Different Modes of Presentation in Children with Celiac Disease

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ABSTRACT

Objectives: Analysis of clinical presentation of celiac disease and effects of gluten free diet on clinical symptoms among children aging 2-15 years
Study design: Descriptive observational study.
Setting: Department of Paediatric Shaikh Zayed Hospital/FPGMI, Lahore.
Sample size: One hundred and fifty patients.
Subjects and methods: Duration of study was one year. Total 150 cases fulfilling the inclusion criteria were enrolled randomly. The immunological assays and proximal bowel biopsy were performed in every case. Results noted on proforma and analyzed.
Results: Chief complains were diarrhea (79.3%) and failure to thrive (80.7%). Main findings were short stature (87%) and anemia (72%). All had typical biopsy findings. Majority of biopsy proven cases responded to gluten free diet (80%) and showed marked improvement in their clinical and laboratory parameters.
Conclusion: Diarrhea and growth parameters improved markedly after treatment with gradual decline of titers of serum antigliadin and anti tissue transglutaminase. Patients with poor compliance to treatment were mostly early teenagers in whom disease remained unresolved..
Key Words: Celiac disease, Failure to thrive, Gluten

INTRODUCTION

Celiac disease is a disorder in which small bowel mucosa damages as the result of permanent sensitivity to gliadin fraction of dietary gluten[1]. Enteropathy associated with celiac disease predominantly involves proximal small intestine[2]. Presence of gluten in diet leads to self-perpetuating mucosal damage whereas elimination of gluten results in full mucosal recovery[3]. Disease is genetically determined with prominent HLA DQ2, DQ3, DR3, DR, association and restricted T-cell mediated reaction in intestinal epithelium[2,4].

Risk factors for celiac disease include dermatitis herpetiformis, type I diabetes mellitus, autoimmune thyroid disease, selective IgA deficiency, connective-tissue diseases, Down syndrome, collagenous or lymphocytic colitis, neurological and neuromuscular disorders, and a family history of celiac disease[5,6]. There are variable modes and age of presentation of celiac disease. Mean age of presentation Is between 3 to 5 years with typical disease presenting between 6 to 18 months and atypical between 5 to 6 years of age[3,7].

Classically children present with chronic diarrhea, failure to thrive, abdominal distension, anorexia and muscle wasting[1,6]. Stools are charactristically pale, loose, bulky and highly offensive due to fat malabsorption. A small number of Infants also have severe hypoproteinemia and edema and may present in shook like state that is termed as “celiac crisis” [3].

ETHODOLOGY

One hundred and fifty patients were included in the study. The study was conducted in Department of Paediatric Shaikh Zayed Hospital /FPGMI, Lahore from December, 2004 to November, 2005. All patients with clinical suspicion of celiac disease between age 2-15 years of either sex, biopsy proven cases of celiac disease and anti-gliadin antibodies and anti-tissue transglutaminase antibodies proven cases were

included in the study. All those patients with dysentery, persistent diarrhea, undergone major intestinal surgery, developmentally delayed children, patients with abdominal tuberculosis and In whom weaning has not yet been started were excluded.

General physical examination and detailed systemic examination was done and findings were noted. Investigations which were done to support diagnosis included complete blood count, peripheral smear, Erythrocyte sedimentation rate (ESR), stool microscopy, routine examination and culture sensitivity, serum total protein, albumin and electrolytes. Other diagnostic tests included qualitative analysis of serum anti gliadin (IgA, IgG) and serum tissue transglutaminase (IgA and IgG) along with distal duodenal biopsy done.

Histopathological findings of total/subtotal atrophy of villi with lymphocytic proliferation in submucosa was taken as gold standard diagnostic test. All the findings were recorded in a specifically designed proforma.

