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Case Report

Multiple Faces of Celiac Disease-A Case Report

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Abstract

Celiac disease is also called as gluten-sensitive enteropathy and non-tropical sprue. It was first described by Dr. Samuel Gee in an 1888. The term "coeliac" was derived from Greek word *koiliakaos*-abdominal. The similar description by Aretaeus from Turkey reaches as far back as the second century AD. The cause of celiac disease was unexplained until 1950 when Dicke recognized an association between the consumption of bread, cereals and relapsing diarrhea. This observation was corroborated when, during periods of food shortage in the Second World War, the symptoms of patients improved once bread was replaced by unconventional, non-cereal containing foods like pure fruit, potatoes, banana, milk or meat. It is a chronic immune-mediated disorder of small intestine that occurs in genetically predisposed populations due to permanent intolerance to wheat gliadins and other cereal prolamins. They found that wheat, rye, barley and to a lesser degree oats, triggered malabsorption, which could be reversed after exclusion of the "toxic" cereals from the diet. Shortly after, the toxic agents were found to be present in gluten, the alcohol-soluble fraction of wheat protein. The epidemiology of CD has iceberg characteristics with more undiagnosed cases. The diagnosis of CD is currently based on both typical small bowel biopsy findings with clinical and serological parameters. The compliance rate regarding strict gluten restricted diet is variable and ranges from 50% to 90%. was Ninety (89.77%) and is more in females and in older age group in comparison to males and younger age group.

Keywords: *Celiac Disease; Non Tropical Sprue; Duodenal Biopsy; Gluten; Compliance*

Introduction and Discussion

Celiac disease (CD) is a chronic immune-mediated disorder of small intestine that occurs in genetically predisposed populations [1]. The pathogenesis entails a T cell mediated immune response with production of auto antibodies directed against tissue transglutaminase or endomysium. Wheat, rye, oat and barley prolamins are toxic for celiac patients due to their high glutamine and proline content [2]. Celiac disease is a multifactorial, autoimmune disorder that occurs in genetically susceptible individuals. The Trigger is an environmental agent-gliadin component of gluten. The enzyme tissue transglutaminase (tTG) has been discovered to be the auto antigen against which the abnormal immune response is directed. Gliadin is a glycoprotein present in wheat and other grains such as rye, barley and to some degree, oats. Gluten is a composite of the proteins gliadin and glutenin which comprise about 80% of the protein contained in wheat seed. The pathogenesis is an abnormal permeability allowing the entry of gliadin peptides not entirely degraded by the Intraluminal and brush-border bound peptides. Many fractions of gliadin are very resistant to digestion by gastric, pancreatic, and mucosa-associated enzymes. The intestinal epithelium is a barrier to the passage of these macromolecules; however in CD there is loosening of the tight junctions lead-

ing to increased permeability to macromolecules. Due to increased permeability of the macromolecules there are two pathways involved in the pathogenesis. The early pathway involves the innate immune system and the subsequent pathway involves the T cells. When the toxic gliadin peptides reach the serosal side of the intestinal epithelium an early response by the innate immune system causes crucial modifications of the mucosal microenvironment. The stage is then set for the subsequent involvement of the pathogenic T cells and an inflammatory response. Initially the suspicion of CD was based on clinical gastrointestinal (GI) symptoms. Subsequently, the disease has been found with variety of atypical symptoms and even in asymptomatic subjects [3]. Celiac disease affects people in all parts of the world. The histologic changes in CD vary from severe villous atrophy to more subtle changes (with or without increased density of intraepithelial lymphocytes and crypt hyperplasia). Although villous atrophy is not specific to CD. Serology has become increasingly relevant to CD diagnosis. Anti-tissue transglutaminase antibodies are the most sensitive test for CD [2]. Celiac Disease is also known as intestinal infantilism, idiopathic steatorrhea, non-tropical sprue, and gluten sensitive enteropathy. Various etiological factors are considered for CD including genetic (HLA class II antigen) and environmental risk factors [4] including GI infections [5]. Gliadin is a glycoprotein extract from gluten, directly toxic to the enterocytes of individuals with CD. Transglutaminase enzyme crosslinks gliadin and causes specific deamination of glutamine into glutamic acid. With such deamination, the gliadin peptides are able to be more efficiently presented to gliadin-reactive CD-4 T cells. Without Transglutaminase, it is believed that gliadin is less immunogenic. Thus, transglutaminase auto antibodies play a role in disease pathogenesis, but lacks sufficient supportive evidence [6]. The epidemiology of CD has iceberg characteristics as there are more undiagnosed cases. The female-to male ratio is 2:1. The prevalence of CD is globally 1%. The prevalence of CD in India is nearly the same as that in Western Caucasian populations [7].

