

Celiac crisis and hemorrhagic diathesis in an adult

Crise celíaca e diátese hemorrágica em um adulto

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ABSTRACT

Aims: Case-report of a patient with a rare presentation of celiac disease.

Case description: A 49 year-old woman, with a long-standing course of diarrhea and weight loss of 35 kg in a three-year period, presented to the emergency department complaining of an increase in diarrheal depositions, hematemesis, melena, lethargy, anasarca and syncope episodes. After clinical evaluation, upper gastrointestinal endoscopy and laboratorial exams, a rare illness known as celiac crisis was diagnosed. This is a rare presentation of celiac disease, associated with profuse diarrhea and severe metabolic/electrolyte disturbances, more frequent in children under two years, and rarely described in adults.

Conclusions: Celiac disease usually manifests itself through chronic diarrhea, abdominal pain and weight loss, but celiac crisis and hemorrhagic diathesis, although rarely, may also occur.

KEY WORDS: CELIAC DISEASE; CELIAC CRISIS; ENTEROPATHY; MALABSORPTION; HEMORRHAGE.

RESUMO

Objetivos: Relatar o caso de uma paciente com apresentação rara de doença celíaca.

Descrição do caso: Apresentamos o caso de uma mulher de 49 anos, com diarreia e emagrecimento de 35 kg em um período de três anos, trazida à emergência por anasarca, aumento nas eliminações diarreicas, vômitos com sangue, melena e episódios de síncope. Além de sintomas neurológicos, apresentava também distúrbios hidroeletrólíticos e perda de função renal. Após investigação clínica com exames endoscópicos e laboratoriais foi diagnosticada uma rara enfermidade conhecida com crise celíaca. Esta é uma apresentação rara da doença celíaca, cursando com diarreia profusa e distúrbios metabólicos/eletrolíticos severos, mais frequente em crianças de menos de dois anos e pouco descrita em adultos.

Conclusões: A doença celíaca manifesta-se classicamente por diarreia crônica, dor abdominal e perda de peso, embora apresentações mais raras, como crise celíaca e diátese hemorrágica, possam ocorrer.

DESCRIPTORIOS: DOENÇA CELÍACA; ENTEROPATIAS; HEMORRAGIA.

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INTRODUCTION

Celiac disease is an autoimmune gastrointestinal disorder in which ingestion of gluten present in wheat, rye and barley causes damage to mucosa of the small intestine.¹ The prevalence of celiac disease in the world is about 1% of individuals being one of the commonest genetically induced chronic disorder.²

Celiac crisis is a life-threatening complication that causes acute dramatic metabolic derangements, including severe diarrhea, hypoproteinemia, metabolic and electrolyte disorders significant enough to require hospitalization. Celiac crisis continues to be associated with high morbidity mandating identification and treatment.³ Treatment of celiac crisis is about parenteral fluid replacement and nutritional support, gluten free diet and in most cases use of corticosteroids.¹

The present case report has been reviewed and approved by a human research ethics committee from Hospital Nossa Senhora da Conceição, protocol number # 14-043.

CASE DESCRIPTION

A 49-years-old woman, with a long-standing course of diarrhea, presented to the emergency room complaining of complaining of an increase in diarrheal depositions, (more than 20 daily), accompanied of episodes of hematemesis and melena, which started 3 days before seeking for medical care. At the admission, she presented lethargic, pale and with sweaty skin, dehydrated (dry mouth) and in anasarca. Vital signs were remarkable for low blood pressure (50/40 mmHg), tachycardia (120 bpm) and tachypnea (26 mpm). At general inspection she had diffuse spontaneous ecchymosis in the upper and lower limbs and in the back. Laboratory workup showed hemoglobin 7.2 g/dL, creatinine 2.0 mg/dL, urea 36 mg/dL, sodium 122 mEq/L, potassium 2.9 mEq/L, magnesium 0.8 mEq/L, total calcium 4.5 mg/dL, albumin 1.2 mg/dL, glucose 43 mg/dL, platelets count 257,000/mm³, prothrombin time / international normalized ratio (INR) >15, activated partial thromboplastin time (aPTT) 154 seconds. Arterial blood gas showed metabolic acidosis. Thoracic X-ray showed bilateral pleural effusion and an abdomen ultrasound presented ascites. An upper gastrointestinal endoscopy (UGE) was performed showing only mild body-fundic enanthematous gastritis, according to the Sydney classification.⁴ Biopsies were not carried out at the time due to severe coagulopathy. Patient was managed initially with vigorous crystalloid resuscitation and transfusion of

