Celiac Disease with Celiac Crisis

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Eur J Gen Med 2010;7(2):213-215

Received: 12.01.2009

Accepted: 08.04.2009

ABSTRACT

Celiac crisis is a life threatening and very rare complication of Celiac disease. Clinically, it is characterized by severe diarrhea, dehydration and metabolic disturbances like hypokalemia, hypomagnesemia, hypocalcemia, hypoproteinemia. A 2.5-years-old-girl with diarrhea, weight loss, abdominal distention, extremities edema, vomit which occurred after the beginning of supplementary nutrition was presented to emphasize that celiac crisis should be keep in mind in patient with gastrointestinal symptoms, which are best known in celiac disease, metabolic disturbance and abdominal distention.

Key words: *Celiac crisis; malnutrition; children*

Çöliak Kriz ile Seyreden Çöliak Hastalığı

Çöliak krizi; Çöliak hastalığının hayatı tehdit eden nadir bir komplikasyonudur. Klinikte şiddetli ishal, dehidratasyon ve hipokalemi, hipomagnezemi, hipokalsemi ve hipoprotenemi benzeri metabolik bozukluk ile karakterizedir. 2.5 yaşında kız hasta ishal, kilo kaybı, karın şişkinliği, extremitelerde ödem, beslendikten sonra kusma tablosu geldi. Bu vaka çöliak hastalığı olduğu iyi bilinen hastalarda metabolik bozukluk ve karın şişkinliği gibi gastrointestinal semptomlarının varlığında çöliak krizinin olabileceğini akılda tutmayı vurgulamak için sunuldu.

Anahtar kelimeler: *Çöliak krizi, malnütrisyon, çocuk*

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INTRODUCTION

Celiac disease (CD) is a disease of the small intestine caused by an immune response to ingested gluten. This response results in characteristic damage to the villi, leading to malabsorption (1). CD is manifested by a variety of clinical signs and symptoms that may begin in either childhood or adult life. Some individuals are completely asymptomatic (2). A 2.5-years-old-girl with diarrhea, weight loss, abdominal distention, extremities edema, vomit which occurred after the beginning of supplementary nutrition was presented to emphasize that celiac crisis should be keep in mind in patient with metabolic disturbance and abdominal distention.

CASE

A 2.5-years-old-girl was brought to our hospital due to weakness, weight loss, vomit and chronic diarrhea, abdominal distention and legs edema. From the history, it was learnt that these complaints were begun four months of her life when the time of supplementary nutrition beginning. On physical examination, body weight and height were 8.9 kg (-2.6 SDS), 78 cm (-2.5 SDS) respectively. Vital signs were stabile. The subcutaneous fat tissue was markedly decreased. Pink-brown colored desquamates macular rash were determined at abdomen and back. Abdominal distention was observed. On laboratory examination, complete blood count analysis was normal except mildly anemia (Hemoglobin 9.9 g/dL, Mean Corpusculer Volume 77 fL, Mean corpusculer hemoglobin 24, Mean corpuscular hemoglobin concentration 31, red cell distribution width 30). Erythrocyte sedimentation rate was normal and C-reactive protein was negative too. Liver and renal function tests were in normal range. Other biochemical examinations were normal except hyponatremia (122 mEq/L), hypopotassemia (2.5 mEq/L), and hypoalbuminemia (1.5 g/L). Thyroid function test and parathyroid hormone levels were in normal range. However, ferritin level (10 ng/mL) was found to be low. The serologies of human immunodeficiency virus, brucella, salmonella, hepatitis markers were negative. Cortisole and adrenocorticotrophic hormone levels were measured as normal (41.8 ng/mL, 17.3 pg/mL respectively). The abdomen and urinary systems' ultrasonographic examination and urine analysis were normal. The amount of protein excretions for 24 hours urine was under the 4 mg/m^2/hour. Serum folate and vitamin B_{12} levels were normal. Urine and blood culture were negative. The serologic markers of celiac disese were positive (tissue transglutaminase immunoglobulin (Ig) A and G, anti-gliadin Ig A and G, anti endomysium Ig A). Therefore, endoscopy was performed and biopsy was taken. The examination of intestinal biopsy was revealed the CD (Villous atrophy with hyperplasia of the crypts and increased intraepithelial lymphocyte count was found on examination of biopsy). Gluten free diet was begun to her. But after the diet, the metabolic disturbances could not be corrected (Serum sodium, potassium, albumin, calcium, phosphorus and blood pH, HCO_3, pCO_2 were measured as 116 mEq/L, 2.7 mEq/L, 1.8 g/L, 6.54 mg/dL, 2.77 mg/dL, 7.03, 6.7 mmol/L, 26 mmHg) and four days after, the patient was dead because of celiac crisis.

