Chronic diarrhea because of villous atrophy unrelated to celiac disease

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. 1 a) and ileum (Fig. 1 b). The mucosa appeared fragile, with ulcers after biopsies (Fig. 1 c). 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. 4 a, b). Infectious, ischemic, and malignant disorders were excluded. Serum anti-tTG 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 × 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.

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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.

Reference

Bibliography
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