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A Celiac Crisis in an Adult: Raising Awareness of a Life-threatening Condition

Rogério Ferreira, Rui Pina, Nelson Cunha, Armando Carvalho
Department of Internal Medicine, Centro Hospitalar e Universitário de Coimbra, Portugal

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ABSTRACT

Objectives: The Authors report the case of a 56-year-old man with celiac disease, who after ingesting a food containing gluten and experiencing a flu-like syndrome, developed severe diarrhea, vomiting, weight loss (15 kg), hypotension, renal dysfunction, hypokalemia and metabolic acidosis.
Materials and Methods: Admission to the Intensive Care Unit and exclusion of an infectious cause was determined.
Results: After receiving noradrenaline, methylprednisolone and correction of ionic disturbances, the patient recovered rapidly and had no further complication.
Conclusion: The authors intend to increase awareness of celiac crisis, because despite being extremely rare in adults, it is potentially fatal and an quick diagnosis and treatment are crucial.

LEARNING POINTS

- Celiac crisis is an acute, life-threatening presentation of celiac disease.
- Its clinical presentation consists of severe diarrhea, metabolic disturbances (namely hypokalemia, hyponatremia, hypomagnesemia, hypocalcemia and metabolic acidosis), hypoproteinemia and dehydration.
- Hospitalization and correction of metabolic imbalance with intravenous fluids are required, with corticotherapy being necessary in some cases.

KEYWORDS

Celiac crisis, celiac disease.

INTRODUCTION

Celiac crisis is very rare in the adult population, with only about 20 cases reported in medical literature[1]. Its main clinical manifestations are profuse diarrhea, dehydration and severe metabolic disturbances. Despite its very significant morbidity, if recognised and properly treated, adult patients with celiac crisis described in literature have excellent clinical evolution[1,2]. Although the patient described in this case had a previous diagnosis of celiac disease and presented with severe symptoms of the disease, due to its rarity, celiac crisis was determined and treated with a vasopressor drug and corticotherapy only after a long period.
This case intends to increase awareness of this potentially life-threatening presentation among clinicians treating adult patients with severe diarrhea and important metabolic disturbances, especially if celiac disease is already known.

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CASE REPORT

A 56-year-old man presented to the Emergency Department (ED) with generalized myalgia, nasal congestion and rhinorrhea with 6 days of evolution, and a subfebrile temperature, nausea, intense vomiting and non-bloody diarrhea in the last three days.

His previous medical history included idiopathic recurrent acute pericarditis (7 episodes between 2000 and 2007); acute lithiasic pancreatitis (3 years before); and celiac disease (diagnosed two years prior, after being tested for chronic diarrhea and recurrent hypokalemia and with tests showing positive HLA-DQ2.5 alleles, negative anti-endomysial antibody and normal tissue transglutaminase IgA and IgG levels, and with duodenal biopsies compatible with Marsh's type 4 hypoplastic lesions and clinical improvement once a gluten-free diet was initiated). He had been on a gluten-free diet ever since, with the exception of some consumption of food containing gluten two months before this episode.

His physical examination showed no significant alterations and his analysis revealed leucocytosis (9.1 x109/L) and elevated C-Reactive Protein (117 mg/L), with normal renal function and no electrolyte disturbance. He was medicated with metoclopramide and intravenous fluids and after symptomatic improvement, he was discharged with the diagnosis of a flu-like syndrome.

Two weeks later, he returned to hospital for continuing diarrhea (3 to 4 episodes a day) and vomiting (3 to 5 episodes a day, and progressive worsening) and weight loss of 15 kg in the previous two-week period. He was hypotensive (84/45 mmHg), with no other alterations in the physical examination and had hypokalemia (2.9 mmol/L), discrete creatinine elevation (107.848 µmol/L), leucocytosis (14.2 x109/L) with normal C-Reactive Protein (2.3 mg/L) and compensated metabolic acidosis. After receiving an intravenous saline solution with potassium supplementation and feeling symptomatic improvement, he was again discharged with a prescription of oral potassium and a follow-up medical appointment.

