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DOI: https://doi.org/10.5114/reum/219173

A rare pulmonary manifestation of immunoglobulin G4-related disease refractory to standard treatment

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Key words: IgG4-related disease, pulmonary manifestations, treatment resistance, lymphadenopathy

Introduction: Immunoglobulin G4-related disease (IgG4-RD) is a chronic immune-mediated disease that typically affects organs such as the pancreas, bile ducts, salivary glands, and lymph nodes. Involvement of the lungs was also reported. The disease usually responds well to standard immunosuppressive and glucocorticosteroid (GC) therapy, and, in refractory cases, biologic treatment, as in the case of the presented patient.

Case description: A 48-year-old patient presented with lymphadenopathy persisting for one year, located in the head, neck, and supraclavicular areas, accompanied by increased sweating and weight loss. Oncological and infectious diseases were investigated, with no identifiable cause of the reported symptoms. A cervical lymph node biopsy revealed: plasma cell density of 150–200 focal cells/HPF, predominant CylgG(+) cells with 40–50% IgG4(+) subclass content, and typical storiform fibrosis pattern. This, along with elevated serum IgG4 concentrations, led to the diagnosis of IgG4-RD disease in October 2023. Treatment with methylprednisolone and methotrexate was initiated, but due to the lack of improvement after 2 months, cyclosporine was added (to a maximum dose of 5 mg/kg), without achieving remission. The patient's clinical condition did not improve; significantly elevated acute-phase markers persisted in the serum. During GC dose reduction, new foci of lymphadenopathy (mainly paraaortic nodes and along the iliac vessels) and fibrotic nodules in the lungs appeared. A verification biopsy of the inguinal lymph node was performed, confirming the diagnosis of IgG4-RD. Due to the ineffectiveness of standard treatment and exhaustion of therapeutic options, rituximab (RTX) therapy was decided upon. Three cycles of RTX were administered: in January and July 2025, and in January 2026. Between cycles of RTX administration, clinical improvement was observed, along with a reduction in the size of the cervical, axillary, inguinal, and abdominal lymph nodes, but with persistent progression of pulmonary lesions (intensification of nodular lesions and atelectasis of the right middle lobe).

Conclusions: The presented case report illustrates the course of IgG4-RD refractory to standard treatment that poses a clinical challenge. The anti-CD19 antibody, inebilizumab, recently recognised as a breakthrough in the treatment of this disease, may be a new therapeutic proposition for such patients.