blood components, besides vitamin K replacement, with normalization of coagulation tests after 5 days, when another UGE – this time with biopsies of the second portion of the duodenum as part of the investigation of chronic diarrhea. Histopathological analysis showed chronic duodenitis, lymphocytosis, hyperplastic crypts and marked atrophy of the intestinal villi (type 3c of the Marsh classification).⁵ Anti-endomysium antibodies IgA and IgA anti-transglutaminase tissue antibodies were positive, confirming the diagnosis of celiac disease. Due to chronic diarrhea over the past 3 years with a severe weight loss (approximately 35 kg), the patient had done already, in outpatient clinic, a colonoscopy with random biopsies of the colon, which showed no abnormalities. During hospitalization, lactose intolerance tested positive. After restrictive gluten and lactose diet, the patient presented progressive clinical and laboratory improvement, being discharged with a stable clinical status. At follow-up consultation in our outpatient clinic, three months after discharge, the patient was asymptomatic, with normalization of exams and referred having already regained about 25 kg of her previous normal weight. At last follow-up consultation at six months, she remained asymptomatic, maintaining strict diet.

DISCUSSION

We presented the case report of an adult patient diagnosed with celiac crisis – a rare and life threatening complication of celiac disease. Despite its high morbidity and mortality, there are few reports in adults,⁵⁻⁷ being mostly reported in children.⁸⁻¹⁰ The estimated prevalence of celiac disease is 1%, reaching 1:22 in first-degree relatives, 1:39 in second-degree relatives, 1:56 in symptomatic patients, and 1:133 in the not-at-risk groups.²

In adults, the classic presentation (diarrhea, abdominal pain and weight loss) is present in less than 50% of cases, besides the silent presentation with iron deficiency, osteoporosis and other metabolic disturbances.¹¹ Celiac disease can cause several hematologic disorders, such as anemia, thrombocytopenia, hyposplenism and coagulopathy, with prothrombin time elevation.¹²⁻¹³

Diagnosis of celiac disease is based on serological tests such as IgA anti-endomysium and IgA anti-transglutaminase antibodies which are close to 100% accurate and 90% sensitive,¹³ in addition to endoscopic and histological findings (duodenal flat mucosa, loss of villi and crypt hyperplasia), as well as response to a gluten-free diet.¹¹

The term “celiac crisis” was first described in 1953, by Anderson and di’ Sant’ Agnese, when they described the clinical course of 58 cases of celiac disease in children, of whom 35 presented severe and fulminant course.¹⁴ Celiac crisis is a rare presentation of celiac disease with intense diarrhea associated with metabolic and electrolyte abnormalities. Consensus among investigators have defined it as a condition that requires hospitalization and total parenteral nutrition, and at least two of other characteristics, such as: (a) signs of severe dehydration, including hemodynamic instability and/or orthostatic changes; (b) neurologic dysfunction; (c) elevation of creatinine, higher than 2g/ dL; (d) arterial blood gas with pH lower than 7.35; (e) albumin lower than 3 g/dL; (f) electrolyte disorders; and (g) weight loss higher than 4.5kg.¹⁵ This patient presented most of the symptoms and characteristics described above, such as electrolyte disturbances, loss of kidney function, acidosis, hypoalbuminemia, mental disorder and marked weight loss.

Due to the earlier diagnosis of celiac disease, celiac crisis is becoming less frequent.² In Dickey and McConnell study it was demonstrated that workup for celiac disease ended-up in diagnosis after average time of 6 years, in 9 out of 39 patients.¹⁶ Celiac crisis can be the first manifestation of the disease and, in most cases there is a triggering factor, such as surgeries, infections, bacterial overgrowth, and even pregnancy.¹⁷⁻¹⁸

Coagulation disorder is not a common manifestation of the celiac disease. Cavallaro et al.¹⁹ showed 18.5% of patients with celiac disease having an international normalized ratio (INR) greater than 1.4, due to injury in the surface of the small intestine, where bacteria synthesize and absorb vitamin K. There is a correlation between TP extent and other signs of bad absorption, such as hypoalbuminemia, iron deficiency and calcium reduction, without correlation with the time of disease, different from osteopenia, which has a direct correlation with the duration of the illness.^{17,19} The manifestation of hypothermia is ecchymosis, though there might be diffuse bleeding, such as the gastrointestinal tract, brain, joints, muscles, among others.¹⁹ In our case, we describe a hemorrhagic diathesis with upper digestive tract bleeding and ecchymosis associated with celiac crisis, what makes it a very rare case.

Since celiac crisis is an uncommon complication, there is no consensus about the treatment. Some authors recommend, besides removing gluten from the diet, the use of immunosuppressive drugs, such as corticosteroid and azathioprin²⁰⁻²¹ well-known for its anti-inflammatory effect to the intestine. In the reported case, immunosuppressive drugs were not used; even so, there was a progressive clinical and laboratorial improvement after removing gluten from the diet.

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