He returned to the ED three days later with diarrhea (about 10 episodes each day), vomiting and asthenia. The physical examination revealed only slight pain in the palpation of the lower abdominal quadrants, with no other significant alterations, including fever. The laboratory test results showed elevated creatinine (334.1 µmol/L), leucocytosis (15.4 x109/L, with 80% neutrophils) with a C-Reactive Protein of 22.7 mg/L, hyponatremia (133 mmol/L) and a normal potassium (3.7 mmol/L). He was medicated with intravenous ciprofloxacin and electrolytes and admitted in the Internal Medicine Department.

In his first day there, he was hypotensive (73/44 mmHg), but initially responded to crystalloid fluids. Blood cultures were collected (both negative) and stool was collected for examination and culture; they were negative for *Salmonella*, *Shigella*, *Campylobacter*, *Yersinia*, *Clostridium difficile* toxin and ova, cysts and parasites. Auto-immunity associated with inflammatory bowel disease was negative. Abdominal radiography and ultra-sound revealed no significant alterations. In his second day of admission, he developed severe hypotension, metabolic acidosis, hypokalemia (2.7 mmol/L), hypophosphatemia (0.9 mmol/L) and oliguria. A central venous catheter was inserted and noradrenalin was initiated, along with correction of the ionic disturbances and intravenous methylprednisolone.

Due to the severity of the case, the patient was transferred to the Intensive Care Unit (ICU). In the initial phase of admission in the ICU, he presented hypokalemia, hypomagnesemia, hypophosphatemia and hypocalcaemia (which were all corrected), normocytic anemia and prolonged prothrombin time. He continued receiving intravenous methylprednisolone (80mg, every 8h); 24h after admission noradrenaline was suspended and the patient was able to restart a gluten-free diet. At the fifth day of admission, the dose of methylprednisolone was reduced to half and ciprofloxacin was stopped because there had still been no clinical or laboratory alterations suggestive of an infectious etiology.

During his seven-day period of admission in the ICU, the patient's renal function improved and normalised, metabolic disturbances were corrected and diarrhea progressively stopped. He was then transferred to the Internal Medicine Department and discharged seven days later, asymptomatic and with no metabolic disturbances.

At the 9-month follow-up, the patient maintained a gluten-free diet and had no further complications.

DISCUSSION

Celiac disease is an autoimmune enteropathy triggered by gluten proteins in genetically susceptible persons, characterised by malabsorption and villous atrophy[3-5]. The acute, life-threatening form of celiac disease has been described in literature as "celiac crisis"[1,2].

In adults, only about 20 cases have been described in literature, with a 3:1 proportion between women and men and a mean age of 51. 4 years at the time of diagnosis[1]. Proposed criteria for its definition require the acute start or rapid progression of gastrointestinal symptoms (severe diarrhea, vomiting), attributable to celiac disease requiring hospital admission and/or nutrition with at least two of the following:

1) Signs of severe dehydration (hemodynamic instability and/or orthostatic changes);
2) Neurologic dysfunction (peripheral neuropathy and tetany, due to hypocalcemia);
3) Renal dysfunction (creatinine > 176.8 µmol/L or 2.0 g/dL);
4) Metabolic acidosis (pH < 7.35);
5) Albumin < 3.0 g/dL;
6) Electrolyte disturbances (hypokalemia, hyper/hyponatremia, hypocalcemia, hypomagnesemia);
7) Weight loss > 10 lbs (4.5 Kg)[2].

Some patients also presented with prolonged prothrombin time[1]. In this clinical case, the patient had five of the criteria.
It remains unclear why, some individuals develop celiac crisis, contrary to the majority of adult patients who present a mild course of the disease. In the majority of cases, an immune stimulus (infection, severe inflammation, surgery, pregnancy or not following a gluten-free diet) happened prior to the crisis, which lead to the hypothesis that a combination of activation of the immune system, severe mucosal inflammation and disruption of normal intestinal motility could be involved[1,2]. In this particular case report, a combination of gluten consumption and infection preceded the onset of the crisis.
Due to its high morbidity rate, a quick diagnosis and treatment is crucial. However, similar to the case herein described, this diagnosis usually occurs later on andafter all infectious causes have been excluded. In all reported cases, hospitalization and correction of metabolic imbalance with intravenous fluids was required. About half the patients had to receive corticosteroids for treatment (with different corticoids and dosages being reported, and their reduction over the following period of months)[1,2]. Our patient, like all previously reported cases, had a quick response once proper treatment was initiated[1,2].

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