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(CASE REPORT)

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Celiac crisis as an initial presentation of adult celiac disease: A case report and brief literature review

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Abstract

Celiac crisis is a rare, life-threatening manifestation of celiac disease characterized by acute, profuse diarrhea, severe dehydration, and profound metabolic disturbances. Although traditionally considered a pediatric complication, it is increasingly recognized in adults, often heralding the onset of the disease. We present the case of a 32-year-old previously healthy female who developed a celiac crisis. Extensive evaluations, including serology and duodenal biopsy, confirmed the diagnosis of celiac disease with marked villous atrophy, while effectively excluding enteropathy-associated T-cell lymphoma (EATL). The patient required intensive resuscitation, parenteral nutrition, and a short course of systemic corticosteroids alongside a strict gluten-free diet to achieve clinical and metabolic stabilization.

This case underscores the critical importance of considering celiac crisis in the differential diagnosis of severe acute malabsorptive syndromes in adults. Early recognition and prompt initiation of dietary and supportive therapies are paramount to prevent serious complications, such as refeeding syndrome or cardiovascular collapse. Furthermore, the successful integration of corticosteroids underscores their utility in refractory or critically ill presentations when dietary withdrawal alone is insufficient to reverse mucosal inflammation rapidly.

Keywords: Celiac disease; Celiac crisis; Immune-mediated enteropathy; Villous atrophy; Gluten-free diet

1. Introduction

Celiac disease is chronic, immune-mediated enteropathy triggered by dietary gluten in genetically susceptible individuals, classically presenting with an indolent course of malabsorption or extraintestinal manifestations. [1] However, a rare and potentially fatal fulminant presentation known as celiac crisis can occur, demanding immediate medical intervention. [2] Initially described in the pediatric population in 1953, celiac crisis is characterized by explosive diarrhea, profound dehydration, hemodynamic instability, and severe electrolyte derangements, such as hypokalemia, hypocalcemia, and metabolic acidosis. [3,4]

While advances in diagnostic modalities have led to earlier detection of celiac disease, adult cases of celiac crisis remain exceedingly uncommon, with fewer than 50 cases reported in modern literature. [5,6] This acute presentation can either herald the initial onset of celiac disease or result from severe dietary non-compliance in previously diagnosed patients. [7] The high morbidity and historical mortality associated with celiac crisis necessitate a high index of clinical suspicion, especially when common infectious etiologies for acute diarrhea have been excluded. Recognizing this entity is crucial,

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as the profound malabsorptive state and subsequent malnutrition predispose patients to life-threatening complications, including refeeding syndrome during initial nutritional rehabilitation. [8]

This report details a severe case of celiac crisis in a young adult female, emphasizing the diagnostic challenges, the multidisciplinary approach to excluding malignancy, and the successful implementation of corticosteroids as an adjunct to a gluten-free diet.

2. Case Presentation

2.1. Clinical Presentation

A 32-year-old previously healthy female presented to the emergency department with a 10-day history of profuse, watery diarrhea (up to 12 episodes/day), generalized fatigue, and a 7-kg weight loss. For three months prior, she had noted intermittent bloating and iron unresponsiveness (Refractory iron deficiency anemia), managed conservatively with oral iron and antispasmodics by her primary care physician without improvement. She sought care urgently after developing orthostatic dizziness, muscle cramps, and two episodes of hematemesis. She reported no NSAID use, travel, or sick contacts. On arrival, she was hypotensive (80/50 mmHg), tachycardic (120 bpm), and oliguric. Initial assessment observed severe dehydration and metabolic derangement, prompting admission for suspected acute gastroenteritis versus a malabsorptive crisis.

2.2. Physical Exam, History, Labs, Imaging, Differential

Physical examination revealed an emaciated, pale patient with dry mucous membranes and reduced skin turgor. The abdomen was distended with hyperactive bowel sounds, without evidence of edema or organomegaly. Neurologic examination was notable for generalized weakness, although the patient remained otherwise intact. Past medical history was unremarkable. Family history was significant for autoimmune thyroiditis in the mother and a sister with anemia of unknown etiology.

Laboratory evaluation demonstrated severe hypokalemia (2.1 mEq/L), hypomagnesemia (1.1 mg/dL), hypoalbuminemia (2.0 g/dL), and an elevated INR (1.8). A metabolic alkalosis was present (HCO3 34) in the setting of acute kidney injury (creatinine 2.5 mg/dL). Complete blood count revealed microcytic anemia (hemoglobin 7.2 g/dL, MCV 72) and thrombocytosis (550,000). Tissue transglutaminase IgA was markedly elevated (>200 U/mL) with a normal total IgA level.

