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CASE REPORT

CELIAC DISEASE, A RARE CAUSE OF MALABSORPTION SYNDROME IN CHILDREN; THE FIRST CASE REPORT IN AN ETHIOPIAN CHILDREN

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ABSTRACT

Celiac disease Celiac disease is a multifactorial, autoimmune disorder that occurs in genetically susceptible individuals. The recognition of the complex clinical picture of the disease helps doctors to search and diagnose celiac disease even if the gastrointestinal symptoms are lacking. Individuals at risk for celiac disease should be thoroughly investigated and unusual manifestations of the disease should be screened actively.

Keywords: *Celiac disease ,Children, Ethiopia*

INTRODUCTION

Celiac disease is defined as an autoimmune disorder causing a permanent intolerance to gluten; a cereal protein present in grain from wheat, barley and rye. It is mainly occurs in whites and rare among Africans, Japanese and Chinese. Populations with certain genetic risk factors, i.e. HLA-DQ2 and –DQ8 are susceptible to the disease (1). The population based screening studies have shown that the overall prevalence of celiac disease among most European populations is at least 1% (2,3).

After absorption in the small intestine gluten interacts with the antigen-presenting cells in the lamina propria causing an inflammatory reaction that targets the mucosa of the small intestine. Manifestations range from no symptoms to overt malabsorption with involvement of multiple organ systems as extraintestinal manifestations and an increased risk of certain malignancies.

According to the current criteria, the diagnosis is based on a typical finding of villous atrophy and crypt hyperplasia in small-bowel mucosal specimen (4). Endomysial and tissue transglutaminase autoantibody tests correlate well with the small bowel mucosal findings (5). The diagnosis is presumptively established when there is concordance between the serologic results and the biopsy findings. It is confirmed when symptoms resolve subsequently on a gluten-free diet (6).

The major problem often encountered in diagnosing celiac disease is the multifaceted clinical picture of the condition. The disease is generally considered to affect mainly the gastrointestinal tract even though the severity of symptoms may vary from mild to severe irrespective of the occurrence of a manifest gluten dependent small-bowel mucosal lesion (7).

The disease is associated with symptoms of malabsorption such as steatorrhea, weight loss or other signs of nutrient or vitamin deficiency. Withdrawal of gluten-containing foods results in resolution of the mucosal lesions and symptoms (4). Therefore active serological screening and life-long gluten-free diet has paramount importance to diagnose and treat this disease (7).

CASE REPORT

This is a 4 yrs and 2 months old female child who presented with intermittent diarrhea and generalized body swelling since the age of 2 years. The diarrhea is non-bloody, yellowish to brownish in color, mucoid, and foul smelling and occurs 4-5 time per day. It is accompanied by occasional vomiting. She had on and off type of body swelling which started 6 months after the onset of the diarrhea. She was admitted with for nutritional rehabilitation as stated above She was taken to different clinics and treated with different syrups on various occasions, but illness did not show any improvement. She was admitted at Yekatit 12 hospital for nutritional rehabilitation and stayed for one month and discharged improved.

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The edema she had at admission subsided by the time she was discharged. Subsequently she was referred to Tikur Anbessa Specialized Teaching Hospital (TASTH), pediatrics Gastro intestinal (GI) clinic where she was investigated further.

Physical examination at TASTH showed a pulse rate (PR) of 102/minute, respiratory rate (RR) of 21/per minute, and axillary temperature of (T) of 36^0_C.Anthropometric measurements showed Weight (Wt) of 18kg, Wt for age that above standard for children of her age, height (Ht) of 98 cm, Ht for age that was between the $50^{th}$and 75^{th}percentile, and a mid-arm circumference (MUAC) of 14 centimeters. She had puffy face, grossly distended abdomen with positive fluid thrill and shifting dullness,+3 pedal, tibial and sacral pitting edema, and macular, hyper pigmented skin rash on the upper and lower extremities.

A clinical diagnosis of severe acutr malnutrition (SAM) associated with chronic diarrhea secondary to malabsorption syndrome and possible Allergic enteropathy was made. Investigation showed a white blood cell count WBC of 4,370 with a differential, neutrophil (N) count of 44.1%, and lymphocyte (L) count of 40.3. platelet (PLT) count of 432,000, hemoglobin (Hgb) of 16.4 g/dl, and hematocrit (Hct) of 46.7%. Urinanalysis was non- revealing. BUN was 9 mg/dl,creatinine(Cr) 0.3 mg/dl,serum glutaminoxaloacetic acetic transferrase (SGOT) 47 mg/dl, serum glutamin phosphate transferrase (SGPT) 29 mg/dl, Alanine Phosphate (ALP) 1060 mg/dl, bilirubin (Direct) 0.03 and (Total) 0.9 mg/dl, total serum protein (TSP)4gm/dl(albumin2.0gm/dl), serum cholesterol87 mg/dl, and antinuclear antibody (ANA) test negative. Abdominal ultrasound (U/S) examination showed moderate ascites and ascitic fluid analysis was non-revealing. Cytology examination showed reactive effusion Serum IgE of 20 IU (normal value <60IU). No ova or parasite (O/P) was seen on microscopic examination of the stools and stool culture was negative. Stool α_1-antitrypsin was 0.03 mg/dl (normal value <0.20 mg/dl).

