A 71-year-old woman was admitted with a 7-month history of watery diarrhea, which had led to an unintentional 30 kg (27%) of weight loss and admittance to the intensive care unit (ICU) on two separate occasions as a result of dehydration. Endoscopy and video capsule evaluation revealed villous atrophy of the entire small bowel, with fissuring, nodularity, and loss of folds, as shown for both duodenum (Fig. 1a) and ileum (Fig. 1b). The mucosa appeared fragile, with ulcers after biopsies (Fig. 1c). Histology confirmed complete villous atrophy and showed lengthened regenerative crypts, only a few intraepithelial lymphocytes, and thickening of the basal membrane in both proximal (Fig. 2) and distal small-bowel biopsies. Colonoscopy revealed a pale and edematous mucosa with superficial ulcerations, more pronounced distally (Fig. 3). Microscopic evaluation showed subtle inflammation in colon biopsies with focal erosion, a focally thickened basal membrane, and some apoptotic cells in the epithelium (Fig. 4a, b). Infectious, ischemic, and malignant disorders were excluded. Serum anti-βTg IgA and anti-gliadin IgG were negative during and after gluten exposure, ruling out celiac disease. The clinical presentation and diagnostic findings were most compatible with adult-onset autoimmune enteropathy [1], affecting an extensive part of the digestive tract (stomach to rectum). Immunosuppressive therapy was started; however high dose prednisolone, increasing doses of azathioprine, and immunoglobulins failed to induce any clinical response. The patient continued to produce voluminous diarrhea; however 3 weeks after starting therapy with oral budesonide (3 x 3 mg daily, pulverized in the morning, granules at noon, capsule at night) [1], the patient recovered, with formed stools, clinical improvement, and weight gain. Duodenal biopsies revealed completely restored villous architecture (Fig. 5). The patient has remained well for 20 months of follow-up. Thus, autoimmune enteropathy should be considered after exclusion of celiac disease when severe diarrhea is associated with villous atrophy. Topical immunosuppressive treatment should be applied.

Competing interests: None

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Fig. 1 A 71-year-old woman was admitted with a 7-month history of watery diarrhea and weight loss. Endoscopic images showing: a villous atrophy in the duodenum; b villous atrophy in the ileum; c ulcers after biopsies in the duodenum.

Fig. 2 Proximal small-bowel biopsy (hematoxylin and eosin (H&E) stain) of duodenal mucosa demonstrating severe villous atrophy, some inflammation, and thickening of the basal membrane.

Fig. 3 Colonoscopy of the distal colon showing pale and edematous mucosa with superficial ulcerations.
Reference


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Fig. 4 Colonic mucosa (H&E staining) showing: a focal erosion with homogenization of the lamina propria; b some apoptotic cells (arrows) in the epithelium.

Fig. 5 Restored villous architecture after topical budesonide treatment: a endoscopic view; b microscopic view.