

Celiac Crisis in a 64-Year-Old Woman: An Unusual Cause of Severe Diarrhea, Acidosis, and Malabsorption

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Abstract

Celiac disease (CD) rarely presents with life-threatening complications in older individuals. We report a 64-year-old woman who presented with profuse diarrhea, weight loss, hemodynamic instability, hypokalemia, hypoproteinemia, acidosis, and vitamin and iron deficiency. Pathologic and serologic studies confirmed CD presenting with celiac crisis with extensive and severe intestinal disease. Although celiac crisis occurs mostly in childhood and early adulthood, it should be considered in adults presenting with acute severe diarrheal illness, electrolyte abnormalities, and malabsorption.

Introduction

Celiac disease (CD) is an immune-mediated enteropathy triggered by an inappropriate response to dietary gluten and characterized by malabsorption and villous atrophy.¹ It is a common cause of diarrhea and malabsorption in adults.² Typical gastrointestinal manifestations of CD in adults include diarrhea, abdominal pain, bloating, and to a lesser extent, steatorrhea and weight loss.³ It is rare for adults to present with life-threatening complications, but presentation with celiac crisis is associated with high morbidity.

Case Report

A 64-year-old woman presented with profuse, watery, non-bloody, foul-smelling diarrhea of 2 months' duration with episodic vomiting, fatigue, and a 10-kg weight loss. There was no fever or abdominal pain. Her symptoms were preceded by an upper respiratory tract infection, and she had been taking iron tablets for anemia. She was afebrile, tachycardic, and had orthostatic hypotension. Her exam was significant for a cachectic appearance, dry skin and mucous membranes, and hyperactive bowel sounds.

The patient was resuscitated with intravenous normal saline, bicarbonate, and potassium chloride, and she was kept without oral intake. Laboratory values after resuscitation showed microcytic anemia (hemoglobin 8.7 g/dL, mean corpuscular volume 68 fL), WBC count 12,500/mm³, CRP 4.3 mg/dL, potassium 3.1 mmol/L, arterial pH 7.23, albumin 31 g/L, iron 29 µg/dL, ferritin 8.6 ng/mL, prothrombin time 16.5 s, and vitamin D at 4.7 ng/mL. She had normal renal and thyroid function. Vitamin B12 and folate levels were normal. Stool culture and microscopy on 3 stool specimens were negative. Stool Sudan 3 stain revealed numerous fat globules. Her diarrhea started to improve in the hospital.

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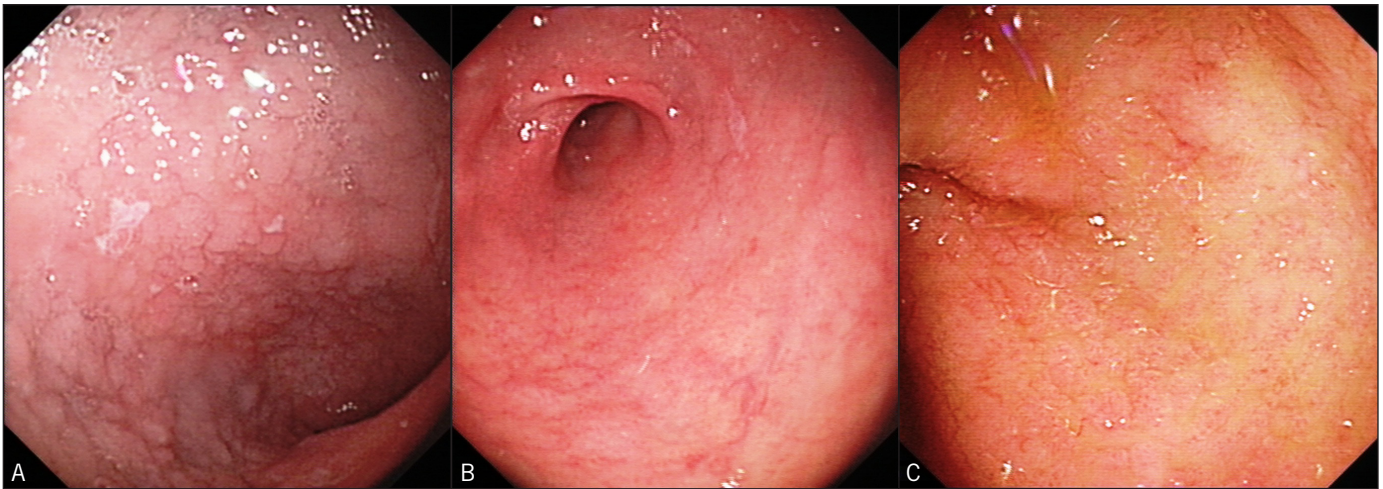


Figure 1. (A) Duodenal bulb showing mucosal nodularity and erosions on endoscopy. (B) Second part of the duodenum showing total loss of mucosal folds on endoscopy. (C) Terminal ileum showing ileal mucosal atrophy on endoscopy.