Based on the above findings, a diagnosis of SAM secondary to protein loosing enteropathyand possible allergic enteropathy was made. The patient was started on prednisolone and plumpy nut. She continued to have generalized body swelling and diarrhea after one month of treatment. A duodenal biopsy and serological markers testing for celiac disease were then done. The following findings were observed:

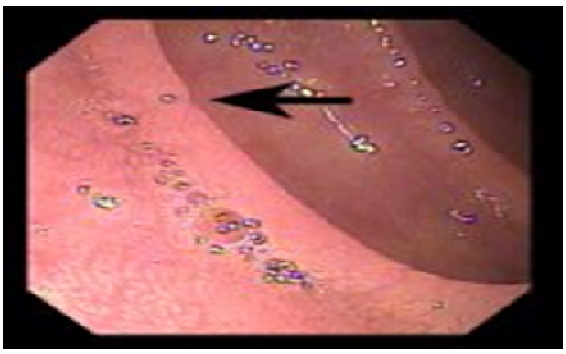

Figure 1: Duodenum Denuded and eaten up mucosae with scalloping on gastroscopy

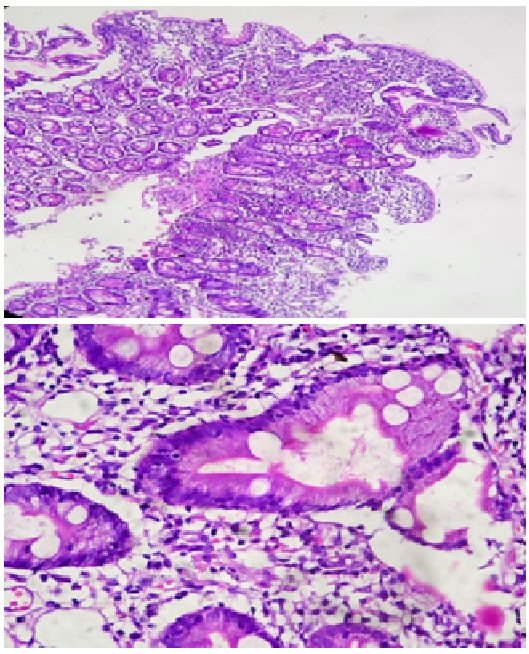

Figure 2. Total loss (atrophy) of the villiin dudenal biopsy specimens

Serological markers determined overseas with support from the International Clinical Laboratories showed anti endomysial antibody (AEMA IgA) and anti-tissue transglutaminase (tTG) antibodies were significantly elevated. Noticeable clinical improvement was observed on a gluten-free diet ('Teff') within several days, which substantiated the diagnosis of celiac disease.

DISCUSSION

The combination of a carefully taken medical history, meticulous conducted physical examination, and selected diagnostic tests (serologic results and the biopsy findings), establishes the diagnosis of ce-

liac disease and collagen spruein the majority of patients. Celiac disease is confirmed when a rapid response to a gluten-free diet is achieved (8), however, patients with Collagen sprue generally do not respond to a gluten-free diet (9). In our case, the patient was diagnosed with celiac disease based upon serological and histological findings with compatible clinical and laboratory manifestations and the clinical improvement observed with 'Teff') diet.

Celiac disease (luten-sensitive enteropathy) or nontropicalspue was first described in 1888 by GeeS This autoimmune disorder is one of the rare causes of Malabsorption syndrome in children with many extraintestinal manifestations like Iron deficiency anemia, hypocalcemia, and *Dermatitis herpetiformis*. Our patient showed noticeable response on Teff, a local gluten free diet (GFD).

The current criteria for the diagnosis of celiac disease may give the false impression that celiac disease is purely a gastrointestinal disorder with manifest small-bowel mucosal lesions. Based what is known today the diagnosis of the disease is much more complex. In the course of the disease, the small intestinal mucosal damage develops gradually, from normal morphology through inflammation to the so-called flat lesion (subtotal villous atrophy with crypt hyperplasia) (7). This process or mucosadeterioration may take years or even decades. This means that when a child, an adult, or an elderly person on a gluten-containing diet has been shown to have a normal small intestinal mucosal morphology, even with no cellular inflammation, the disease may not be excluded

***Conclusion**:* Duodenal findings with biopsy and serological markers are suggestive of celiac disease, in our case. Gastroscopy with intestinal biopsy and serologic markers (like AEMA, tTG antibody) have an important role in diagnosing celiac disease. This finding plus improvements on gluten free diet as in our patient makes a definitive diagnosis.

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