Computed tomography of the abdomen with enterography demonstrated diffuse small bowel wall thickening, luminal dilation, mesenteric lymphadenopathy, and transient intussusception without evidence of obstruction. The differential diagnosis included severe Crohn's disease, tropical sprue, common variable immunodeficiency–associated enteropathy, autoimmune enteropathy, and lymphoma considering the lymphadenopathy and weight loss.

2.3. Multidisciplinary Discussion and Management Decisions

Following initial resuscitation with IV fluids, electrolyte repletion, and parenteral nutrition, the case was discussed at a multidisciplinary tumor board (gastroenterology, nutrition, radiology, and hematology-oncology). The discussion centered on three key questions: (1) Was there an underlying lymphoma mimicking or complicating celiac disease? (2) Should steroids be initiated for refractory celiac crisis, given ongoing diarrhea despite gluten withdrawal? (3) How to safely transition from parenteral to enteral nutrition.

After ruling out infectious etiology, the team decided to proceed with urgent upper endoscopy with deep duodenal biopsies to exclude EATL before initiating corticosteroids. Management consensus was to start an empirical gluten-free diet (GFD) immediately, with IV methylprednisolone (1 mg/kg/day) for 5 days if there is no response to GFD and hydration alone after 48 hours. Parenteral nutrition was continued until oral tolerance improved. Hematology advised PET/CT if biopsies suggested lymphoma.

2.4. Special Studies, Pathology, IHC, Molecular

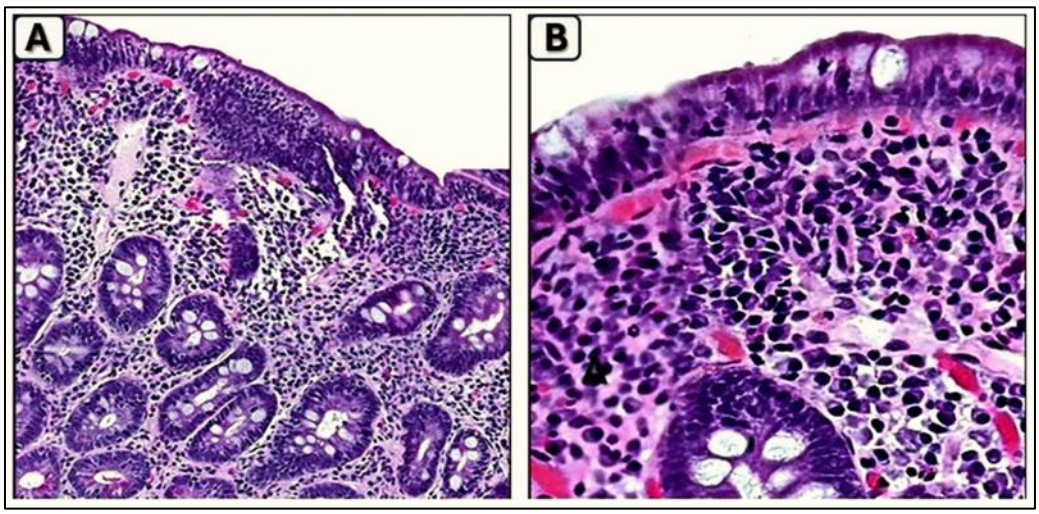
Upper endoscopy revealed scalloping and fissuring of duodenal mucosa. Histopathology showed subtotal villous atrophy indicating Marsh 3c (Modified Marsh-Oberhuber scale), crypt hyperplasia, and marked intraepithelial lymphocytosis (>40 lymphocytes/100 enterocytes). Immunohistochemistry (IHC) of duodenal biopsies showed CD3+ T-lymphocytes with retained CD8 expression and absent CD30 or CD56, arguing against EATL.

A concurrent capsule endoscopy (after stabilization) revealed patchy atrophy extending into the proximal Jejunum but no ulcerative strictures or masses. Iron studies confirmed absolute iron deficiency; anti-tissue transglutaminase IgA levels served as the monitoring biomarker.

2.5. Treatment, Outcome, Follow-Up, 18-Month Condition

Treatment started with IV methylprednisolone (5 days) followed by rapid oral prednisone taper over 6 weeks. Strict lifelong GFD with dietitian counseling. IV iron, vitamin D, calcium, and multivitamin supplementation. Proton-pump inhibitor for two weeks. Diarrhea resolved by day 4. Electrolytes normalized by day 6. She was discharged on day 12 on oral GFD and tapering prednisone.

