

Intestinal Pseudo-Obstruction and Total Villous Atrophy of the Terminal Ileum: An Unusual Presentation of Untreated Celiac Disease

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Abstract

Intestinal pseudo-obstruction (IPO) is a rare complication of celiac disease (CD) and has often resulted in laparotomy for diagnosis. We report an adult case of CD presenting as IPO with severe protein calorie malnutrition (PCM) and negative endomysial as well as tissue transglutaminase antibodies. This is the first case report of CD presenting with combined IPO, severe PCM, negative first-line celiac serologies, and terminal ileal atrophy that was diagnosed without laparotomy. A non-surgical diagnosis was achieved by expanded laboratory and endoscopic methods, including video capsule endoscopy. Extent of pathologic gut involvement and response to treatment with budesonide and gluten-free diet is described.

Introduction

Intestinal pseudo-obstruction (IPO), by definition, presents with signs and symptoms of bowel obstruction in the absence of any identifiable occluding gut lesion. Not only is this condition quite unusual, but it is associated with significant morbidity and a broad differential diagnosis. The etiology of IPO may be idiopathic or secondary to a host of well-recognized underlying pathological conditions, including endocrine, autoimmune, neurologic, paraneoplastic, and inflammatory/infectious diseases. IPO has rarely been reported in adults as a manifestation of untreated celiac disease (CD) and was first described by Ingelfinger in 1943.¹ Only 7 additional adult cases have been described in the medical literature.²⁻⁶ All but one previous report underwent laparotomy to exclude mechanical obstruction.⁵

Case Report

A 52-year-old man reported being treated with sporadic courses of budesonide over a 6-year period for abdominal pain, diarrhea, and gradual weight loss, presumptively on the basis of inflammatory bowel disease. Financial restraints and non-compliance had precluded his previous gastroenterologists from thorough evaluation and continuity of care. He had supplemented his prescriptions of budesonide from those obtained by his daughter, who was being treated for Crohn's disease. The patient presented to our hospital service after several weeks of increasing diarrhea, abdominal pain, vomiting, and 6.8-kg weight loss. On physical examination, he was found to be afebrile and hypotensive, with a systolic pressure of 93 mmHg. He weighed 65.3 kg with a calculated BMI of 21 kg/m² and appeared chronically ill with somatic muscle wasting. The abdomen was modestly distended and diffusely tender to deep palpation without peritoneal signs. His bowel sounds were cavernous. Plain abdominal radiographs and CT abdomen (Figure 1) revealed diffuse small and large bowel dilation with significant edema of a large segment of the distal ileum.

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Figure 1. CT abdomen demonstrates diffuse gut dilation and significant edema of the distal ileum.

Comprehensive laboratory interrogation was remarkable for iron deficiency anemia (Hgb 8.1 gm/dL, iron 38 mcg/dL, ferritin 13 ng/mL) and hypoproteinemia (albumin 2.5 gm/dL, total protein 5.1 gm/dL). Serum immunoglobulin levels were normal. IgA tissue transglutaminase antibody (tTG) and antiendomysial antibodies (EMA) were negative; however, both IgG deaminated gliadin peptide (DGP) antibodies (105.0 U) and IgA DGP antibodies (74.6 U) were strongly positive (<20 U is considered negative for both indices). HLA DQ typing was permissive for CD. Neuromuscular markers for IPO including anti-neuronal nuclear Ab; type 1 ANNA-1, s; striational (striated muscle) Ab, s; N-type calcium channel Ab, acetylcholine receptor (muscle) binding Ab; AChR ganglionic neuronal Ab, s; and GAD65 Ab assay were negative.

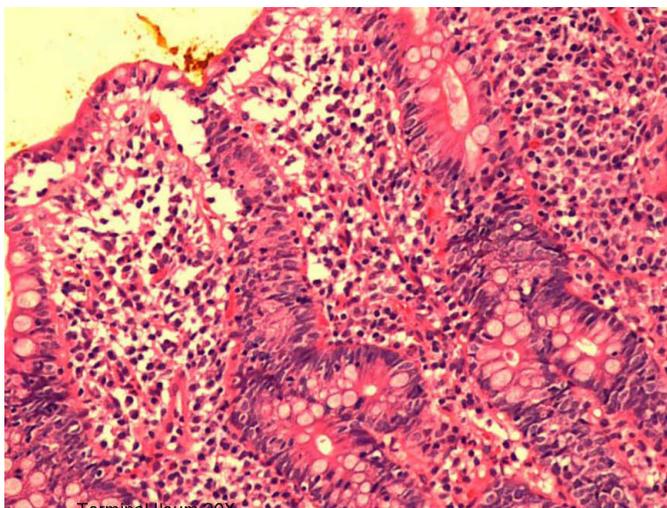


Figure 2. Histology of terminal ileum demonstrating total villous atrophy.

