Dental Enamel Defects as the Oral Clinical Sign for the Diagnosis of Celiac Disease: A Case Report and Review of Literature

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ABSTRACT

The phrase ‘mouth is a mirror of health and disease’ proves true with the present case of a 10 year old girl with undiagnosed celiac disease who was timely diagnosed with the help of oral signs of dental enamel defects and treated satisfactorily. Systemic diseases often manifest in oral cavity involving the mucosa and/or dentition and thus allow a skilled oral physician to diagnose such cases effectively. Celiac disease, a diagnosis often missed, presents with oral manifestations like dental enamel defects which may range in severity, recurrent aphthous stomatitis, missing teeth and higher incidence of dental caries. Such is the association of oral symptoms of this disease that sometimes dental enamel defects and recurrent aphthous stomatitis may be the only manifestation of the disorder. So oral findings in such cases can serve as a clinical guide for the oral physician to reach a correct and timely diagnosis and provide proper treatment.

Key words: celiac disease; dental enamel defects; gluten; management.

INTRODUCTION

Celiac disease (CD) also known as celiac sprue or gluten sensitive enteropathy is one of the most common chronic gastrointestinal disorders affecting the global population. It is a genetic, immunologically mediated condition characterized by typical intestinal lesions leading to poor digestion and nutrient malabsorption. The disease is precipitated by the consumption of gluten diet (a protein present in wheat, rye and barley). It is a T cell mediated disease resulting in inflammation and atrophy of the mucosa of the small intestine. The prevalence varies from country to country and white individuals are known to be affected more. The incidence of celiac disease in first (approximately 10%) and second degree relatives of the patient is significantly higher when compared to the control population. Mac Donald et al found four biopsy proven cases of celiac disease in a single family, the relatives were not aware of any abnormality because of the lack or mild symptoms. Other high-risk groups include patients with autoimmune disorders, such as type 1 diabetes mellitus, Down syndrome and thyroid disorders.

It manifests itself in sixth month of life, coinciding with the introduction of cereals in the diet. Two types are 1) classical form, which is more frequent and manifests with gastrointestinal presentation (chronic diarrhea, abdominal distension and pain, vomiting and weight loss) and 2) atypical form (“non-gastroenterological”, “silent form”) in which lack of symptoms and presentations make the diagnosis more difficult. In children it presents as delayed growth and puberty, vomiting and dental enamel defects. Secondary immunologic illnesses, such as dermatitis herpetiformis, atopic dermatitis and alopecia may be seen in CD patients as a primary presentation. Highly sensitive and specific serologic tests are available now to screen for celiac disease. Antigliadin (AGA), antireticulin (ARA) and antiendomysium (EmA) antibodies are described as major serological markers. EmA antibodies have been reported to show a high degree of specificity and sensitivity at various stages of the CD disease hence it can be considered as a valid tracer test.

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Distal duodenal biopsy is considered as the gold standard for the diagnosis of CD which demonstrates villous atrophy, increased crypt lengths and number of intraepithelial lymphocytes. This villous atrophy leads to malabsorption of nutrients like iron, calcium, folic acid and fat soluble vitamins. Management of CD is basically dietary, which includes a strict gluten free food which generally results in rapid and complete healing of small-bowel inflammation. Complications such as hepatic diseases, sterility, osteoporosis, endocrinopathies, neurological and psychiatric disturbances and intestinal lymphoma have been reported in untreated CD patients.

**CASE REPORT**

A 10-year-old Indian girl reported to the Department of Oral Medicine and Radiology, Jodhpur Dental College and Hospital, Jodhpur National University, Jodhpur, with a chief complaint of several decayed teeth in her mouth. Physical examination of the patient revealed a height of 160 cms (which was short for that age), a distended abdomen and pallor of eyes. Intraoral examination revealed grossly carious 84 and 85. Other teeth with caries involvement were 54, 55, 16, 62, 63, 64, 65, 26, 36 and 16, clinically 12 was missing. Generalized enamel hypoplasia was also noted, which was severe on maxillary central incisors. OPG confirmed congenitally missing 12. Differential diagnosis of fluorosis, amelogenesis imperfecta, hypoplasia due to systemic disease and chronic infection was considered. Patient’s anamnesis revealed occasional fever, nausea, vomiting, chronic diarrhoea and abdominal bloating. Her parents did not report any history of systemic disease or, in particular, any gastrointestinal symptoms.

