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Mini Review

Celiac Disease - A Multisystemic Disease

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Abstract

Celiac disease is an immune mediated enteropathy triggered by ingestion of gluten containing cereals such as wheat, barley or rye in genetically predisposed individuals. It is also known as celiac sprue or gluten sensitive enteropathy. Affected people may develop intestinal as well as extraintestinal manifestations. Enteropathy associated with symptoms of bloating, chronic diarrhoea, anorexia, weight loss and sequalae of malabsorption while extraintestinal manifestations of disease include iron deficiency anemia, osteoporosis, arthritis, hepatitis, dermatitis herpetiformis or malignancy. The disease is thought to be underdiagnosed, owing to the fact that coeliac disease is often characterized by diarrohea or associated malabsorption conditions. The extraintestinal manifestations are not considered as a part of the disease can misdirect and impede diagnosis.

Keywords: *Celiac Disease; Gluten; Enteropathy*

Coeliac disease is widely regarded as an autoimmune disease that arises from an aberrant immune response towards derivatives of gluten, which is present in wheat, barley and rye, in genetically susceptible people [1,2]. Digestion of wheat, rye and barley may expose the bowel mucosa to immunoreactive epitopes that subsequently initiate a maladaptive immune response [3]. During the process some products may remain undigested on presentation to the small bowel [4] which in turn contribute to the activation of specific populations of T cells in the mucosa [5]. A variety of immune mediators contribute to coeliac disease, these include mast cells, macrophages, plasma cells, eosinophils, CD4+ helper T-cells, CD8+ cytotoxic T cells and natural killer (NK) cells [6]. These T cells produce cytokines that contribute to tissue damage and characteristic mucosal pathology.

Extraintestinal symptoms include abnormal liver enzymes, arthritis, dermatitis herpetiformis, alopecia, fatigue, iron deficiency anemia, stomatitis, myalgias, psychiatric disorders, rashes, seizures, neuropathy, short stature, delayed puberty, osteoporosis, and infertility. Thus, any organ from the central nervous system to joints, liver or teeth can be affected therefore celiac disease is a multisystemic disease.

These extraintestinal manifestations have different pathogenesis such as dermatitis herpetiformis or gluten ataxia has undergone direct autoimmune process whereas others are indirectly related to inflammation or malabsorption including iron deficiency anaemia, osteoporosis, short stature and delayed puberty.

Criteria for the diagnosis of coeliac disease vary. The United European Gastroenterology Week [7] emphasised the importance of small bowel biopsy in the diagnosis of coeliac disease and suggest that circulating antibodies supports the diagnosis on tissue biopsy. The American Gastroenterological Association mandates a biopsy to confirm the diagnosis [8]. Although many still consider the small bowel

biopsy as a "gold standard" in the diagnosis of coeliac disease [8,9]. The US National Institutes of Health [10] recently released a consensus statement recommending biopsies only after a positive serology or when indeterminate serological results.

Our concern regarding review on extraintestinal manifestation of celiac disease is that the celiac disease is a multisystemic disorder rather than a primary gastrointestinal disease. Over the past two decades the percentage of patients presenting in atypical ways, with so-called "silent celiac disease," has increased [11]. The spectrum of disease presentation and severity is varied. Some patients are totally asymptomatic as picked up by screening in at-risk populations, while others may present with a malabsorption syndrome. The reason for the tremendously varied presentation is unclear. However, clinicians need to be aware that celiac disease does not only present with intestinal manifestation and they should be able to recognize the variety of its presentations. The Classical celiac disease is of diarrhea-predominant presentation, whereas silent celiac disease may be atypical which present as iron deficiency or osteoporosis or ataxia or asymptomatic that is detected by screening of at-risk groups, like first-degree relatives or patients with type 1 diabetes or primary biliary cirrhosis.

As in celiac disease any organ from the central nervous system to joints, liver or teeth may be affected. The presence of gastrointestinal symptoms, mainly diarrhea is the key presentation while in few cases, extraintestinal symptoms are the only clinical manifestations of celiac disease or occur in conjunction with diarrhoea and malabsorptive symptoms. An increased awareness among medical practitioners of the variety of extraintestinal manifestations of coeliac disease is essential to improve diagnosis and treatment. Thus, in nutshell Coeliac disease is often accompanied by extraintestinal manifestations, which can be the result of immune responses and malabsorption also. It affect various systems and organs, and include manifestations in the skin, musculoskeletal and central nervous system thus it is considered to be a multisystem disorder. Anaemia, osteoporosis, dermatitis herpetiformis and gluten ataxia are among the most commonly seen characteristics and in paediatric population, coeliac disease can lead to severe growth disorders, such as short stature and delayed puberty due to hypogonadism. Hence clinicians must be updated for the disease manifestations to treat patients promptly before getting complicated.

Conclusion

Thus, celiac disease is a multisystem autoimmune disorder that not only involves gastrointestinal tract but other organ systems such as the skin and bones also. Osteoporosis and osteopenia due to malabsorption of vitamin D are seen commonly in celiac disease along with deficiencies of vitamins A, B12, D, E and K and copper, zinc, folic acid, and iron. Thus, the treating physician must aware of all these clinical findings of celiac disease rather only have tubular vision on gastrointestinal symptoms.

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