Follow-up at 18 months: the patient was asymptomatic, fully active, and had returned to baseline weight. Anti-tTG IgA was undetectable. Repeat DXA showed significant improvement (Z-score -1.0, compared to earlier score of -2.5 at the lumbar spine). Iron studies are normal. No signs of autoimmune disease or malignancy. She was advised to remain on annual follow-up with adherence monitoring and screening for osteoporosis and associated autoimmune conditions.



1A**:** Intermediate-power view showing villous atrophy and crypt hyperplasia (H&E X40); 1B**:** High-power view showing marked intraepithelial lymphocytosis (H&E X60)

Figure 1 Microscopic features of Clear Cell Hidradenoma

3. Discussion

3.1. Background (History, epidemiology, risk factors)

Celiac disease (also known as celiac sprue or gluten-sensitive enteropathy) is characterized as an immune-mediated malabsorptive pathology that affects approximately 1% of the global population. [9] It has been well documented, with a clear pathogenesis and a thorough classification system that relies on histological (Modified Marsh-Oberhuber scale) and clinical (Oslo System) descriptions. [10] It is proposed that the disease first arose in humans during the first agricultural revolution approximately 10,000 years ago, when early human diets expanded to include gluten-containing cereals such as wild wheat and barley. [9,11] Archaeological studies have even shown evidence of cases of celiac disease in the first century AD. [12]

Celiac disease is vastly prevalent in highly populated countries in Europe and North America where the main inhabitants are of white ancestry. However, it also affects non-white populations to a lesser degree. Developing Celiac disease is highly dependent on family history and concurrent health conditions, mainly autoimmune conditions such as type 1 diabetes mellitus and autoimmune thyroid disease. It is also more common in individuals with chromosomal abnormalities like Down syndrome and Turner's syndrome. Females are disproportionately affected, with a female-to-male ratio of approximately 2-3:1. The age distribution of onset is bimodal, with the first peak in young children aged 9 months to 2 years and a second peak in the fourth decade of life. It is estimated that approximately two-thirds of patients remain undiagnosed due to atypical symptoms or being asymptomatic. [9,13]

Celiac crisis is a life-threatening complication that affects individuals with Celiac disease. Acute, severe gastrointestinal symptoms including profuse diarrhea, electrolyte imbalance, and metabolic disturbance characterize it. It was initially described in 1953 as a fulminant form of Celiac disease that exclusively affected the pediatric population and carried a mortality rate of almost 9%. [2] It is rare among adults and is almost exclusively documented as an isolated medical emergency, with only a few dozen cases described globally. [2,14]

3.2. Pathogenesis, Pathophysiology

The pathogenesis of celiac disease involves a complex interplay between environmental triggers, genetic susceptibility, and a dysregulated immune response. The primary environmental trigger is gluten, a proline-rich protein resistant to complete degradation by luminal proteases. [15]

In genetically predisposed individuals, typically carrying human leukocyte antigen (HLA) DQ2 or DQ8 alleles, these undigested gluten peptides cross the intestinal epithelium. Within the lamina propria, the enzyme tissue transglutaminase 2 (TG2) deamidates the gluten peptides, converting specific glutamine residues to glutamic acid. This modification significantly enhances the binding affinity of the peptides to the HLA-DQ2 or DQ8 molecules on antigen-presenting cells. [16,17] The presentation of these deamidated peptides activates gluten-specific CD4+ T cells, which subsequently release a cascade of pro-inflammatory cytokines, predominantly interferon-gamma (IFN-γ) and interleukin-21 (IL-21). [18] This inflammatory milieu drives the activation of intraepithelial cytotoxic CD8+ T cells (IE-CTLs) and the overexpression of interleukin-15 (IL-15) by stressed enterocytes. [19] IL-15 licenses the IE-CTLs to induce epithelial apoptosis, culminating in the characteristic crypt hyperplasia and villous atrophy. [20]

In the context of celiac crisis, this inflammatory cascade is profoundly amplified, often precipitated by an immune stimulus such as surgery, infection, or profound physiological stress, leading to rapid and extensive mucosal destruction, severe malabsorption, and the acute metabolic derangements observed clinically. [21]

