A case of Immunoglobulin G4-related disease (IgG4-RD) treated with rituximab

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Immunoglobulin G4-related disease(IgG4-RD) is a recently recognized immune-mediated fibro-inflammatory disorder with multiorgan involvement. It is characterized by elevated serum IgG4 concentration, infiltration of IgG4+ plasma cells or lymphocytes, and fibrosis. Most IgG4-RD cases have a good therapeutic response to corticosteroids, and when this is insufficient the use of immunosuppressants may be effective. According to recent studies, Rituximab(RTX) is known to be effective in inducing remission and reducing corticosteroid use in IgG4-RD. Herein, we describe a case of IgG4-RD successfully treated with RTX. A 52-year-old male with a history of taking corticosteroids due to sinusitis presented to a gastroenterology clinic in 2011 with poor oral intake and weight loss of 6 kg during the previous 4 months. On abdominal computed tomography(CT) diffuse pancreas swelling was observed, following which an endoscopic ultrasound-guided biopsy was performed. The pancreas biopsy revealed chronic pancreatitis with massive infiltration of lymphoplasma cells and fibrosis, and elevated immunohistochemical expression of IgG4+ plasma cells. The patient was diagnosed with IgG4-related autoimmune pancreatitis, and was treated with oral prednisolone and azathioprine. In 2017, he was referred to a rheumatology clinic to evaluate Sjogren's syndrome because of dry eyes. Despite taking medication, his clinical symptoms were aggravated and serum IgG4 levels were elevated (3150 mg/dL; range: 3–201 mg/dL). Rituximab was started in November 2017 (2 cycles of 1 g at 15 days apart), in response to which his symptoms improved and IgG4 levels decreased (1230 mg/dL). In January 2020, RTX was administered one more time due to maculopapular rash on the lower extremities. Currently, the patient's disease status remains stable. In conclusion, several of the clinical manifestations of IgG4-RD, such as pancreatitis, dry eye syndrome, etc., can occur in the same patient. Corticosteroids remain the first line of IgG4-RD therapy. However, in recurrent or refractory cases despite administration of steroids or other immunosuppressants, treatment with RTX may lead to clinical and serologic improvement.

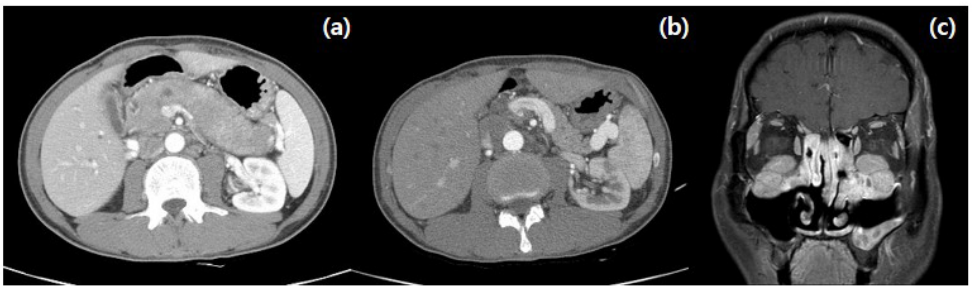

Figure I. (a-b) Abdomen computed tomography (CT) – (a) Diffuse pancreas swelling, (b) Improvement of diffuse pancreas swelling after steroid administration, (c) Orbit MR shows diffuse enhancing soft tissue lesion involving bilateral extraocular muscle

	17.02	17.03	17.06	17.09	17.11	18.02	18.07	19.11	20.01	20.04	21.07	22.06
IgG4 (mg/dL)	1790.0	3110.0	4000.0	3150.0	1230.0	1410.0	1220.0	758.0	653.0	451.0	1000.0	943.0