ORIGINAL ARTICLE

Clinical Presentation of Celiac Disease in Children

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ABSTRACT

Objectives: To evaluate clinical features of celiac disease in pediatric patients.
Materials & methods: The study was conducted at Department of Pediatrics, Services Institute of Medical Sciences (SIMS) and Services Hospital, Lahore. We performed retrospective data analysis of 70 patients in whom small bowel biopsy (SBB) was done with the suspicion of celiac disease. Forty six children had confirmed celiac disease on the basis of European Society for Pediatric Gastroenterology & Nutrition (ESPGAN) criteria. Twenty four patients due to lack of follow-up, equivocal response to gluten free diet (GFD) and non-specific histology were excluded from study. Clinical symptoms and signs, anthropometry and lab investigations were analyzed.
Results: A total of 46 children with M:F = 1:1.2 fulfill the diagnostic criteria for celiac disease. Mean age at diagnosis and onset of symptoms was 6.7 years and 3.4 years respectively. Chronic diarrhea, failure to thrive and pallor were the commonest clinical features present in 82.6%, 89% and 95.6% children respectively. Abdominal distension and clubbing was noted in 65.2% and 17.4% patients. Four children presented with rickets and three had edema feet on clinical examination. Weight & height <5th centile was present in 91.3% and 86.9% patients respectively. Hemoglobin <10 g/dl was found in 45 children and raised ALT >40 I.U. was present in 13 patients.
Conclusion: Chronic diarrhea, failure to thrive and anemia are three major clinical features in children with celiac disease. Diagnostic criteria should be fulfilled before labeling the child as celiac because GFD is recommended lifelong.
Keywords: Celiac disease, diarrhea, pallor, failure to thrive, children

INTRODUCTION

Celiac disease (CD) is a lifelong Gluten sensitive enteropathy. It has worldwide distribution with high prevalence in Caucasians. In Europe and United States prevalence estimates range from 1:80 to 1:300 children[1]. Punjabies from India living in England and eating a gluten rich diet have celiac disease, 2.9 times more often than Europeans[2]. Celiac disease has familial tendency with 10% risk of recurrence in first degree relatives. Concordance rate in homozygous twins is >70%.

Celiac disease results in autoimmune damage to intestinal mucosa from gliadine fraction of gluten in wheat, barley and rye. Both humoral and cell mediated immune mechanisms are involved. Histological features include villous atrophy, crypt hyperplasia and infiltration of lamina propria with chronic inflammatory cells. Mucosal damage is classified according to Marsh criteria as increased intra-epithelia lymphocytes (IELs) (Marsh type-I), partial villous atrophy (Marsh type-II), flat mucosa with complete loss of villi and crypt hyperplasia (Marsh type-III). Marsh type-IV lesions have the same histological features seen in type-III lesion except that the crypts are hypoplastic.

The patients with celiac disease present with gastrointestinal and non-gastrointestinal symptoms. Gastrointestinal symptoms, common in younger children (<3 years of age) include chronic diarrhea, anorexia, abdominal distension and vomiting. These symptoms are usually accompanied by consequences of malabsorption, like growth failure, weight loss, anemia and osteopenia. Non-gastrointestinal symptoms are more common in older children and adolescents[7] and include short stature, unexplained Iron deficiency anemia, delayed puberty and CNS manifestations like ataxia, peripheral neuropathy, headache, anxiety and depression. Some patients, usually first degree relatives of diagnosed case of celiac disease are asymptomatic and are classified as silent and latent celiacs. The patients with silent CD are asymptomatic but have positive serology and biopsy evidence of villous atrophy while the patients with latent CD have normal jejunal mucosa but positive serological tests.

Prevalence of celiac disease is frequently high in 1st and 2nd degree relatives of patients with CD, Turner syndrome, IgA deficiency[5], type-I Diabetes Mellitus[4] and thyroid disease[6]. There is increased risk for some malignancies, particularly Non Hodgkin Lymphoma in patients with CD as compared to general population.

ESPGAN (European Society for Pediatric Gastroenterology & Nutrition) criteria for diagnosis of

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CD includes small bowel biopsy (SBB) consistent with CD, and response to gluten free diet (GFD) (mandatory) and positive serology (supportive evidence) in the form of anti-gliadine antibodies (AGAs), anti-endomysial antibodies (EMAs) and anti-tissue transglutaminase antibodies (tTG), in patients with clinical suspension of CD. In addition, laboratory evidence of mal-absorption in the form of micronutrient deficiency is usually present in most of the patients.

MATERIALS AND METHODS

The study was conducted at Department of Pediatrics, Services Institute of Medical Sciences (SIMS) and Services Hospital, Lahore. We retrospectively analyzed the data of 70 patients in whom SBB was done with suspicion of CD. 46 children who had confirmed CD on the basis of SBB and response to GFD on follow-up and positive serology (in most of the patients) were included in the study. 24 patients with lack of follow-up, equivocal response to GFD and nonspecific histology were excluded from the study. Age at presentation and onset of illness, clinical symptoms and signs, anthropometry and laboratory investigations including complete blood counts with peripheral smear, S/E, S/albumin, Stool C/E, LFTs and viral markers were analyzed.