Pan endoscopy with duodenal, terminal ileal, and universal colon biopsies revealed endoscopic features of CD involving the duodenum. The colonic mucosa was edematous throughout. Histology revealed total villous atrophy of the duodenum and terminal ileum (Figure 2), with typical features of lymphocytic colitis involving the colon. A patency capsule procedure was negative. Subsequent video capsule endoscopy (VCE) identified stacking of folds and scalloped mucosa (Figure 3). No obstruction was identified.

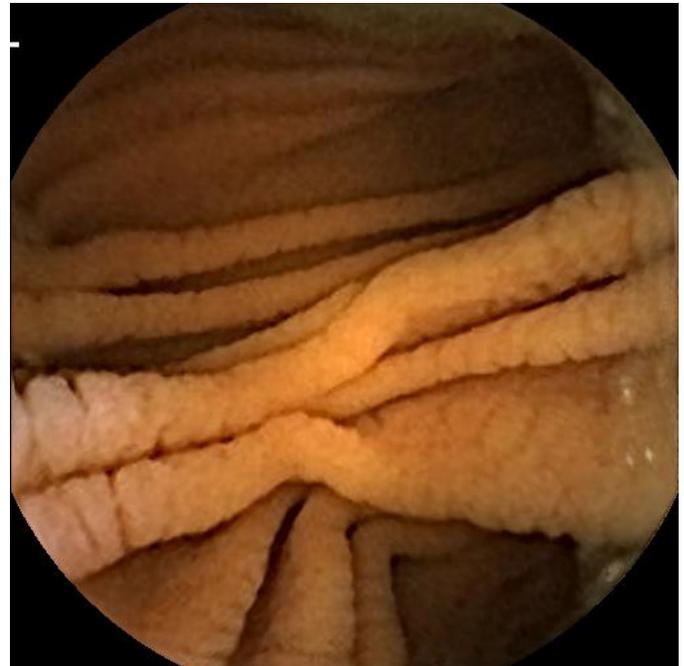


Figure 3. Stacking of folds and scalloped mucosa as seen on video capsule endoscopy (VCE).

The patient was placed on a gluten-free diet and prescribed budesonide 9 mg daily for 8 weeks. He returned for follow-up duodenal biopsies after 9 weeks of therapy. He was asymptomatic and had gained 4.5 kg in weight. Duodenal pathology had markedly improved and revealed only partial villous atrophy. The patient was seen again 5 months after discontinuation of budesonide therapy and remained free of gut symptoms on a gluten-free diet, with return to his pre-morbid weight of 85.3 kg.

Discussion

Intestinal pseudo-obstruction is a rare complication of CD and has often resulted in laparotomy for diagnosis. Pathogenesis of intestinal pseudo-instruction may involve neuromyopathic mechanisms as well as injury to the interstitial cells of Cajal.⁷ Celiac disease is an autoimmune disorder with IgA antibodies to the intestinal smooth muscle connective tissue known as the endomysium, as well as tissue transglutaminase, which is found within the endomysium.

These antibodies serve as markers for the diagnosis of celiac disease and would suggest a myopathic basis for IPO in untreated patients. Absence of these markers in our case would suggest an alternative mechanism(s) for the development of IPO. Perhaps direct gut injury related to the diffuse inflammatory process, as demonstrated in our case, was responsible for the development of IPO. Whether this inflammatory-related injury was primarily neuropathic, myopathic, or related to injury of the interstitial cells of Cajal is unknown.

In our patient, VCE excluded mechanical obstruction and provided images highly suggestive of CD, obviating unnecessary surgery. Our case illustrates the diffuse pathologic nature of untreated CD, in particular, total villous atrophy of the terminal ileum, which has not been described in previous cases of celiac-induced IPO. Despite our high index of suspicion, a confident diagnosis of CD was not supported by positive endomysial or tissue transglutaminase antibodies. Serologic exclusion for known neuromuscular markers of IPO allowed us to focus on the differential diagnosis of villous atrophy, which included autoimmune enteropathy and common variable immunodeficiency syndrome; negative anti-enterocyte antibody and normal immunoglobulins, respectively, excluded these causes of small intestinal villous atrophy. Tropical sprue, Crohn's disease, eosinophilic gastroenteritis, Zollinger-Ellison syndrome, giardiasis, bacterial overgrowth, and lymphoma as causes for small intestinal villous atrophy were reasonably excluded on clinical and/or histopathologic grounds. Permissive HLA typing results and positive deaminated gliadin antibodies supported our clinical diagnosis of CD as the most likely etiology of the pathologic and endoscopic findings. The diagnosis of CD was confirmed by the patient's resolution of symptoms and normalization of duodenal histology after treatment with gluten-free diet and tapering course of oral budesonide.

Disclosures

Author contributions: E.C. Gwillim participated in manuscript design, acquisition of data, and draft of manuscript; B.A. Bowyer participated manuscript design, acquisition of data, critical revision for intellectual content, and is the guarantor of the article.

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