A detailed investigation was carried out in co-ordination with a gastroenterologist, the findings were, mild anemia (microcytic, hypochromic), presence of IgA and IgG endomysial antibodies, IgA and IgG gliadin antibodies, IgA and IgG human tissue transglutaminase antibodies in the blood. Ultrasonography of the abdomen revealed mild hepatomegaly. Apart from these findings, small intestine biopsy revealed villous atrophy with crypt hyperplasia and increase in intraepithelial lymphocytes. The findings were suggestive of Celiac disease.

Gastroenterologist instructed the patient to follow a gluten free diet and prescribed iron tablets and multivitamin. Grossly carious teeth were extracted under local anesthesia and other carious teeth were restored with composites. Direct composite veneer restorations were planned for the teeth with hypoplastic enamel defects.
DISCUSSION

Oral cavity which is easily accessible for examination constitutes an integral part of gastrointestinal system and manifests various changes secondary to CD. Most common are recurrent aphthous stomatitis and dental enamel defects. These occur so frequently in a patient with CD that dental enamel defects and recurrent aphthous ulcers are usually considered as the only oral manifestation of this disorder. Angular cheilitis, glossitis and depapillated tongue (due to vitamin B12, folic acid and iron deficiency) are also considered as main oral signs associated with CD. Enamel hypoplasia is a developmental dental defect which manifests due to disturbances during enamel formation. It is considered as a quantitative defect which is associated with reduced thickness of enamel formed during the secretory stage of amelogenesis.

Dental enamel defects (DEDs) are common in children who develop symptoms of celiac disease during the development of permanent dentition. These defects are seen most commonly in the permanent dentition and tend to appear symmetrically and chronologically in all 4 quadrants, with more defects in the maxillary and mandibular incisors and molars as seen in the present case. Defects range from pitting and grooving to complete loss of enamel. The exact mechanism leading to these defects is not clear, but immune mediated damage leading to formation of antibodies against the matrix of enamel organ is suspected to be the primary cause. Nutritional disturbances, including hypocalcemia, may also play a role. DEDs are also found in healthy first degree relatives of patients with celiac disease but no such association was noted in the present case.

A classification of these defects in CD patients was given by Aine et al who graded these defects into 5 types from 0-IV. Our patient presented with grade III enamel defects according to the classification. DEDs are not specific for celiac disease but the possibility of celiac disease should be considered as one of the causes along with the more accepted explanations such as fluorosis, amelogenesis imperfecta, deficiency of vitamin D and chronic infections such as measles, chicken pox and mumps. Studies comparing the prevalence of DEDs in children and adults with celiac disease found that children had higher rate of enamel defects compared to adults.

Even though a wide range of frequencies of enamel defects in CD patients has been reported in literature, there are also studies which did not find any relationship between enamel defects and CD patients.

The prevalence of dental caries in CD patients has been found to vary. While one study found that the prevalence was higher compared to general population, another study found no difference. Other oral alterations found in CD patients are delayed tooth eruption, diminished size of the teeth, and salivary gland dysfunction. A study which aimed to investigate the oral manifestations of CD patients with control group found that percentage of missing teeth was found to be 13.6 %. Also the prevalence was higher in typical CD patients. Our patient also presented with clinically missing 12 which was later found to be congenitally missing.

CONCLUSION

Children with celiac disease may present with more subtle features which may lead to a delay in diagnosis. As the oral cavity is very easy to examine, oral lesions can provide a valuable clinical clue for early diagnosis of unrecognized cases of celiac disease, thus allowing immediate treatment with a gluten free diet and restoring health. It also leads to a reduction in the complications of the disease. Dental enamel defects are not specific for celiac disease but the possibility of celiac disease should be considered as one of the causes of dental enamel defects.
Case Report

REFERENCES


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