Distinction in Clinical Manifestations and Outcomes between Probable and Definite Autoimmune Hepatitis by Revised Original Diagnostic Scoring Criteria

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Background: The diagnostic scoring systems for autoimmune hepatitis (AIH) categorize some patients as having probable disease. This designation can affect treatment strategies and outcomes.

Objective: To define the clinical phenotype of probable AIH, and to evaluate the distinctions between patients with probable and definite AIH diagnoses by the revised original scoring system of the International AIH Group.

Methods: This was a retrospective cohort study. During 2005-2015, 358 patients satisfied the codified clinical criteria for the diagnosis of AIH at presentation. The comparison of survival between the groups was performed using the Cox proportional model.

Results: A total of 208 patients (mean age 58±15 years and female 89%) were graded as having probable AIH. The mean pretreatment score for AIH by this system was 13 points (range, 10-15 points; median 14 points). Patients with probable AIH were distinguished from 150 patients designated as having definite AIH by fewer concurrent immune diseases (2.9% vs 15.3%, p<0.001), lower serum globulin (median 4.4 vs 4.8 g/dL, p=0.001), immunoglobulin G levels (median 2,530 vs 2,986 mg/dl, p=0.002), lower frequency of high serum titers (>1:80) of antinuclear antibodies (75% vs 91%, p<0.001), and smooth muscle antibodies (27% vs 41%, p=0.02). The frequency of cirrhosis at presentation was similar in patients with probable and definite AIH at presentation (43% vs 37%, p=0.3). A number of 332 patients with probable (n=186) or definite (n=146) AIH were treated with high-dose prednisolone alone or low-dose in conjunction with azathioprine. The rate of full response was lower in patients with probable AIH compared to those with definite AIH (32% vs 42%, p=0.046). The frequencies of partial response (26% vs 33%, p=0.2) and no response (8.2% vs 4.7%, p=0.2) were comparable between patients with probable and definite designations. Twenty-six patients with probable AIH and six patients with definite AIH died during the median follow-up of 4.3 years (interquartile range, 1.9-8.2 years). Patients with probable AIH had higher mortality rate compared to those with definite AIH (hazard ratio, 3.33; 95% CI, 1.37-8.09, p=0.008).

Conclusion: The pretreatment classification of AIH as probable or definite by the revised original diagnostic scoring system can characterize subgroups of the disease with different severity, treatment response, and prognosis.

Keywords: Definite autoimmune hepatitis, Probable autoimmune hepatitis, Pretreatment classification, revised original scoring system, cirrhosis, full response, partial response, no response