DISCUSSION

Toddlers and young children classically present with chronic diarrhea, vomiting, poor appetite, abdominal distension, abdominal pain, irritability, and failure to thrive sometime after the introduction of gluten in the diet (3). More frequently, the child with CD presents with subtle gastrointestinal symptoms such as constipation. The child may also present with non-gastrointestinal symptoms (e.g., short stature) or be asymptomatic but have a parent with CD. Some children may be simply cranky or have sleep disturbance (4) In general, CD should be included in the differential diagnosis of most patients seen in a pediatric gastroenterology practice. Our patient had classic symptom of celiac disease (weakness, weight loss, vomit and diarrhea, abdominal distention). The symptoms were begun after the supplementary nutrition, which included gluten. Our patient's results of serologic testing were positive. So endoscopic examination and biopsy were performed. Villous atrophy with hyperplasia of the crypts and increased intraepithelial lymphocyte count was found on examination of biopsy. Therefore, gluten free diet was given to her. The diagnosis of CD is established by positive results of serological testing and evidence of characteristic histopathology on intestinal biopsy (5). Characteristic histologic features of CD include varying degrees of villous atrophy, with hyperplasia of the crypts and increased intraepithelial lymphocyte count. The criteria proposed by Marsh are often used to grade the disease (from 0 to 4) in terms of these features (6). Most symptomatic patients have partial, subtotal or total villous atrophy, which are Marsh type 3 lesions.

Positive identification of these abnormalities leads to a presumptive diagnosis of CD and institution of a gluten-free diet. Clear clinical improvement while the patient is following the diet yields a definitive diagnosis. Celiac crisis is the term that has been applied to patients with CD of acute onset that is severe enough to be potentially fatal. It may arise in patients with established CD or, as in our patient, it may be the initial presentation of their disease (7). Clinically, it is characterized by severe diarrhea, dehydration and metabolic disturbances like hypokalemia, hypomagnesemia, hypocalcemia, hypoproteinemia (7). Various precipitating factors identified for crisis are severe malnutrition, infections, hypoproteinemia, and poor compliance to gluten free diet, bacterial overgrowth in setting of altered motility in CD and anticholinergic drugs.

Celiac crisis may not respond to a gluten-free diet alone. Some cases in literature, authors have used corticosteroids for variable periods. In severely ill children with celiac crisis, the use of corticosteroids may cause dramatic improvement (8). In 1952, Anderson and di'Sant-Agnese followed the clinical course of 58 children with CD (9). They observed 35 episodes of celiac crisis and 3 fatalities among these patients. In 1951 Adlersberg et al reported that the use of corticosteroids in adults with CD was effective therapy but relapse occurred when treatment was withdrawn (10). In 1972 Lloyd-Still described 3 cases of celiac crisis in children who presented with profound diarrhea, dehydration, metabolic abnormalities, and weight loss (11). They were successfully treated with corticosteroids. After that, Baranwal et al reported celiac crisis in a 5-year old girl with chronic intermittent diarrhea and gastrointestinal complaints who was treated with corticosteroids (12). But over the time it was realized that early recognition of CD and then gluten free diet in these patients is quite helpful to tilt the balance. In our patient celiac crisis was associated with hypoproteinemia and severe malnutrition. After the gluten free diet, the problems did not resolve. Therefore, other supporting treatment (albumin infusion, sodium replacement, gluten free diet, total parenteral nutrition) were given to her. However, despite these treatments, she was dead. Steroid could not given to our patient. It is not clear why celiac crisis has become a rare event during the past 50 years. It may be because of the changing nature of CD in general (ie, toward a more subtle and indolent disease). This may be the result of natural selection (13). It has also been hypothesized that factors such as the increased incidence of breast-feeding, the delayed introduction of baby food in general, and the introduction of gluten-containing cereals at an older age have contributed to the changing nature of CD (13). In conclusion, in some patients, the lesions of CD may be extensive and may therefore result in more severe symptoms.

In conclusion, we emphasized that; gastrointestinal symptoms, which are best known in celiac disease, metabolic disturbance and abdominal distention should preoccupy celiac crisis and besides supporting treatment, corticosteroids might be used for treatment.

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