CASE REPORT

CELIAC CRISIS IN AN IGA DEFICIENT CHILD

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Abstract

Celiac crisis is a life threatening and very rare complication of Celiac disease. It is characterized by severe diarrhea, dehydration and metabolic disturbances like hypokalemia, hypomagnesemia, hypocalcemia and hypoproteinemia. We present a 7 years old female child in celiac crisis and IgA deficiency.

Introduction

Celiac disease (CD) is a disease of the small intestine caused by an immune response to ingested gluten resulting in atrophy of the villi and malabsorption (1). CD is manifested by a variety of clinical signs and symptoms that begin early in childhood after introduction of gluten in diet. Celiac crisis is a rare complication of CD which must be kept in mind when treating a child with malabsorption, abdominal distension and metabolic disturbances.

Case Report

A 7 years old female child was admitted with 7-8 episodes of foul smelling yellow stools, non bilious non projectile vomiting, abdominal distension, fever, easy fatigability, cough and difficulty in breathing since 1 month. There was no contact with a patient having tuberculosis. She had received blood transfusion 2 months back. There was decreased appetite and weight loss. She had similar episodes of loose stools, vomiting and abdominal distension in the past that required multiple hospitalizations since the age of 2 years. Patient was diagnosed as a case of abdominal tuberculosis by a physician and was started on Anti tubercular therapy (ATT) with 4 drugs of Isoniazid, Rifampicin, Ethambutol and Pyrazinamide 20 days back but no improvement in the symptoms and general condition was noticed. On examination the patient was dehydrated and had pallor, frontal bossing, chelitis, angular stomatitis, aphthous ulcers, abdominal distension with shifting dullness, pitting edema over the legs, generalized wasting with loss of subcutaneous fat, desquamation of skin over the body including perianal area and significant axillary lymphadenopathy. Weight and height was less than 3rd percentile for age and mid arm circumference was 12.3cm. She had tachypnea with mild retractions. Auscultation of chest revealed bilaterally reduced air entry and fine rales. Investigations revealed hemoglobin of 7.2gm%, white cell count of 5000/cumm (50% polymorphs, 45% lymphocytes, 2% monocytes, 3% eosinophils), ESR(Westergren) 50 mm in 1st hr. Peripheral blood smear examination showed dimorphic anemia. Blood urea was 16mg/dl and serum creatinine was 0.6 mg/dl. Serum transaminases were normal, serum protein level was 2.5 gm/dl with albumin of 1.2 gm/dl. Serum sodium was 124 meq/l, serum potassium was 2.3 meq/l & serum calcium was 7.5 mg/dl. Urine culture had a growth of pseudomonas aeruginosa. Blood culture and 3 samples of sputum for acid fast bacilli (AFB) were negative. CSF examination was normal. Chest X-ray showed bilateral pleural effusion. USG abdomen revealed ascitis with mild hepatomegaly. The amount of protein excretion in 24 hours urine was less than 4 mg/m^2/hour. Tissue transglutaminase antibody (IgA) levels were 6.0 U/ml (Reference range 0-7 U/ml) and serum IgA levels were 29 mg/dl (Reference range 34- 305 mg/dl). Anti-gliadin IgG antibody titres were raised. Endoscopic biopsy revealed villous atrophy with hyperplasia of the crypts and increased intraepithelial lymphocyte count. The patient was put on a gluten free diet, IV antibiotics and ATT was stopped. Multivitamins, protein supplements and good nutritional diet were started .Within 15 days the symptoms subsided with improvement in the general condition of the patient. Child gained 2 kgs of weight, edema subsided and skin changes were reversed. Patient was discharged on gluten free diet, multivitamins and iron supplements. Corticosteroids were not given to the patient.

Discussion

Incidence of celiac disease is on rise in North India. Prevalence of celiac disease is found to be 1 in 310 children (2). 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). Atypical presentations of celiac disease include constipation, refractory anemia, short stature, clubbing and seizures (4). Some children may be simply cranky or have sleep disturbances (5). The prevalence of IgA deficiency is 1 in 500 in general population and IgA deficiency in patients with CD is 10 to 15 times higher than that in the general population (6).

Tissue transglutaminase antibody tests were done on our patient by local practitioners thrice earlier before patient presented to us, results of which were negative. But biopsy or IgA levels were never done, as a result of which patient landed in celiac crisis. This case report highlights the fact that serological tests alone should not be considered as the only criteria for ruling out celiac disease and every suspected patient of celiac disease must undergo serum IgA levels and endoscopic biopsy before labeling it as a non celiac.

Celiac crisis is a life threatening complication of celiac disease. Clinically, it is characterized by severe diarrhea, dehydration and metabolic disturbances like hypokalemia, hypomagnesemia, hypocalcemia, hypoproteinemia and metabolic acidosis. Various precipitating factors identified for crisis are severe malnutrition, infections, hypoproteinemia, and poor compliance to gluten free diet and anticholinergic drugs (7). The possible precipitating factors in present patient are severe hypokalemia, hypoproteinemia and infection. Celiac crisis may not respond to a gluten-free diet alone. In severely ill children with celiac crisis, the use of corticosteroids may cause dramatic improvement (8). Lloyd-Still described 3 cases of celiac crisis successfully treated with corticosteroids (9).The

role of steroids now is controversial as gluten free and good nutritional diet are considered good enough to tide over the crisis (10).

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Conflicts of Interest: None

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