RESULTS

A total of 46 children, 25 girls and 21 boys (M: F = 1:1.2) fulfill the diagnostic criteria for CD. Mean age at diagnosis was 6.7 years and 15 patients (32.6%) were <5 years, 21 (45.7%) were 5-10 years and 10 patients (21.7%) were >10 years (Table-1). Regarding age at onset of symptoms the mean age was 3.4 years and most of the patients, 26(56.5%) had symptom onset under 2 years of age, 7 (15.2%) at 2-5 years of age, 11 (23.9%) at 6-10 years and 2 patients (4.3%) after the age of 10 years (Table-2)

Chronic diarrhea was the major complaint for seeking medical treatment. It was present in 38 (82.6%) children. Pallor and failure to thrive (FTT) were the two commonest clinical signs and symptoms, noted in 44 (95.6%) and 41 (89%) children respectively. Among patients with FTT 34 patients had complaints of diarrhea while 7 children presented within explained short stature. Irritability and abdominal distension were noted in 33 (71.7%) and 30 (65.2%) patients. Finger clubbing was present in 8 (17.4%) children. 4(8.7%) children presented with rickets and 3 (6.5%) had edema feet on clinical examination (Table-3).

Regarding weight and height, it was <5th centile in 42 (91.3%) and 40 (86.9%), 5th – 10th centile in 2 (4.3%) and 5 (10.9%) and >10th centile in 2 and 1 patients respectively (Table-4). Hemoglobin <10 g/dl was found in 45 children [it was <7 g/dl in 18 (39.1%) and 7-10 g/dl in 27 (58.7%)] and more than 10 g/dl in only one child (Table-5). In this group peripheral smear revealed microcytic hypochromic red blood cells in 39 (84.8%) patients.

Raised alanine aminotransaminase (ALT) >40 I.U. was found in 13 (28.2%) patients. Among these ALT was 41 – 60 I.U. in 11 patients and in only 2 patients it was >60 I.U. (Table-6).

Table 1: Age at Diagnosis (n = 46)

Age (years)	=n	%age
< 5	15	32.66
5 – 10	21	45.7
>10	10	21.7

Mean age = 6.7 years

Table 2: Age at Onset of Symptoms (n = 46)

Age (years)	=n	%age
<2	26	56.5
2 – 5	7	15.2
6 – 10	11	23.9
>10	2	4.3

Mean age = 3.4 years

Table 3: Clinical Presentation (n = 46)

	=n	%age
Chronic Diarrhoea	38	82.6
Failure to Thrive	41 (34+7)	89.1
Pallor	44	95.6
Irritability	33	71.7
Abdominal distension	30	65.2
Clubbing	8	17.4
Rickets	4	8.7
Oedema	3	6.5

Table 4: Anthropometric Measurements (n=46)

	<5th Centile	5th–10th Centile	>10th Centile
Weight	42 (91.3%)	2 (4.3%)	2
Height	40 (86.95%)	5 (10.9%)	1

Table 5: Hemoglobin Level (g/dl) (n=46)

Hb (g/dl)	=n	%age
<7	18	39.1
7 – 10	25	54.3
>10	3	6.5

Table 6: Alanin Transaminase Level (n=46)

ALT (I.U.)	=n	%age
<40	33	71.7
41 - 60	11	23.9
>60	2	4.2

DISCUSSION

A total of 46 children were found to be confirmed cases of celiac disease. Mean age at diagnosis was 6.7 years. It is consistent with a study from India where the mean age at diagnosis of CD in children

was also 6.7 years[8]. It is also comparable to a study from Switzerland[9] where this age was 6.8 years. In a study from Jordan, the mean age at the time of diagnosis was 8.4 years[10]. The mean age at onset of symptoms in our study was 3.4 years, which is close to the study from Jordan where this age is reported to be 4.6 years[10].

A female preponderance (25 girls & 21 boys) was found in our study, and it is similar to data from most of the countries. It is explained on the basis of autoimmune nature of celiac disease and the higher incidence of immunological disorders in females as compared to males.

Chronic diarrhea, FTT and progressive pallor were the major clinical presentations in our study, present in 82.6%, 89.1% and 95.6% children respectively. These findings are comparable with an Indian study where these ratios were 84%, 84% and 91% [(8)]. In a study from Turkey, diarrhea was the commonest clinical feature present in 81.7% patients, which is again consistent with our study[11].

Abdominal distension was present in 65.2% children and this finding is similar to Turkish study where abdominal distension was documented in 60.6% patients. Clubbing was noted in 17.4% patients in our study while this figure is reported as 26% in the international data[12].

Rickets and edema was found in 8.7% and 6.5% children respectively. In the report from Jordan, 26% patients with CD had rickets. It may be explained on the basis of late diagnosis in Jordan as compared to we people as mentioned previously (mean age at diagnosis, 6.8 years in our study versus 8.4 years in Jordanian study). Among patients with edema, 2 patients had associated chronic diarrhea and in one adolescent boy unexplained edema and hypoalbuminemia was the only clinical presentation of celiac disease, which settled on GFD.

Weight and height <5th centile were present in 91.3% and 86.9% children in our study. These findings are similar to Indian study where wasting was present in 87% and stunting in 60% of cases[8]. Data from Turkey reported short stature in 45.2% of cases, which is very low as compared to our study. It may be explained on the basis of very high incidence of malnutrition in our country which augments stunting in children with celiac disease.

Hemoglobin <10 g/dl was present in 93.5% children. 84.8% patients with anemia had microcytic hypochromic picture. It is consistent with the international data regarding nutritional anemia in patients with CD. Mild derangements in Alanine aminotransminase levels (ALT) was noted in 28.1% cases, which is consistent with another study where this value was 38.3%[11].

We had 2 patients with type-I diabetes mellitus (IDDM) and one child with Down syndrome, which are definite associations of CD. We did not find any case with dermatitis herpetiformis which is also a clinical manifestation of CD and more common in adults as compared to childhood CD.

CONCLUSIONS

- Chronic diarrhea, failure to thrive and anemia are three major clinical presentations in children with celiac disease.
- Diagnostic criteria should be fulfilled before labeling the child as celiac because gluten free diet is recommended lifelong.

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