3.3. Comparative Analysis of Our Case with Existing Literature

In adults, celiac crisis is a striking departure from the usually gradual course of adult-onset celiac disease. In a systematic review of 42 adult cases, Balaban et al. reported a median age at presentation of 50 years and a clear female predominance. [5] Our 32-year-old female patient fits this demographic pattern, although her illness was unusually severe. As reported in the literature, 88% of adult celiac crisis cases occur before a formal diagnosis of celiac disease; similarly, our patient's crisis was the first recognized manifestation of the disease, preceded only by refractory iron deficiency anemia. [5] Her profuse diarrhea, marked weight loss, and severe electrolyte abnormalities, particularly hypokalemia and hypomagnesemia, are consistent with the classic presentation described by Jamma et al. in the modern diagnostic criteria for celiac crisis. [2,3]

The diagnostic workup in our case was complicated by the presence of mesenteric lymphadenopathy and severe systemic symptoms, necessitating a thorough exclusion of EATL. While EATL is a known complication of refractory celiac disease, its presentation can mimic celiac crisis. [22] The multidisciplinary tumor board's decision to pursue deep duodenal biopsies with IHC was critical; the retained CD8 expression and absence of CD30/CD56 on intraepithelial lymphocytes effectively ruled out EATL, allowing for the safe initiation of immunosuppressive therapy.

Therapeutically, while a strict gluten-free diet and aggressive fluid resuscitation remain the cornerstones of management, the role of corticosteroids is increasingly recognized in severe or refractory cases. Early reports by Lloyd-Still et al. demonstrated the efficacy of corticosteroids in stabilizing pediatric celiac crisis. [23] In adults, Jamma et al. reported that 50% of their celiac crisis cohort required corticosteroid therapy due to an inadequate initial response to dietary restriction alone. [2,3] Our patient's rapid clinical improvement following the initiation of intravenous methylprednisolone underscores the utility of short-course systemic steroids in rapidly downregulating the intense mucosal inflammation characteristic of a crisis. Furthermore, the careful transition from parenteral to enteral nutrition in our patient highlights the critical need to anticipate and prevent refeeding syndrome, a potentially fatal complication documented in severely malnourished celiac crisis patients. [8,24]

4. What Have We Learned from This Case

This case provides several actionable clinical insights for the management of acute malabsorptive syndromes. First, it emphasizes that celiac disease must be included in the differential diagnosis of acute, severe, non-infectious diarrhea in adults, particularly when accompanied by profound metabolic derangements and a history of unexplained anemia.

Second, it highlights the diagnostic imperative to differentiate severe celiac crisis from EATL through rigorous histopathological and immunohistochemical evaluation before initiating immunosuppression. Third, the case reinforces the efficacy of systemic corticosteroids as a vital adjunct therapy when the response to a gluten-free diet is delayed, providing rapid stabilization of the mucosal inflammatory cascade.

Finally, the management trajectory underscores the necessity of a multidisciplinary approach, particularly concerning nutritional rehabilitation, where vigilant monitoring for refeeding syndrome is as critical as correcting the initial electrolyte deficits.

Abbreviations

- Enteropathy-associated T-cell lymphoma (EATL)
- Gluten-free diet (GFD)
- Modified Marsh-Oberhuber scale (Marsh 3c)
- Immunohistochemistry (IHC)

5. Conclusion

Celiac crisis is an exceptionally rare but life-threatening presentation of celiac disease in adulthood that requires prompt recognition and aggressive multidisciplinary management. This case report contributes to the sparse literature by detailing a severe initial presentation of celiac disease complicated by acute kidney injury and profound metabolic derangements, successfully managed with a combination of parenteral nutrition, systemic corticosteroids, and a strict gluten-free diet.

The rationale for reporting this case is to heighten clinical awareness among emergency and internal medicine physicians regarding this fulminant manifestation, ensuring that celiac disease is considered early in the differential diagnosis of acute severe diarrhea. By disseminating these findings, we aim to improve diagnostic vigilance, promote the safe and effective use of adjunctive corticosteroid therapy, and highlight the critical importance of preventing refeeding syndrome in critically malnourished patients, ultimately reducing morbidity and potential mortality associated with this severe clinical entity.

Compliance with ethical standards

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Disclosure of conflict of interest

All authors make the following declarations

- Payment/services information: All authors have declared that they received no financial support from any organization for the submitted work.
- Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might be interested in the submitted work.

Statement of informed consent

The patient was lost to follow-up, and all attempts to reach the patient were unsuccessful. Therefore, the paper has been sufficiently anonymized to maintain patient confidentiality.

Data access statement

All relevant data are included in the paper.

Author contributions

All authors contributed equally to producing this manuscript.

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