and toxoplasmosis were excluded. Laboratory tests revealed: leukopenia –  $2.9 \times 10^3/\mu\text{l}$ , anemia – Hb 10.9 g/dl with positive DAT, [a]hypergammaglobulinemia; low C3, C4 complement, positive rheumatoid factor, antibodies to citrullinated protein antigens (–), anti-nuclear antibodies (ANA) (HEp-2) high-titer 1 : 10,240 (homogeneous) and 1 : 320 (speckled); anti-dsDNA elevated (393.2 IU/ml); anti-Ro60/Ro52 high (quantitative method); anti-SSB/La and AMA-M2 (+++; semiquantitative method); negative antiphospholipid antibodies, circulating immune complexes present. Based on the cervical lymph node biopsy, a proliferative process was excluded – reactive lymphoid hyperplasia. The patient was diagnosed with juvenile systemic lupus erythematosus (28 points European Alliance

of Associations for Rheumatology/American College of Rheumatology classification criteria for SLE). Sjögren's disease (SjD) diagnosis supported despite no minor salivary gland biopsy: typical Abs (anti-Ro60, anti-AMA-M2), SGUS score 2, recurrent sialadenitis. The SLEDAI 2K indicate high SLE activity (13 pts). Treatment with megadoses of glucocorticosteroid (GC) – methylprednisolone, maintenance therapy with oral GC (prednisone), methotrexate and hydroxychloroquine were introduced. Child lupus low disease activity was achieved. In 2024, GC was discontinued. The only clinical symptom was a discrete malar rash, peripheral blood counts, urinalysis were in norm, ANA titer 1 : 5,120 homogenous; 1 : 320 speckled pattern. Continued low C4 complement component and high Ro60 and Ro52 antibody concentrations without sicca symptoms.

**Conclusions:** Features of SLE appear to be identical to the adult form, but this is less obvious in children with SjD, especially with the absence of dryness symptoms. Recurrent parotitis, cutaneous manifestations, and similar laboratory abnormalities complicate the diagnostic process. Prior infections may act as a triggering factor, potentially explaining some atypical symptoms for both diseases (elevation of inflammatory markers). Treatment involves the same groups of drugs, and as in the case described, improvement was achieved in most domains.