Case Report

CELIAC DISEASE

Zahra T[1], Memon AR[2] & Afsar SS[3]

SUMMARY

A 17 year old female, presented with complaint of vomiting, weakness and lethargy since one week. She had history of leg deformities and protruded chest since age three. There was also a history of progressive weight loss and growth retardation.On examination, she was anemic (Hb. 3.2g/dl) with protruded eyes, short stature and pigeon chest deformity. Serum calcium and albumin were decreased and serum globulins were increased. Tissue Trans Glutaminase(TTG) antibodies were elevated. Duodenal biopsy revealed features consistent with celiac sprue.

KEY WORDS: Anemia, hypoalbuminemia, growth retardation, weight loss, celiac disease.

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INTRODUCTION

Celiac disease (also known as celiac sprue and gluten sensitive enteropathy) is an autoimmune condition. It is triggered by ingestion of gluten containing grains (including wheat, rye, barley) in genetically susceptible individuals[1]. This particular case was reported because the patient was diagnosed after ten years. During this time, she was being treated for a skeletal disorder.

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CASE REPORT

A 17 year old female, presented with complaints of vomiting, since one week. The patient had developed bowing of legs, swelling of wrists and prominent protruded chest since the age of three. A history of progressive weight loss was also given. On presentation she could not walk with or without support. She had been hospitalized previously and diagnosed as having a skeletal disorder (most probably metabolic bone disease). On examination, she was anemic(Hb 3.2 g/dl) and dehydrated. There was prominent rib cage, retarded growth and short stature with bowing of legs. (Fig. 1)

Investigations showed microcytic, hypochromic anemia. Serum calcium was decreased with a value of 7.5mg/dl. Albumin and A/G ratio were also decreased (2.8g/dl and 0.56 respectively). Serum globulins were 5g/dl and alkaline phosphatase was 1090u/L.

The X-Rays of skull (Fig. 2), chest (Fig. 3), pelvis (Fig. 4) and both hands (Fig. 5) shows generalized osteopenia with paucity of secondary trabeculation. Multiple pseudofractures are identified in the rib cage and femora at different stages of healing. There is evidence of

