

Autoimmune hepatitis as a complication of common variable immunodeficiency

Key words: Autoimmune hepatitis. Common variable immunodeficiency.

Dear Editor,

Common variable immunodeficiency (CVID) is associated with a digestive morbidity, particularly diarrheal syndrome. Some cases have been previously reported in this journal (1,2). Liver involvement is frequent, but less reported. Herein we present a case of autoimmune hepatitis associated with CVID.

Case report

A 28-year-old female was admitted due to a condition compatible with acute hepatitis. She was diagnosed with CVID three years previously and had not received specific treatment. There was an increase in the levels of glutamic-pyruvic transaminase (GPT) to 1,474 IU/l and bilirubin at 3.23 mg/dl, in the absence of coagulopathy. An etiological investigation that included autoantibodies and HBV/HCV serology was negative and the IgG levels were normal. In addition, she had neutropenia, and a wide range of studies were performed that included a bone marrow biopsy, but there was no definitive diagnosis. The liver biopsy identified persistent chronic hepatitis, without a definite etiology or fibrosis. Therefore, a possible toxic hepatitis in relation to NSAIDs was suspected. There was a spontaneous biochemical improvement, with occasional transient transaminase level increases up to 2-4 times the normal limit and also oscillating neutropenia. Three years after the initial flare, the transaminase levels increased to 12 times above normal levels. The ultrasound identified homogenous hepatomegaly and splenomegaly. A second liver biopsy was performed that was compatible with autoimmune hepatitis with F1 fibrosis. She started treatment with budesonide 9 mg/day and there was a normalization of transaminase levels for more than two years during follow-up (Fig. 1). In addition,

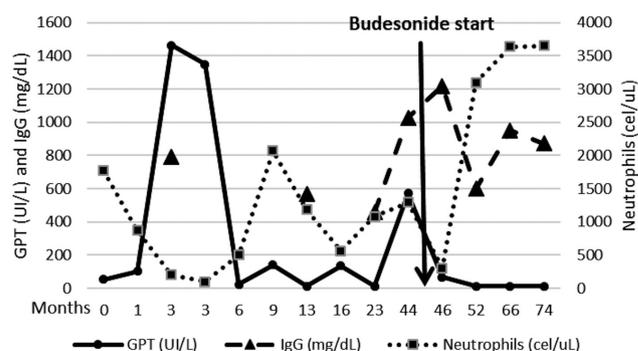


Fig. 1. Analytical evolution: an increase in transaminase levels and a decrease in neutrophils was observed, with a temporal coinciding increase in IgG that was due to the autoimmunity phenomena.

there was a sustained neutrophil normalization. Currently she is under treatment with budesonide at 3 mg/day.

Discussion

Liver involvement is a common complication of CVID, frequently with nodular regenerative hyperplasia and granulomatous disease (3,4). There are a few reported cases of autoimmune hepatitis associated with CVID, thus making the diagnosis difficult as autoantibodies are usually negative and IgG levels are normal or low. Corticosteroid use has been described as a treatment option for autoimmune disorders in the CVID (5). In this case, budesonide resulted in a control of autoimmune hepatitis and a normalization of the neutrophil level.

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