Case Report

Celiac mimicking cirrhosis of liver

A twenty two year old male presented with a prolonged history of vague pain abdomen, bloating and diarrhea for last eight years. He also had symptoms of generalized weakness, easy fatigability distension of abdomen and swelling over bilateral legs for last six months. On evaluation, he was cachexic, frail, and had significant anemia and hypoproteinemia. He had ascites and bilateral pedal edema mimicking cirrhosis of liver with decompensation. His cardiological, respiratory, neurological examination was essentially normal. His serum IgA TTG antibodies were significantly raised i.e. 220 I.U./ml (normal range was below 20 I.U./ml) and endoscopy revealed scalloped duodenal folds, and duodenal biopsies confirmed it to be Marsh grade 3 b celiac disease. He was put on strict gluten restricted diet along with symptomatic treatment with multivitamin, iron and high protein diet. He gradually responded to treatment and within six months, his blood hemoglobin and serum protein levels became normal and his pedal edema and ascites vanished. All his clinical symptoms also improved and he started living a normal healthy life.

Figure 1: *Celiac patient having emaciation, ascites and pedal edema.*

Celiac associated with Sjogren's syndrome

A thirty six year old female, an athlete by profession presented with history of falling of hair, blackening of face and weight loss. She also had symptoms of generalized weakness and easy fatigability for last one year which led to stoppage of her athletic profession completely. On evaluation, she was well built, tall but slightly anemic. Her cardiological, respiratory, neurological and rest systemic examination was essentially normal. Her facial skin was dark in comparison to other parts of body. She had significant loss of hair which were significantly reduced from long hairs to just covering the scalp and were easily pluckable, even during combing of hairs. His serum IgA TTG antibodies were raised to 154 I.U./ml (normal range was below 20 I.U./ml) and endoscopy revealed scalloped duodenal folds, and duodenal biopsies confirmed it to be Marsh grade 3 celiac disease. She was put on strict gluten restricted diet along with symptomatic treatment and gradually responded to treatment. The anemia improved, hair growth normalized and black skin colour partially improved. After few days, she noticed dryness of mouth and difficulty in mastication of food in mouth due to minimal oral secretions. She was further investigated and on autoimmune profile was found to be suffering from Sjogren's syndrome, as her SS-A (RO) and SS-B (LA) antibody were strongly positive. She was put on immunosuppressive by Dental and Rheumatology team which has led to partial improvement in oral symptoms.

