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ORIGINAL ARTICLE

Autoimmune hepatitis in children: Progression of 20 cases in northern Mexico[☆]



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KEYWORDS

Autoimmune hepatitis;
Children;
Progression;
Liver biopsy;
Treatment

Abstract

Background: Autoimmune hepatitis (AIH) is a chronic inflammatory disease of the liver with nonspecific clinical manifestations that causes greater liver damage in children than in adults.

Aims: To analyze the clinical progression, biochemical profiles, histopathologic changes, and treatment response in 20 children with AIH.

Material and methods: A retrospective study was carried out on the variables associated with clinical progression, diagnosis, and treatment response in children seen at the the *Unidad Médica de Alta Especialidad (UMAE) No. 71 IMSS* in Torreón, Coahuila, Mexico, from 1992 to 2012.

Results: Twenty patients were analyzed, 75% with type 1 AIH (AIH-1) and 25% with type 2 AIH (AIH-2). Girls predominated with a 3:1 ratio of girls to boys. The mean age was 10.07 ± 6.53 years for the AIH-1 cases and 6.75 ± 3.77 years for the AIH-2 cases. There was an association with immunologic diseases in 40% of the patients.

The patients in the AIH-2 group had greater biochemical profile alterations and IgA deficiency. Anti-nuclear antibody and anti-smooth muscle antibody were positive in 100% of the patients with AIH-1, and anti-liver kidney microsomal type 1 antibody was positive in 100% of the AIH-2 patients. Liver biopsy revealed interface hepatitis in both groups. The AIH-2 group responded more quickly to treatment, but had a higher recurrence rate.

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Conclusions: Autoimmune hepatitis in the pediatric patient should be suspected in order to make an early diagnosis and thereby establish opportune treatment. Determining the type of AIH is necessary for making adequate diagnosis and for achieving a better outcome in relation to recurrence and complication rates.

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PALABRAS CLAVE

Hepatitis autoinmune;
Niños;
Evolución;
Biopsia hepática;
Tratamiento

Hepatitis autoinmune en niños: evolución de 20 casos del norte de México

Resumen

Antecedentes: La hepatitis autoinmune (HAI) es una enfermedad inflamatoria crónica del hígado con manifestaciones clínicas inespecíficas y mayor daño hepático en niños que en adultos.

Objetivo: Analizar la evolución clínica, los perfiles bioquímicos, los cambios histopatológicos y la respuesta al tratamiento de 20 niños con HAI.

Material y métodos: Estudio retrospectivo de las variables asociadas a evolución clínica, diagnóstico y respuesta al tratamiento en niños atendidos en la UMAE n.º 71 IMSS Torreón, Coahuila, México, en el período comprendido entre 1992 y 2012.

Resultados: Se analizó a 20 pacientes, el 75% con HAI-1 y el 25% con HAI-2. Se observó predominio del sexo femenino 3:1. La edad promedio \pm desviación estándar fue de 10.07 ± 6.53 años para HAI-1 y de 6.75 ± 3.77 para HAI-2. La asociación a enfermedades inmunológicas fue del 40%.

Los pacientes del grupo HAI-2 mostraron mayores alteraciones en su perfil bioquímico y deficiencia de IgA. La positividad para anticuerpos antinucleares y anticuerpos antimúsculo liso fue del 100% en los pacientes con HAI-1, anticuerpos antimicrosomales para hígado y riñón tipo 1 en el 100% de HAI-2. Las biopsias hepáticas mostraron hepatitis de interfase en ambos grupos. Los pacientes del grupo HAI-2 respondieron más rápidamente al tratamiento y tuvieron mayor tasa de recaídas.

Conclusiones: Es necesario sospechar la HAI en pediatría para poder realizar un diagnóstico temprano y así establecer el tratamiento oportuno. Determinar el tipo de HAI nos permitirá establecer el diagnóstico adecuado y elaborar un mejor pronóstico respecto a tasas de recaídas y complicaciones.

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Introduction

Autoimmune hepatitis (AIH) is a chronic inflammatory process involving cell destruction and fibrosis. It predominantly affects women and is characterized by an increase in transaminases and immunoglobulins, in particular immunoglobulin G (IgG).

AIH has 2 main variants: type 1 (AIH-1) and type 2 (AIH-2). AIH-1 is the most common form, affecting children and adults, and is characterized by the presence of anti-nuclear antibody (ANA) and anti-smooth muscle antibody (SMA). AIH-2 mainly affects children and young adults and is characterized by the presence of anti-liver/kidney microsomal type 1 antibody (LKM-1). The diagnostic criteria for AIH were published by the International Autoimmune Hepatitis Group in 1993,¹ in 1999,² and simplified in 2008.^{3,4}

These criteria include: the exclusion of infectious viral processes, antibody profiles (ANA, SMA, LKM-1), IgG levels 1.10-fold above the normal limits, and a histologic pattern of interface hepatitis with plasma cells and rosette formation.

Acute AIH presents in children and young adults and is more aggressive than in older adults.^{5,6}

Aims

The aim of this study was to analyze the clinical progression, biochemical profiles, histopathologic changes, and treatment response in 20 children presenting with AIH.

Methods

Twenty children diagnosed with AIH that were seen at the outpatient service of the Department of Gastroenterology and Pediatric Nutrition of the Advanced Speciality Medical Unit No. 71 of the *Instituto Mexicano del Seguro Social* within the time frame of 1992 and 2012 were studied. In each case, the disease was classified in accordance with the criteria of the International Autoimmune Hepatitis Group (IAIHG) and reclassified with the 2008 simplified diagnostic

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