RESULTS

Details of results are given in tables 1, 2 & 3

Table 1: Presenting complaints (n=150)

Complaints	**n=**	**%age**
Diarrhea	114	79.3
Vomiting	86	57.3
Failure to thrive	121	80.7
Bulky + Foul smelling stools	75	50.0
Abdominal pain	29	19.3
Anorexia	72	48.0

Table 2: Results of immunological assays (n=150)

Immunological Assay	**No. of Patients with raised titer**	**%age**
Serum antigliadin IgA IgG	 123 113	 82.0 75.3
Serum tissue transglutaminase IgA & IgG	146	97.3

Key: IgA = Immunoglobulin A
IgG = Immunoglobulin B

Table 3: Effects of parameters before and after treatment

Parameters	**Before treatment (n=150)**	**After treatment (n=150)**
Diarrhea	114 (79.3%)	99 (66%)
Growth parameters	114 (79.3%)	92 (61.3%)
Titre of anti-gliadin antibodies (IgA)	123 (82%)	110 (73.3%)
Antibodies IgG	114 (79.3%)	91 (60.7%)
Serum tissue transglutaminase IgA	148 (98.7%)	128 (85.3%)

DISCUSSION

Celiac disease can present in multiple ways under 2 years of age. Many malabsorption syndromes present in almost the same way. Celiac disease can present at any age with peak in early childhood and then after 40 years of age. Disease has both typical and atypical presentation patterns.

As far as clinical presentation is concerned majority *of* patients presented with a combination *of* symptoms. In age group of 2 to 5 years, main complaints were diarrhea, vomiting and

failure to thrive while between 5 to 10 years of age, chief symptoms were bulky and foul smelling stools, anorexia and abdominal pain with off and on episodes of diarrhea.

Other studies conducted on presentation of celiac disease showed that although the “classical” gastrointestinal mal-absorption syndrome characterized by diarrhoea, steatorrhoea, weight loss, fatigue, and anaemia, most patients have a mild symptoms such as abdominal discomfort, bloating, indigestion, or non-gastrointestinal symptoms (or no symptoms at all)[8,9].

Weaning is thought to be largely associated with development of symptoms. It was seen that early weaning is a risk factor for development of celiac disease especially in genetically susceptible patients. In this study majority of children have history of weaning at the age of 5 to 6 months usually with rice and wheat based food.

Only 13 patients who got weaning foods after 6 months developed celiac disease later on in life. In another study which was conducted by Clemente et al (2003)[14], it was seen that direct effects of gliadin on enterocytes may also increase intestinal permeability leading to mal-absorption.

We noticed that confirmatory investigations like serum antigliadin (IgG and IgA), serum tissue transglutaminase (IgG and IgA)were raised in all the patients, showing these tests are very helpful in detecting the disease as well as to monitor the response of disease to gluten free diet (Sensitivity of 90% and 98% and specificity of 97% and 95% respectively).

Measuring tissue transglutaminase antibody levels is quicker, easier, and quantitative, so has clear advantages over the anti endomysial antibody test. Both tests have superseded the use of antigliadin antibodies, which although of some use have subsequently been shown to have inferior diagnostic accuracy with sensitivity as low as 76%[12,13].

All the patients were put on gluten free diet and special diet charts were provided to them. Patients who had strict adherence to gluten free diet (wheat, rye and barley) improved in their symptoms of diarrhoea within initial 2 – 3 weeks of treatment while growth parameters were also noticed to be improved both in terms of height velocity and weight gain. Initially barley was thought to be the part of gluten containing diet however, the trial by Janatuinen et al[14] showing no adverse effects after 6-12 months of dietary oats in patients with celiac disease. When growth parameters of the children included in the study were drawn on comparative standard growth charts it was found that majority of these patients had failure to thrive. In 76% of patients, height was less than 10th percentile while weight was less than 10th percentile in 70% of patients. Mid parental height was also taken to rule out any constitutional or familial delay which was on 25th percentile in 66% of patients. Pallor, clubbing, edema, muscle wasting were other symptoms to be notified. Abdominal distension and nutritional deficiencies especially of fat soluble vitamins were also seen.

Systemic examination in most of patient was unremarkable. A case report of 17 years old girl was published who had symptoms since the age of 3. She had bowing of legs, short stature, anemia and hypoalbuminemia, later on turned out to be celiac disease[11]. Laboratory Investigations which support the diagnosis were complete blood count which showed anemia in 72%. Some requires ongoing education of patients and their parents by both doctor and dieticians. More cheaper Immune assays are now available with good diagnostic value which are also helpful In monitoring the response to treatment. Single upper GI endoscopy and biopsy remains gold standard for diagnosis. All patients with celiac disease should be put on lifelong gluten free diet, which leads to prevention of complications as well as primary disease manifestation.

CONCLUSION

Diarrhea and growth parameters improved markedly after treatment with gradual decline of titers of serum antigliadin and anti tissue transglutaminase. Patients with poor compliance to treatment were mostly early teenagers in whom disease remained unresolved..

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