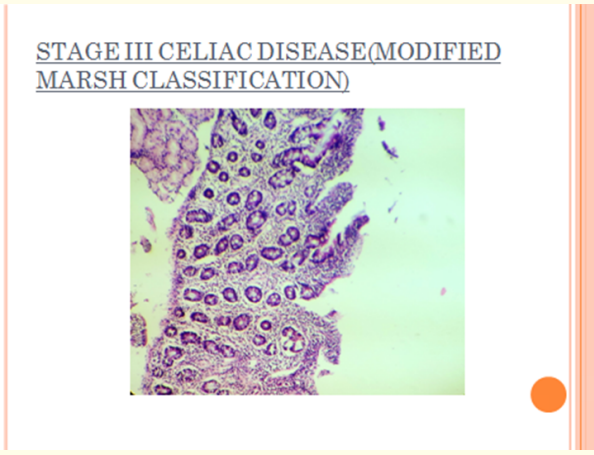

Figure 2: *Showing Marsh stage 3 of celiac disease.*

Celiac associated with ulcerative colitis

A thirty twenty eight year old female presented with history of generalized weakness and diarrhea for last three years. On evaluation, except for pallor, her general and systemic examination including cardiological, respiratory, neurological and rest systemic examination was essentially normal. The complete haemogram was essentially normal, except for mild anemia. The rest biochemical examination was non-contributory. She was subjected to colonoscopy which revealed pan colitis which on histopathological examination proved to be ulcerative colitis. She was started on mesalamine, steroids and immunosuppressive therapy but responded partially. Hence further evaluation was done and serum IgA TTG antibodies were significantly raised i.e. 82.5 I.U./ml (normal range was below 20 I.U./ml) and endoscopy revealed scalloped duodenal folds, and duodenal biopsies confirmed it to be Marsh grade 1 celiac disease. In addition to previous treatment for ulcerative colitis, she was immediately put on strict gluten restricted diet and gradually her stool frequency subsided and within four months, she became totally asymptomatic. At our department, in last twelve years, in total pool of one thousand patients, we have seen four celiac disease patients who have associated ulcerative colitis.

Celiac associated with turner syndrome

A twenty year old female presented with short stature, short and swollen fingers of hands and feet, broad chest with widely spaced nipples and slowed growth. She was diagnosed to be having turner's syndrome. The cardiological, respiratory, neurological and rest systemic examination was normal. Her serum IgA TTG antibodies were elevated i.e. 120 I.U./ml (normal range was below 20 I.U./ml) and endoscopy revealed scalloped duodenal folds and duodenal biopsies confirmed it to be Marsh grade 3 celiac disease. She was put on strict gluten restricted diet but as she had already achieved 20 years of age and in view of turner's syndrome, no improvement occurred in short stature.

