Seronegative autoimmune hepatitis in children

A real diagnostic challenge

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Summary

Background and aim Classical autoimmune hepatitis (AIH) is characterized by the presence of conventional autoantibodies (anti-smooth muscle, antinuclear and anti-liver-kidney-microsomal antibodies). The absence of such autoantibodies in some patients does not preclude AIH diagnosis or the need for its treatment. This group of patients was termed seronegative AIH. Whether non-conventional autoantibodies can identify this group of patients is still elusive. We aimed to study the prevalence of seronegativity of conventional autoantibodies and the occurrence of non-conventional autoantibodies in children with AIH.

Methods In this study, 55 children with AIH were investigated for non-conventional autoantibodies (antineutrophil cytoplasmic antibodies, antibodies to soluble liver antigen, anti-tissue transglutaminase and antiplatelet antibodies). All the patients received immunosuppressive therapy and were assessed for treatment response.

Results Of the patients 44 had classical AIH (type1, 70.09%, type 2, 9.09%) and 20% were seronegative. The four studied non-conventional autoantibodies occurred in four patients, one for each. All nonconventional autoantibodies were exclusively associated with type 1

AIH. The clinical profile, ultrasonographic findings, liver biochemistry and histopathological findings were comparable in the classical and seronegative AIH. The majority of patients with classical (72.7%) and seronegative (54.5%) AIH were treatment responders.

Conclusion Seronegative AIH represents a substantial percentage of pediatric patients diagnosed with AIH. They were even negative for non-conventional autoantibodies. Furthermore, apart from autoantibodies, seronegative AIH is almost indistinguishable from the classical AIH and the majority of patients were treatment responders. This favorable response to immunosuppression deserves sustainable efforts for considering such a diagnosis and start therapy to halt disease progression is worthwhile.

Keywords Autoantibodies · Conventional ·Non-conventional Corticosteroids · Pediatrics