Sinusogenes orbital complication as the first symptom of refractory granulomatosis with polyangiitis and the use of rituximab in treatment. A case report

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ABSTRACT

Granulomatosis with polyangiitis (GPA) is a rare disease involving multiple organs and systems, characterized by necrotizing inflammation of small and medium-sized vessels and formation of granulomas. There are several forms of the disease. In the generalized form (including severe one), cyclophosphamide in combination with steroids is usually used. There are also cases of a particularly severe and refractory course of the disease, which do not respond to the standard treatment. In such cases some other methods of treatment are recommended, including biological agents such as rituximab - anti-CD20 monoclonal antibody. We described a case of a 17-year-old patient with refractory GPA, resistant to standard remission induction therapy. In case of this patient, advanced lesions were mainly localized in the lungs, sinuses and middle ear. The disease progressed despite standard therapy. Therefore, it was decided to use an alternative treatment with a biological agent – rituximab. In the assessment after therapy we observed a significant clinical recovery as well as the improvement in laboratory tests, pulmonary function tests and radiological imaging. Within six months after the administration of rituximab, progression of the disease was not observed, which was evidenced in radiological imaging and pulmonary function tests. The case illustrates how biological agents, including rituximab, are extending the therapeutic options for patients with GPA, especially with severe forms of the disease.

Key words: granulomatosis with polyangiitis, refractory GPA, rituximab