Collagenous Sprue, an Enigma in the Spectrum of Celiac Disease

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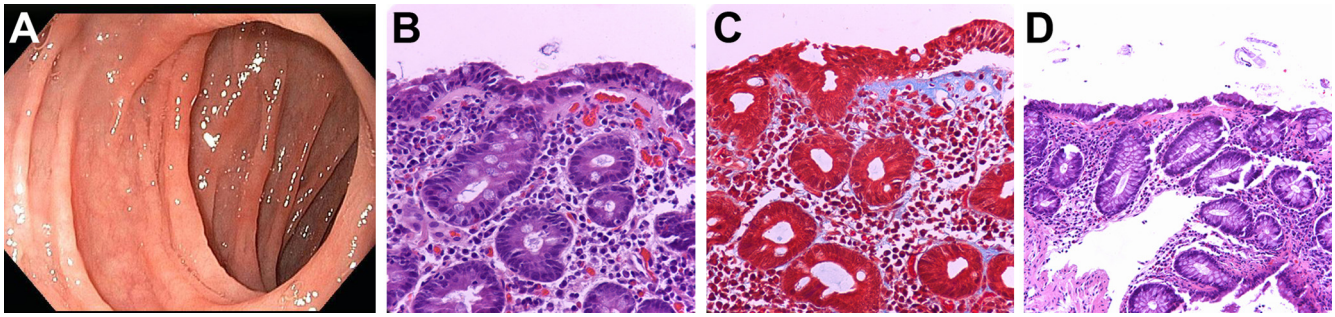

A 64-year-old white woman presented to our clinic with bilateral swelling of her hands and feet and an unintentional 50-pound weight loss over the past year. She was having 1 to 2 bowel movements daily with occasional additional watery stools without dietary variation. Laboratory values were as follows: alanine aminotransferase, 35 IU/L; aspartate aminotransferase, 46 IU/L; alkaline phosphatase, 117 IU/L; total bilirubin, 0.3 mg/dL; albumin, 1.7 g/dL; and hemoglobin, 11.6 g/dL. Her liver function tests later normalized after discontinuing her statin and meloxicam, but given her weight loss she was evaluated endoscopically. Esophagogastroduodenoscopy showed moderately erythematous mucosa of the entire stomach and blunted duodenal mucosa (Figure *A*). Her colonoscopy was normal. A biopsy showed almost complete flattening of the duodenal mucosa and subepithelial collagen deposition in the absence of intraepithelial lymphocytosis (Figure *B*). Collagen was highlighted with a trichrome stain (Figure *C*). Subepithelial collagen deposition also was seen in the cecum and in the left and right colon (Figure *D*). Antral and fundic biopsy specimens showed chronic gastritis. Celiac panel and later HLA-DQ2/DQ8 were negative. She was started on 9-mg oral budesonide capsules and initially a gluten-free diet for collagenous sprue. After 16 weeks of treatment she gained 24 pounds, her albumin level increased to 3.3 g/dL, and her edema resolved. She was started on an 8-week budesonide taper and a regular diet without return of symptoms.

Collagenous sprue (CS) is a rare cause of severe malabsorption. We present a case of collagenous sprue with concomitant collagenous colitis. Most cases of CS are reported in middle-aged to elderly Caucasian women. CS clinically presents similarly to celiac disease—with chronic diarrhea and weight loss. Our patient had an atypical presentation given her lack of diarrhea. Histopathologically, CS is distinguished by the presence of significant subepithelial collagen deposition entrapping blood vessels and inflammatory cells in addition to distorted villous and crypt architecture in the small bowel. There is ongoing debate regarding whether CS is a separate disease entity or simply part of the celiac spectrum. Some experts believe CS to be a histopathologic variant of celiac disease with a poorer outcome. Biopsy-proven celiac disease and CS both can have concomitant collagenous or lymphocytic gastritis and/or colitis. The prevalence of specific HLA haplotypes has not yet been proven in CS.[1,2]

Corticosteroids have been used increasingly to treat CS. The capsule formulation of oral budesonide has at least equal therapeutic efficacy compared with other steroids, with less first-pass hepatic metabolism, showing benefit with enhanced local colonic absorption; and this formulation has been used successfully to treat refractory sprue as well.[3] Response to dietary gluten removal has been shown to be inconsistent, particularly in HLA-DQ2/DQ8–negative patients.[1,2]

Collagenous sprue is a rare cause of severe malabsorption that may be part of the celiac spectrum. It can

involve multiple parts of the gastrointestinal tract and has a favorable response to corticosteroids.

References

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Conflicts of interest
The authors disclose no conflicts.

1542-3565/$36.00
http://dx.doi.org/10.1016/j.cgh.2013.05.031