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Therapeutic challenges of concurrent pneumonia and disease flare in a patient with Still's disease and recurrent macrophage activation syndrome

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Introduction: Still's disease belongs to non-familial autoinflammatory systemic disorders. Prior distinction of this condition into two separate entities: the systemic juvenile idiopathic arthritis (sJIA) and adult-onset Still's disease (AOSD), is obsolete, and European Alliance of Associations for Rheumatology recommends that it should be considered as one disease.

Case description: Patient at age 30, diagnosed with sJIA at 4. The first treatment was high doses of glucocorticosteroids (GCs). At that time, the patient 3 times develop macrophage activation syndrome (MAS). Since 2016, methotrexate (MTX) was prescribed, and in 2022, leflunomide was added as a second drug, but the patients took them irregularly. In 2022, due to a persistent subfebrile state and recurrent arthritis, he was qualified for treatment with tocilizumab at a dose of 400 mg applied in infusions every 4 weeks. Due to a remission state since 2023 multiple attempts to prolong the time between applications were made, but they failed. Finally, in 2025, the duration between dosages was successfully extended to 8 weeks. During therapy, the prior treatment with MTX was recommended but not taken by patients. In January 2026, the patient reported pain in multiple joints, fever, sore throat and rash on his trunk. In addition, patient complains of severe pain in his chest. In laboratory tests, the inflammatory indicators were very high. In the high-resolution computed tomography of the chest, the symptoms of pneumonia were described. Treatment with levofloxacin and a pulse of i.v. methylprednisolone has been introduced with subsequent treatment with prednisolone at a dose 1 mg/kg/day. The regression of changes in the lungs was achieved, the fever did not subside, and inflammatory indicators were still high, pointing to the development of MAS. After administration of tocilizumab, due anakinra was not available, complete remission was achieved. Although the interval from the end of pneumonia treatment was only one week. Due to the reluctance to use MTX, treatment with cyclosporine at a dose of 3 mg/kg/day was proposed.

Conclusions: The occurrence of severe pneumonia raises concerns about the safety of further tocilizumab treatment. A possible therapeutic option for this patient is interleukin-1 (IL-1) blockade, e.g. anakinra, which, according to the recommendations of the British Society for Rheumatology, is preferred in severe systemic disease over IL-6 blockade. In addition, it has been proven to be safe to use anakinra even in patients with sepsis, so it could be a therapeutic option in this case. Similar data for the use of tocilizumab in bacterial infection is lacking.