

Autoimmune Hepatitis in Children, Clinical Features and Biochemical of Iranian Children

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ABSTRACT

Background

There are only a very small number of reports which discuss subtype, sex distribution, clinical features and laboratory characteristics of autoimmune hepatitis in children. The aim of this study was both to define the clinical features, biochemical and histological findings and also to determine the age and sex related distribution of autoimmune hepatitis (AIH).

Materials and Methods

Data of 30 children presenting with AIH (20 girls and 10 boys) have been analyzed for their clinical, serological, and histological profile. The most common presenting signs or symptoms were jaundice (60%), abdominal mass (23.4%) and constitutional symptoms (weakness, anorexia and paleness) (6.7%).

Results

About 10% of patients had an acute hepatitis like clinical presentation. Twenty two children (73.3%) (15 girls, 7 boys, 2.1:1) had AIH type 1 and 4 patients (13.3%) type 2 due to specific autoantibodies. Four children could not be classified. In liver biopsy, 100% of patients had interface hepatitis and fibrosis with or without cirrhosis were found in 60%.

Conclusions

In our cohort the prevalence of AIH was 2:1 in girls. Type 1 was the most frequent diagnosis (73.3%) and was more prevalent in older children. Patients with type 2 were younger. The clinical presentation of AIH in children was unspecific and each type could only be differentiated by the determination of the specific autoantibodies.

Keywords: Autoimmune hepatitis type 1, type 2; Children; Autoimmune liver disease

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INTRODUCTION

Autoimmune liver disease in children progress to cirrhosis and liver failure if not diagnosed in time and treated with immunosuppressant.(1-3), This disease usually has an insidious onset and a slow progressive course. Despite viral infections, as possible trigger factors and the frequent presence of some specific HLA allotypes, reflecting a

genetic background, etiology finally remains unknown.(4-6), According to the pattern of detected antibodies two major forms of autoimmune hepatitis are differentiated. Autoantibodies characterizing type 1 are anti-nuclear antibodies (ANA), anti-smooth muscle antibodies (SMA) and anti-soluble liver protein antibodies (SLA). Type 2 is defined by the detection of liver kidney microsomal antibodies (LKM1) and/or liver cytosol 1 antigen (LC1) antibodies.(1), The prerequisite of diagnosis is the exclusion of other known causes of liver disease and the presence of one of the above autoantibodies.

Unfortunately many patients do not reveal

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