Figure 1

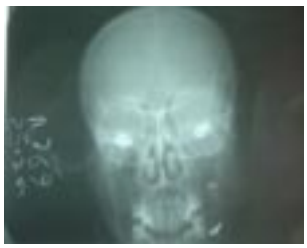

Figure 2

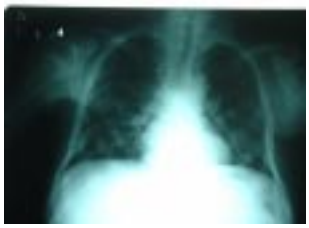

Figure 3

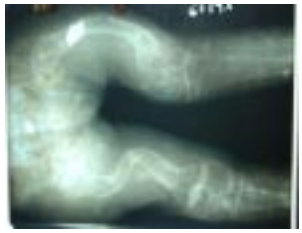

Figure 4

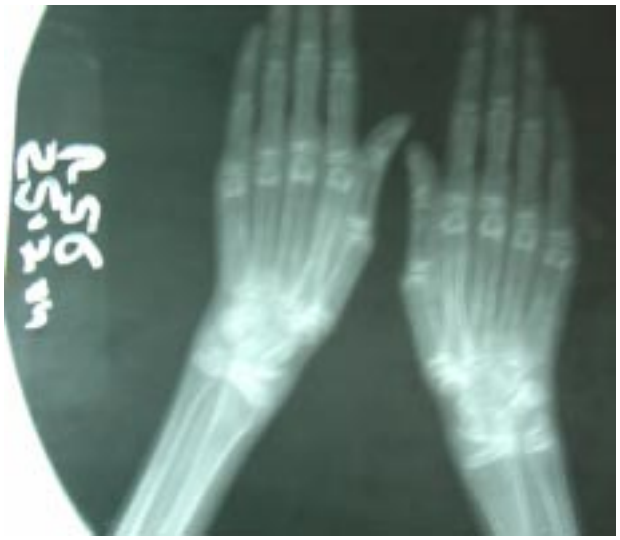

Figure 5

basilar invagination in lateral view of skull. Bulbous endings of costochondral junction and contracted deformed pelvis are reminiscent of a rachitic past. There is gross pubic diastasis, both femora are bowed and deformed consequent to osteopenia and multiple fractures. The bone age was estimated to be between ten to twelve years according to the ossification centers. Findings were supposed to be due to metabolic bone disease while osteogenesis imperfecta was a less likely differential.

The presence of iron deficiency anemia, hypoalbuminemia and osteopenia alongwith ricket raised the probability of a malabsorption syndrome. Further investigations revealed elevation of antigliaden and antiendomysial antibodies (IgA was 76.5U / ml, Ig G was 45U / ml).

Duodenal biopsy revealed severe villous stunting and increased intraepithelial lymphocytes with infiltration of lamina propria by lymphoplasma cells. The features were consistent with celiac sprue. She was counseled for a gluten free diet and oral calcium, active form of vitamin D and iron supplements were also started. Within two weeks the patient showed a dramatic improvement as her Hb went from 3.2 to 9.2 gm / dl. Physiotherapy and rehabilitation were also started in consultation with physiotherapy department. As a result, now she can move with support.

DISCUSSION

Celiac disease is self perpetuating in the continued presence of gluten. According to recent evidence, gluten induced upregulation of zonulin, an intestinal peptide involved in regulation of tight junctions, is partly responsible for the increase in gut permeability that is present in early phase of celiac disease and subsequent passage of gluten into lamina propria. Many epidemiological studies now indicate that this disease occurs more in females than males, which is applicable in the presented case[2]. The delay in diagnosis in this case was due to the severe skeletal deformities which misguided clinicians that she was suffering from some hereditary skeletal disorder.

Fasaro A[3] has mentioned clinical manifestations including chronic diarrhea, weight loss, and abdominal distension alongwith atypical manifestations of diabetes, microcytic / macrocytic anemia, osteoporosis, chronic fatigue, autoimmune disorders behavioral changes and intestinal lymphoma. Our patient had complaints of fatigue and lethargy,weight loss and muscle wasting, microcytic anemia and evidence of osteoporosis.

Laboratory findings of malabsorption include iron deficiency anemia, hypoalbuminemia and hypocalcemia all of which were present in our patient. Iron deficiency anemia

can occur because of impaired absorption and increased loss into gastrointestinal tract due to rapid epithelial cell turnover or associated with blood loss.

Short stature is well established as the only symptom of celiac disease in some older children and adolescents and it is believed that about 9-10% of those with idiopathic short stature have celiac disease. In them, both bone age and growth velocity are significantly impaired. These findings also correlate well with our presented case[4]. Also applicable in this case is the result of a study by Walters JR[5], which mentions that patients with celiac disease have a high risk of developing low bone mineral density and bone turnover impairment resulting in osteopenia.

Dieterich W[6] mentions the presence of endomysial and serum antigliaden antibodies as indicators of celiac disease. This is consistent with the findings in our patient. According to Schuppan D[7] small bowel biopsy is the investigation of choice. The results of this biopsy were the key to final diagnosis in the case of our patient. Histological changes in celiac disease range from minor villous blunting to subtotal or total villous atrophy. A gluten free diet is the most adequate treatment.

CONCLUSION

In view of high morbidity in case of untreated celiac disease, increased awareness is needed on the part of health care professionals so as to increase the quality of life of patients suffering from this condition. In our country diagnosis is delayed due to poverty and illiteracy. Patients generally do not seek medical advice. There is also less awareness of disease among clinicians as well as for quality of diagnostic tools. Hence it is important to develop awareness of this disease both in public and general practitioners because simple withdrawal of gluten from diet results in a dramatic clinical response besides prevention of many complications of celiac disease.

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