IgG4-related disease mimicking a liver abscess

Keywords: IgG4-related disease. Liver abscess. Inflammatory pseudotumor.

Dear Editor,

In recent years, IgG4-related disease (IGRD) has emerged as a fascinating and complex medical condition that affects various organs, including the liver. This immune-mediated disorder is characterized by excessive production of IgG4 antibodies, leading to chronic inflammation and tissue damage.

Case report

A 37-year-old male with a medical history of chronic pancreatitis of unknown etiology presented to the Emergency Department with a four-week history of fever, right upper quadrant abdominal pain, and anorexia. He was an ex-smoker and had no history of alcohol consumption or trips to endemic areas.

The physical examination was unremarkable. Laboratory test results showed C-reactive protein 23 mg/dl (< 0.5 mg/dl), 9.70 x 103 leukocytes (3.9-11.00), total bilirubin 1.9 mg/dl (< 1.2 U/l), alkaline phosphatase 174 U/l (40-150 U/l), gamma-glutamyl transpeptidase 127 U/l (12-64 U/l), and normal aminotransferases. Abdominal computed tomography (CT) revealed a 10-cm heterogeneous mass involving liver segments VII and VIII, along with smaller lesions that were aimed at segments V and VIII (Fig. 1A). The biliary tree was normal. The initial diagnosis was a possible liver abscess, and treatment with piperacillin and tazobactam 4 g every eight hours plus metronidazole 500 mg every eight hours was started. The patient evolved unfavorably with persistent fever, so he was admitted to our center to complete the study. Serologic tests were negative for *Entamoeba histolytica* and *Echinococcus.* Blood and urine cultures were also negative. Tumor markers were normal. Finally, an ultrasound-guided biopsy of the lesion was performed. Histology revealed increased IgG-4 positive cells identified by immunohistochemical studies (Fig. 1B and C). The anaplastic lymphoma kinase - fluorescence *in situ* hybridization (ALK FISH) study was negative. IgG4 levels were also elevated at

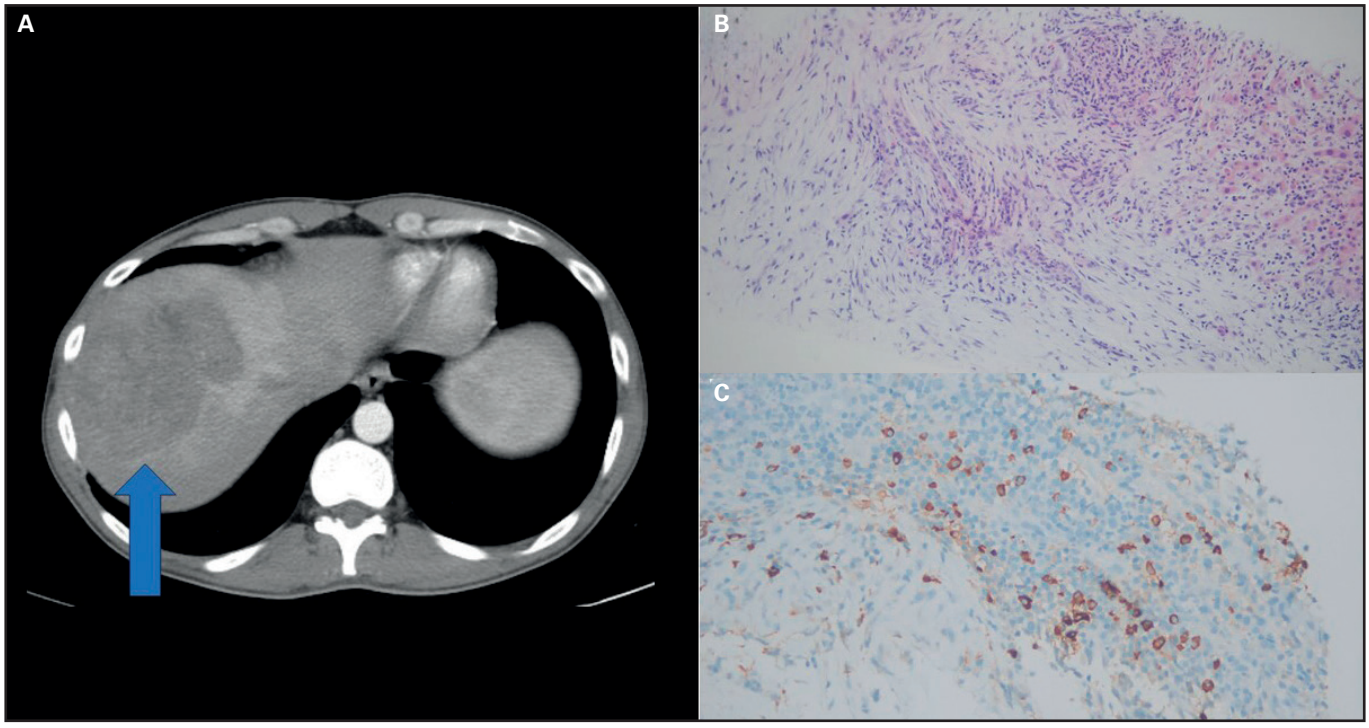

Fig. 1. *A. Abdominal computed tomography (CT) showing a 10-cm heterogeneous liver mass involving liver segments VII-VIII (blue solid arrow). B. Image of a liver mass at 20x magnification hematoxylin and eosin stain with fibroinflammatory proliferation and abundant plasma cells. There was no evidence of microorganisms. C. IgG4 immunohistochemistry from a liver mass. The brown staining shows positivity in a significant number of plasma cells, with > 10 plasma cells per high-power field (40x magnification).*

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236 mg/dl (8-140 mg/dl). Therefore, a diagnosis of an inflammatory pseudotumor associated with IGRD was made. He started prednisone taper therapy and follow-up abdominal ultrasound revealed complete resolution of the liver lesion after six months of corticoid treatment.

Discussion

IGRD is an uncommon autoimmune condition that appears as increased IgG4 levels, inflammation, and organ fibrosis. Its clinical symptoms will depend on the affected organs, and it might mimic other diseases like neoplasms. As soon as it is detected, treatment must be initiated to stop fibrosis and must be continued once remission is reached to prevent relapses. The main treatment is systemic glucocorticoids. It is essential to be aware of this disease, be suspicious of it, and obtain a proper diagnosis to prevent irreversible fibrosis and organ failure that follow.

Conflict of interest: the authors declare no conflict of interest.

Artificial intelligence: the authors declare that they did not use artificial intelligence (AI) or any AI-assisted technologies in the elaboration of the article.

Pere Vaquer-Grimalt[1], Irene Micaela García[2], Ester Antón[2], Aina Escarda[1], Margalida Vanrell[1], Lucía Bonet[1], Lydia Sastre[1]
Departments of [1]Hepatology and [2]Gastroentelogy. Hospital Universitario Son Espases. Palma de Mallorca

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