Esophagogastroduodenoscopy and ileoscopy revealed duodenal bulb erosions and mucosal nodularity (Figure 1A), total loss of mucosal folds in the second part of the duodenum (Figure 1B), and absence of villi and of granularity in the terminal ileum (Figure 1C). Duodenal biopsies showed total villous atrophy and hypoplasia (Marsh IV) with greater than 50 lymphocytes per 100 epithelial cells and crypt hyperplasia (Figure 2A). Ileal biopsies also showed villous atrophy (Figure 2B), and random colonic biopsies showed lymphocytic colitis (Figure 2C). She tested positive for IgA anti-endomysial antibodies with high titers of IgA anti-tissue transglutaminase antibodies (>200 U/mL, normal: <10 U/mL). She was diagnosed with CD presenting with a celiac crisis and was started on a gluten-free diet. She had a gradual recovery with a decrease in diarrhea and normalization of her electrolyte abnormalities. She was discharged in stable condition on a gluten-free diet with vitamin D and iron supplementation. On follow-up, she had weight gain with resolution of her diarrhea, anemia, and electrolyte abnormalities.

Discussion

Celiac crisis is a rare and potentially life-threatening complication of CD. It is characterized by the acute onset or the rapid progression of diarrhea with dehydration, hemodynamic instability, hypoproteinemia, weight loss, acidosis, and abnormal electrolyte levels.⁴ In 1953, Andersen and Di Sant'Agnese reported the clinical course of celiac crisis in 35 children; mortality was 9%.⁵ The condition has been described in 22 adults to date.⁴ The treatment of celiac crisis is gluten-free diet, but some patients require steroids.⁴

The cause of our patient's sudden deterioration is unclear, but celiac crisis has been described in association with or following surgery, pregnancy, immunosuppressive therapy, and infection. She had a rare atrophic form of celiac disease (Marsh IV),⁶ although it is not clear if small intestinal hypoplasia is associated with a severe clinical presentation.⁷ Ileal atrophy is rare and occurs in less than 5% of patients with CD,⁸ and likely contributed to our patient's

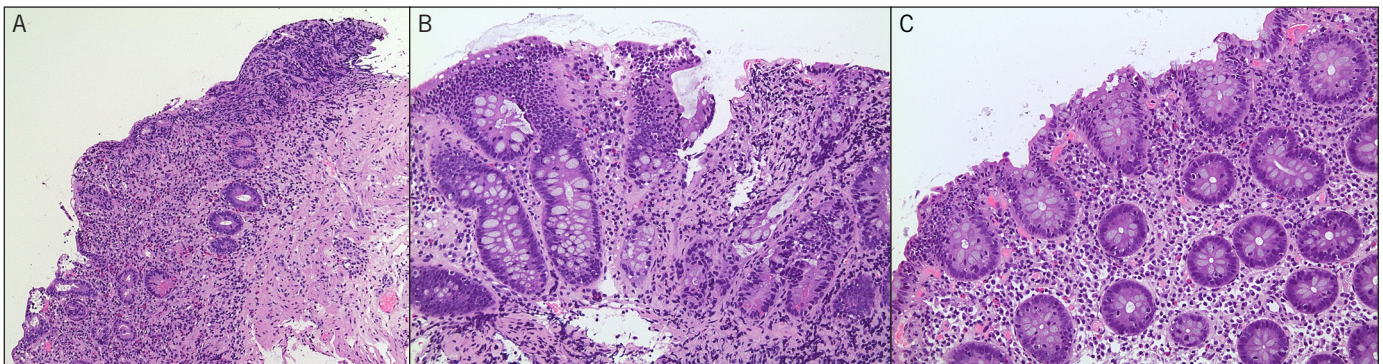


Figure 2. (A) Duodenal biopsy showing total villous atrophy and hypoplasia (Marsh IV). (B) Ileal biopsy showing villous atrophy. (C) Colonic biopsy revealing microscopic colitis.

deficient vitamin D and K. Microscopic colitis is associated with CD and affects about 4% of patients, and may have contributed to our patient's severe diarrhea.⁹

Though rare, celiac crisis should be considered in adults presenting with acute severe diarrheal illness, metabolic derangements, and evidence of malabsorption. In our patient, the atrophic form of the disease, extensive involvement of her small bowel, and microscopic colitis likely contributed to her dramatic presentation in celiac crisis.

Disclosures

Author contributions: R. Abou Mrad captured the images, performed the literature review, and drafted, edited, and finalized the manuscript. H. Abou Ghaddara performed the literature review, and drafted, edited, and finalized the manuscript. PH Green and N. El-Majzoub drafted, edited, and finalized the manuscript. KA Barada captured the images, edited and finalized the manuscript, and is the article guarantor.

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