Immunoglobulin G4 (IgG4)-related disease (IgG4-RD) is a rare systemic fibro-inflammatory 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. 1a–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.