Revista de Gastroenterología de México. 2013;78(4):211–218

REVISTA DE GASTROENTEROLOGÍA DE MÉXICO

www.elsevier.es/rgmx

ORIGINAL ARTICLE

Celiac disease in children from the northwest of Mexico: Clinical characteristics of 24 cases☆

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Received 8 February 2013; accepted 31 July 2013
Available online 7 March 2014

KEYWORDS
Celiac disease;
Gluten;
Diagnosis;
Children

Abstract
Background: Celiac disease (CD) is an autoimmune enteropathy induced by dietary wheat gluten that can have serious consequences if not diagnosed and treated early. It is important to be familiar with other alterations associated with gluten ingestion due to the multiplicity of clinical presentations.
Objectives: To describe the most common CD presentation patterns and alterations associated with gluten in children from the northwest region of Mexico, with an incipient knowledge of its prevalence.
Patients and methods: Age, sex, family history, and gastrointestinal and extraintestinal symptoms were recorded in 24 patients within the time frame of 2006 to 2010. Biochemical and hematologic data were collected. Anti-gliadin and anti-transglutaminase antibodies were analyzed in all the cases, and haplotypes (HLA-DQ2/DQ8) and duodenal biopsy were evaluated in some of the cases.
Results: Of the 24 patients (14 girls and 10 boys), 13 presented with typical CD with symptoms of poor gastrointestinal absorption; 7 patients with a mean age of 5 years presented with atypical CD; 2 had disease onset with gastrointestinal and extraintestinal (neurologic) problems; and 2 with other gluten-related disorders. All of the patients had positive serology; 11/15 presented with HLA-DQ2/DQ8 and 4 with at least one allele; damaged mucosa was observed in the 6 biopsies taken. A third of the patients were anemic, 6 presented with an albumin value of < 3.5 g/dL, and 4 with mineral deficiencies. A total of 83% of the patients improved with a gluten-free diet.

☆ Please cite this article as: Sotelo Cruz N, Calderón de la Barca AM, Hurtado Valenzuela JG. Enfermedad celiaca en niños del noroeste de México: características clínicas de 24 casos. Revista de Gastroenterología de México. 2013;78:211–218.

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Conclusions: The presentation patterns were: *1)* typical CD, *2)* atypical CD, *3)* CD with gastrointestinal and extraintestinal (neurologic) symptoms, and *4)* gluten-related disorders other than CD.

PALABRAS CLAVE
Enfermedad celiaca;
Gluten;
Diagnóstico;
Niños

Enfermedad celiaca en niños del noroeste de México: características clínicas de 24 casos

Resumen
Antecedentes: La enfermedad celiaca (EC) es una enteropatía autoinmune inducida por el gluten del trigo dietético, con serias consecuencias si no se diagnostica y trata tempranamente. Hay además otras alteraciones asociadas a la ingestión de gluten, que es importante conocer, por su multiplicidad de presentaciones clínicas.
Objetivos: Describir los patrones más comunes de presentación de EC y alteraciones asociadas al gluten en niños de la región noroeste de México, con incipiente conocimiento de su prevalencia.
Pacientes y métodos: Se registraron la edad, el género, la historia familiar y los síntomas gastro y extraintestinales, en 24 pacientes, entre 2006 y 2010. Se recogieron datos bioquímicos y hematológicos. Se analizaron anticuerpos antigliadinas y antitransglutaminasa en todos los casos; haplotipos (HLA-DQ2/DQ8) y biopsia duodenal en parte de los mismos.
Resultados: De los 24 pacientes (14 mujeres y 10 varones), 13 presentaron EC típica con síntomas de mala absorción gastrointestinal; 7 promediando 5 años de edad, con EC no típica; 2 iniciaron con problemas gastro y extraintestinales (neurológicos), y 2 con otros desórdenes asociados al gluten. Todos presentaron serología positiva; 11/15 presentaron HLA-DQ2/DQ8 y 4 al menos un alelo; las 6 biopsias tomadas, mostraron mucosa dañada. Una tercera parte estaban anémicos, 6 con albúmina < 3.5 g/dL, 4 con deficiencias de minerales. El 83% de los pacientes mejoró con la dieta sin gluten.
Conclusiones: Los patrones de presentación fueron: *1)* EC típica; *2)* EC no típica; *3)* EC con síntomas gastro y extraintestinales (neurológicos), y *4)* sin EC, con otros desórdenes relacionados con el gluten.

Introduction

Celiac disease (CD) is systemic, it is mediated immunologically, precipitated by exposure to dietary gluten, and it develops in genetically susceptible individuals. It is characterized by diverse clinical manifestations, specific antibodies, haplotypes HLA-DQ2 and DQ8, and enteropathy.[1,2]

CD presents with a wide variety of non-specific signs and symptoms that can be gastrointestinal or extraintestinal and thus has been classified as typical, atypical, asymptomatic, subclinical, and potential.[2–4] Typical pediatric CD is characterized by delayed growth, diarrhea, emaciation, loss of appetite, and abdominal bloating; when it appears with any other sign or symptom it is atypical. Asymptomatic, or silent, CD does not present with clinically suspicious signs or symptoms, and is subclinical when it is below the threshold of clinical detection. Potential CD, also called latent CD, is defined by the presence of compatible antibodies and haplotypes, but with no abnormalities in the duodenal mucosa; it may or may not present with symptoms or enteropathy.[1,2]

There is also a recently recognized entity called non-celiac sensitivity or hypersensitivity to gluten.[2,5] It is characterized by clinical symptoms (gastrointestinal) that are very similar to those of CD, the patients present with positive anti-gliadin antibody titers, but negative anti-transglutaminase antibody titers.[5] They do not present with atrophy of the intestinal villi, but they do present with eosinophil infiltration into the duodenal and colonic mucosa; in addition, they do not present with allergies linked to IgE. In many cases of hypersensitivity, the patients have the HLA-DQ2 and DQ8 haplotypes, and in all cases their symptoms are resolved by a gluten-free diet.

Refractory CD is also well characterized and is defined by its symptoms of persistent or recurrent malabsorption and atrophy of the intestinal villi, despite a strict gluten-free diet during 6-12 months.[2,6] Refractory CD can be type 1 or type 2. In type 1, the patients do not respond to a gluten-free diet, but their intraepithelial lymphocytes are normal. Type 2 is characterized by clones of abnormal intraepithelial lymphocytes that do not present with the CD3 and CD8 markers and the T-cell receptor, but they express CD3 intracellularly. It is associated with poor outcome due to the fact that it can progress to T-cell lymphoma.[6,7]

With this diversity of signs and symptoms, the diagnosis of CD and other gluten-associated alterations tends to be complicated, obscuring the dimension of the problem. It is estimated that between 1:100 and 1:200 individuals of any given population suffer from some form of CD; however, there are differences among the published data. For example, for the Mexican population, there was a 1:37 prevalence of the IgA anti-transglutaminase antibody resulting from the analysis of serum from healthy donors;[8] when the serum was reanalyzed for anti-endomysial antibodies, prevalence decreased to 1:168[9]. In contrast, in a recent study conducted on 7,798 persons in the United States, 1:141 were positive for celiac disease, whereas among 1,686 Mexican Americans in the same study using the same serologic indicators, not a single individual was positive.[10]