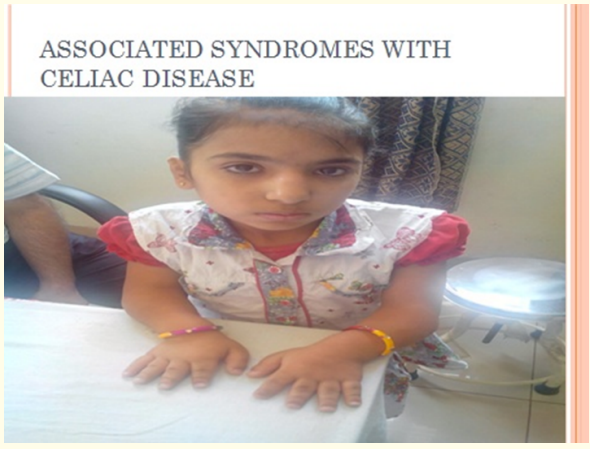

Figure 3: *Showing short stature and fingers in celiac patient.*

Celiac associated with selective iron deficiency anemia

A thirty two year old female, a house wife presented with history of generalized weakness and persistent iron deficiency anemia for which repeatedly blood transfusion and parenteral iron supplementation was given in last seven years. Her iron deficiency anemia was attributed to menorrhagia. On evaluation, except for pallor, her general and systemic examination including cardiological, respiratory, neurological and rest systemic examination was essentially normal. Her serum iron profile was suggestive of iron deficiency as evidenced by low serum iron and increased serum transferrin saturation. The peripheral blood film revealed low hemoglobin of 7 gm% with micro-cytic hypochromic picture and normal total leukocyte and platelet count. serum IgA TTG antibodies were raised to 154 I.U./ml (normal range was below 20 I.U./ml) and endoscopy revealed scalloped duodenal folds, and duodenal biopsies confirmed it to be Marsh grade 2 celiac disease. Surprisingly, this patient on being confirmed to be having celiac disease, brought her twenty eight year old brother who was also having persistent iron deficiency anemia along with diarrhea and was being managed as Irritable bowel syndrome for last fifteen years. He also was proven to be celiac disease on serum IgAttg antibody test, endoscopy and on histopathological examination of duodenal biopsies. Both these brother and sister were put on strict gluten restricted diet and within six months, all symptoms disappeared and their hemoglobin and serum iron parameters normalized and they never needed iron therapy or blood transfusion. At our department, in last twelve years, in total pool of one thousand patients, we have seen twenty celiac disease patients who have presented with selective iron deficiency anemia.

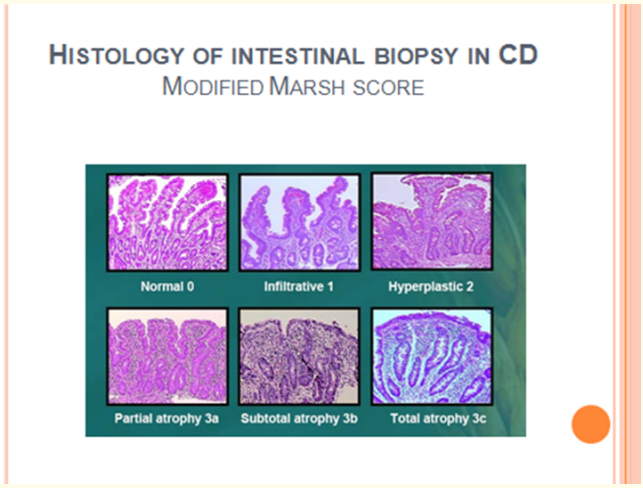

Figure 4: *Showing various stages of celiac on histopathological basis.*

Celiac associated with infertility

At our department, in last twelve years, in total pool of one thousand patients, we have seen five female belonging to age group of 20 - 30 yrs and married for last five to ten years. They were not able to conceive and thus they were being investigated for infertility, along with their life partners. All the tests of both husband and wives were found to be normal and they were being treated as primary infertility. The serum IgAttg antibody test which was done, keeping infertility as rare possibility, was found to be positive in all of them which was further confirmed on Endoscopy and Duodenal biopsies which revealed Marsh Grade 2-3. All these five female patients were put on strict gluten restricted diet and within span of 4 - 6 months of it conceived and two patients delivered by caesarean section and three had normal vaginal delivery. There were no inadvertent complications in both mother and newborn in post-partum or breast feeding period.

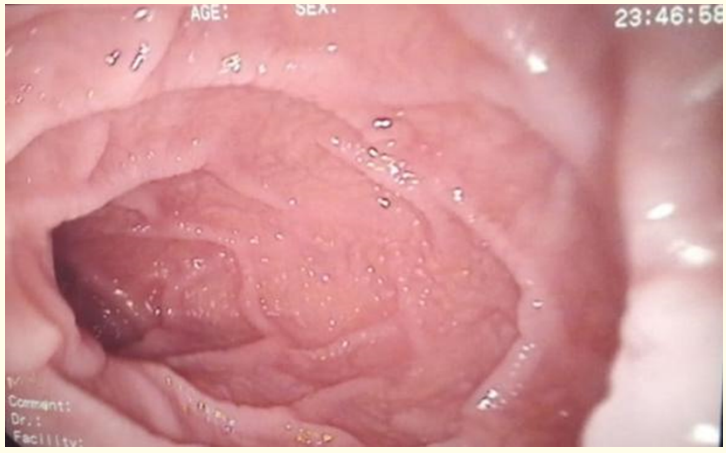

Figure 5: *Showing scalloping of duodenal folds on endoscopy in celiac patient.*

Celiac associated with menstrual problems

At our department, in last twelve years, in total pool of one thousand patients, we have seen fifty female celiac disease patients who have presented with menstrual irregularities like delayed menarche, menorrhagia, menometrorrhagia and oligomenorrhoea. These patients belonged to 14 - 40 yrs of age group and in majority these menstrual irregularities were attributed to anemia which was basically to celiac disease. The strict gluten restriction within 3-6 months showed improvement in 90% of patients.

