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%) 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 intra-epithelial 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-iat 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.

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.
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
DOI http://dx.doi.org/10.1055/s-0034-1391247
Endoscopy 2015; 47: E71–E72
© Georg Thieme Verlag KG Stuttgart · New York
ISSN 0013-726X

Corresponding author
Anja Ursula van Lent, MD, PhD
Department of Gastroenterology and Hepatology Academic Medical Center University of Amsterdam P.O. Box 22700 1100 DE Amsterdam The Netherlands Fax: +31-20-5669608 a.u.vanlent@amc.uva.nl

van Lent Anja Ursula et al. Chronic diarrhea because of villous atrophy unrelated to celiac disease ... Endoscopy 2015; 47: E71–E72