Multifocal ulcerating stenosing enteritis as a novel manifestation of immunoglobulin G4-related disease

Immunoglobulin G4 (IgG4)-related disease (IgG4-RD) is a rare systemic fibroinflammatory disease characterized by the presence of tumefactive lesions with dense infiltration of IgG4-positive plasma cells and sometimes serum elevated IgG4 [1]. Seventy-five percent of patients have two or more organs affected, with frequent involvement of the pancreas and the bile ducts [2]. Small-bowel involvement has rarely been reported, with only a few case reports in the literature [3]. The presence of IgG4-bearing plasma cells is essential for its diagnosis; an additional histological characteristic is eosinophil infiltration [1, 2]. There is uncertainty regarding its clinical presentation, diagnostic criteria, and treatment. Management with glucocorticoids may be an
appropriate option, as well as, in some cases, immunosuppressive maintenance treatment [4].

Herein we present the case of a 28-year-old woman with a history of iron deficiency anemia with no gynecological causes, and recurrent episodes of abdominal pain and bloating. Upper gastrointestinal endoscopy and colonoscopy showed no significant findings. A video capsule endoscopy was performed and revealed congestive mucosa with ulcers, scars, and zones of stenosis at the terminal ileum (▶ Video 1). A retrograde double-balloon enteroscopy was performed and demonstrated multiple areas of concentric irregular ulcers with secondary stenosis and scars (▶ Video 1). Hydropneumatic dilation was performed without complications (▶ Video 1).

The pathology report was consistent with IgG4-associated multifocal ulcerating stenosing enteritis (▶ Fig. 1 a–e). Positron emission tomography-computed tomography scan showed no extraintestinal IgG4-RD involvement. Systemic corticosteroid therapy was started, and long-term follow-up will be given.

In conclusion, we present a rare case of a patient with isolated bowel IgG4-RD, who presented with occult intestinal bleeding and stenosis, and was managed with hydropneumatic dilation and systemic steroid, with a satisfactory outcome at the time of writing this report. Long-term follow-up of these patients is required, as further lesions may appear as late as years after initial manifestation and could be located in distinct organs [4].

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Competing interests

The authors declare that they have no conflict of interest.

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