Celiac associated with hair problems

At our department, in last twelve years, in total pool of one thousand patients, we have seen Ten female celiac disease patients who have presented with hair problems like loss of hair, thinning of hair, loss of texture and easy pluck ability while combing of hair. These patients belonged to 16 - 52 yrs of age group and there was not selective involvement of hairs but other associated manifestations were also seen. The strict gluten restriction within 3 - 6 months showed improvement in hair in all ten patients, as evidenced by normal growth, improvement in texture, no falling of hair during combing. Surprisingly, these hair problem was not seen in any male who formed approximately fifty percent of total pool of patients.

Celiac selectively presenting with constipation

A 62 year old female presented with chief complaint of constipation for last forty years and was evaluated at different centers and battery of tests which were normal at various point of time. She was being managed symptomatically with laxatives, gut motility agents and high fiber diet but for any relief in symptoms. In last forty years, she had been never celiac disease. The Serum IgAttg antibody test done for first time was strongly positive i.e. > 300 I.U. She was immediately put on strict gluten restricted diet and within eight months had substantial relief in symptoms. At present, after six years of diagnosis, she is having just only intermittent constipation and mild dyspepsia symptoms. She was diagnosed to be hypothyroid after three years of being diagnosed to be celiac disease. At our department, in last twelve years, in total pool of one thousand patients, we have seen Fifty two celiac disease patients who have presented with predominantly constipation. All of them have shown significant improvement and in many constipation has completely resolved and there is no need of continuing with laxatives.

Conclusion

Celiac disease can have both typical and atypical presentation, along with many associated diseases. Thus, it has a wide spectrum which require broad vision by treating doctors of various speciality for early recognition of celiac disease, so as to minimize morbidity and mortality associated with it. Once confirmed, then next most important point is to make patient and family members understand the importance of life-long restriction of gluten in diet. It requires proper counseling at time of diagnosis and on regular follow up of patients which increases compliance in celiac patients. There is urgent need of increasing awareness among medical professionals and alternative medicine practitioners, for not restarting gluten in diet under cover of steroids because they temporarily mask the development of symptoms due to re-entry of gluten in body but damage of gluten on intestine goes unabated and even side effects of immunosuppressant's.

Conflict of Interests

The authors declare that they have no conflict of interests.

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Bibliography

1. Harris LA., *et al*. "Celiac disease: clinical, endoscopic and histopathologic review". *Gastrointestinal Endoscopy* 76 (2012): 625-640.
2. Volta U and Villanacci V. "Celiac disease: diagnostic criteria in progress". *Cellular and Molecular Immunology* 8 (2011): 96-102.
3. Serra S and Jani PA. "An approach to duodenal biopsies". *Journal of Clinical Pathology* 59 (2006): 1133-1150.
4. Akobeng AK., *et al*. "Effect of breast feeding on risk of celiac disease: a systematic review and meta-analysis of observational studies". *Archives of Disease in Childhood* 91 (2006): 39-43.
5. Myleus A., *et al*. "Celiac disease revealed in 3% of Swedish 12-year-olds born during an epidemic". *Journal of Pediatric Gastroenterology and Nutrition* 49 (2009): 170-176.
6. Guandalini S. "The influence of gluten: weaning recommendations for healthy children and children at risk for celiac disease". *Nestlé Nutrition Institute Workshop Series: Pediatric Program* 60 (2007): 139-151.
7. Plot L and Amital H. "Infectious associations of celiac disease". *Autoimmunity Reviews* 8 (2009): 316-319.

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