

NOTAS CLÍNICAS

Celiac crisis in adults: a case report and review of the literature focusing in the prevention of refeeding syndrome

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ABSTRACT

Introduction: Celiac crisis is a life-threatening complication of celiac disease that is rarely described in adults.

Case report: We report the case of a 31-year-old man with celiac crisis as a first manifestation of celiac disease. The patient presented with severe diarrhea, metabolic acidosis, and electrolyte disturbances accompanied by electrocardiographic alterations. A satisfactory clinical response was obtained after the correction of electrolyte abnormalities, hydration, and nutritional support with a gluten-free diet according to recommendations for patients at high risk of refeeding syndrome.

Discussion: Celiac crisis generally occurs in patients with no previous diagnosis of celiac disease. The physician should therefore be aware of this diagnosis and consider celiac crisis in cases of unexplained intense secretory diarrhea, metabolic acidosis and severe electrolyte alterations in adults. The risk of refeeding syndrome should be assessed when a gluten-free diet is introduced and treatment of celiac crisis should include prevention and management of this possible complication.

Key words: Celiac crisis. Refeeding syndrome. Celiac disease. Adults.

INTRODUCTION

Celiac crisis is a rare and life-threatening manifestation of celiac disease (1). This condition mainly affects children under the age of 2 years and is uncommon in adults (1,2). Adult celiac disease almost always has an insidious onset. About half of the patients have no overt gastrointestinal symptoms and many are asymptomatic. However, the occurrence of celiac crisis in adults who generally have no previous diagnosis of celiac disease has been described in the literature (2-10). Therefore, this diagnosis should be considered in cases of unexplained severe secretory diarrhea and metabolic acidosis in adults (3). Once the diagnosis of celiac crisis is established, the physician should be aware of the risk of refeeding syndrome. The introduction of a gluten-free diet can precipitate refeeding syndrome since

these patients present several risk factors for its occurrence, such as rapid and significant weight loss, low body mass index, little or no nutritional intake in the recent past, and severe electrolyte disturbance. The occurrence of refeeding syndrome can worsen the prognosis of celiac crisis (11).

CASE REPORT

A 31-year-old man was seen in the Emergency Department with a one-month history of diarrhea and intensification of symptoms in the last 16 days, accompanied by weight loss of 14 kg over the same period. The patient had large volumes of watery stools with steatorrhea, without blood or mucus (frequency of 10 times a day). There was no history of fever or recent travels. The patient reported recurrent episodes of diarrhea for about 10 years accompanied by weight loss and weakness, but no other medical conditions or medication use.

On admission, the patient was dehydrated, his blood pressure was 100/70 mmHg and pulse was 74/min. The respiratory rate was increased, but auscultation was normal. The body mass index was 17 kg/m². Initial laboratory tests revealed severe hypokalemia (K 1.5 mmol/L) associated with electrocardiographic alterations, metabolic acidosis, coagulopathy (INR 3.48), and acute kidney injury.

Vigorous intravenous hydration was initiated and electrolyte and acid-base disorders were corrected. The patient was started on intravenous potassium and enteral nutrition. A low-calorie (500 kcal/day) diet without lactose and gluten was introduced and the patient received thiamine and complex B vitamin reposition since admission. Clinical and laboratory improvement was observed in the following days and the patient was discharged on day 6 of hospitalization.

Laboratory investigation showed hypoalbuminemia (2.8 g/dL), hypogammaglobulinemia (0.34 g/dL), and iron deficiency (serum iron 23 µg/dL). Stool examination did not suggest an infectious etiology. HIV and hepatitis serology was negative. Upper digestive endoscopy showed

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atrophic duodenitis and a biopsy revealed intense villus atrophy and an increased number of intraepithelial lymphocytes. No parasites were found. The diagnosis of celiac disease was made based on the patient's clinical presentation, duodenal biopsy consistent with the disease, and improvement after the introduction of a gluten-free diet.

DISCUSSION

Celiac crisis is a medical emergency and potentially fatal complication of celiac disease. Although rarely described in adults, celiac crisis should be considered within a context of severe diarrhea, hypoproteinemia and electrolyte and metabolic alterations significant enough to require hospitalization. The syndrome is characterized by profuse diarrhea and symptoms related to electrolyte alterations, such as tetany and paralysis. There is generally no history of fever (1-8,10). In most cases, celiac crisis occurs in patients without a previous diagnosis of celiac disease (2-10), as observed in the case reported here. Therefore, a high clinical suspicion is needed in order to reach a diagnosis. Celiac crisis in patients with a previous diagnosis of celiac disease is often associated with dietary transgressions (1,3).

In cases of celiac crisis reported in the literature, the mean age of onset was 53 years, varying from 23 to 83 years, and the disorder was more frequent in women (15 *versus* 7 cases in men) (1-10). The most common electrolyte and acid-base alterations described in celiac crisis are hypokalemia, metabolic acidosis, and hypocalcemia (1-10). The lowest potassium level described was found in the present patient (1.5 mmol/L). Jamma et al. (3) proposed criteria for the diagnosis of celiac crisis, which included an acute onset or rapid progression of gastrointestinal symptoms that could be attributed to celiac disease and require hospitalization or parenteral nutrition, along with at least two objective signs of malnutrition, dehydration or electrolyte disturbance.

During the treatment of celiac crisis, refeeding syndrome is a manifestation that should be actively prevented and treated. The condition is still under-recognized and the fluid, electrolyte and mineral loss caused by diarrhea, associated with malnutrition, can contribute to the occurrence of this syndrome. Refeeding syndrome is defined as a severe metabolic disturbance that can occur in starved or malnourished patients receiving either enteral or parenteral refeeding (12). During refeeding, there is a metabolic shift from catabolism to anabolism in a process that requires minerals such as phosphate and magnesium and cofactors such as thiamine (11). The syndrome typically appears in the first days of refeeding and is potentially fatal if not recognized promptly, considering its possible complications such as hypotension, cardiac arrhythmias and respiratory failure (13). Risk factors for refeeding syndrome include little or no nutritional intake in the recent past, significant weight loss, low body mass index, electrolyte disturbance, specifically low phosphate, potassium or magnesium prior to feeding, and a history of alcohol abuse (12).

Patients with celiac crisis and risk factors for refeeding syndrome should be monitored carefully because clinical deterioration may occur after the introduction of a gluten-free diet. In the present case, nutrition support started with a reduced calorie intake of 500 kcal/day (about 10 kcal/kg/day) and vitamin reposition, which resulted in a good clinical response. Refeeding syndrome has not been described in adults with celiac crisis. Studies suggest a poor prognosis of celiac crisis in children when the condition is associated with refeeding syndrome (13), which can be aggravated by the use of corticosteroids, a therapeutic option described in the literature for celiac crisis, since it contributes to the depletion of potassium, magnesium and phosphate (14). Therefore, nutrition support should be started at an initial rate of 10 kcal/kg/day or 5 kcal/kg/day in case of extreme risk and should be gradually increased thereafter. The management of patients at risk of refeeding syndrome includes safe rate of feeding, thiamine reposition (if intravenous, daily for 3 days, and if enteral, for 10 days) and a general micronutrient supplement, as well as a daily dose of electrolytes in the first 5 days and 3 times weekly until stable (12).

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