

Case Report

The Diagnostic Path beyond Liver Biopsy

Irene Verdasca¹, Susana Franco², Luis Melo³, João Torres⁴,
Susana Peres⁴, Fernando Borges⁴, and Kamal Mansinho⁴

¹Department of Internal Medicine, Hospital São Francisco Xavier, Portugal

²Department of Internal Medicine, Hospital Beatriz Angelo, Portugal

³Department of Internal Medicine, Hospital Fernando da Fonseca, Portugal

⁴Department of Infectious Diseases, Hospital Egas Moniz, Portugal

*Corresponding author

Irene Verdasca, Department of Internal Medicine, Hospital São Francisco Xavier, Rua 2 de Abril 24, Évora, 7005-273, Portugal, Tel: 351 962818918; Email: ireneverdasca@hotmail.com

Submitted: 18 December, 2019

Accepted: 08 January, 2019

Published: 10 January, 2019

Copyright © 2020 Verdasca I, et al.

ISSN: 2373-9819

OPEN ACCESS

Keywords

• Autoimmune hepatitis; liver biopsy; Liver tests; Immunosuppression; Simplified autoimmune hepatitis score

Abstract

Autoimmune hepatitis (AIH) remains a major diagnostic and therapeutic challenge. In addition to being a relatively rare disease, early recognition may be difficult due to its heterogeneous clinical picture and the absence of a specific laboratory finding. Female patient, 67 years old, with human immunodeficiency virus infection under antiretroviral therapy with stable immune status and suppressed viral load. Reported asthenia, yellowish mucosa and nausea for one month. Lived in a rural area with direct contact with unvaccinated animals, consumed unpasteurized food and had occasional contact with rat poison. Laboratory evaluation showed liver biochemical and function tests highly elevated and the autoimmune study positive antinuclear antibody (ANA) and anti-smooth muscle antibody (ASMA). Other causes of inflammatory liver diseases were excluded. Abdominal ultrasound showed no abnormal findings. Liver biopsy (Figure 1) was preceded and revealed chronic hepatitis with marked activity that could be compatible with autoimmune or toxic etiology (Figure 2). By the use of simplified AIH score, diagnosis of AIH was likely (7 points) and probable by using the revised original score for AIH (14 points). The patient started prednisolone 1mg / kg / day and azathioprine 25mg / day with rapid remission. The simplified diagnostic criteria have a high sensitivity and specificity for the diagnosis of AIH. Its application in clinical practice led to the diagnosis of HAI after the result of a non-definitive liver histology and the initiation of immunosuppressive therapy, in this case with remission of the disease.

ABBREVIATIONS

AIH: Autoimmune Hepatitis; ANA: Antinuclear Antibody; ASMA: Anti-Smooth Muscle Antibody

INTRODUCTION

Autoimmune hepatitis (AIH) remains a major diagnostic and therapeutic challenge. In addition to being a relatively rare disease, early recognition may be difficult due to its heterogeneous clinical picture and the absence of a specific laboratory finding.

CASE PRESENTATION

Female patient, 67 years old, reported symptoms of asthenia, nausea and yellowish mucosa present for one month. She was diagnosed with human immunodeficiency virus (HIV) infection in 2010, treated with emtricitabine/tenofovir and raltegravir with good virological and immunological control (viral load <20 copies / mL, Lymphocytes TCD4 + 720 Cells / uL). Of particular note the patient lived in a rural area with direct contact with unvaccinated animals, consumed unpasteurized food and had occasional contact with rat poison. At physical examination presented jaundiced mucosa and abdominal pain on the on palpation on the right hypochondrium.

Laboratory evaluation was carried out and showed highly elevated liver biochemical and function tests: total bilirubin

23mg/dl (<0.9), aspartate aminotransferase (AST) 1969 U/L (<32), alanine transaminase (ALT) 871 U/L (>33), alkaline phosphatase 265 (<104U/L) and gamma glutamyl transferase (GGT) 153 (<42). No other organ/system dysfunction was present. Serum protein electrophoresis was normal. Other causes of viral acute hepatitis were excluded (hepatitis A, B, C, D, E, herpes simplex virus, varicella zoster virus, Epstein-Barr virus, cytomegalovirus serology were negative) as well as zoonotic infection, attending to the social context (serology for *leptospira*,

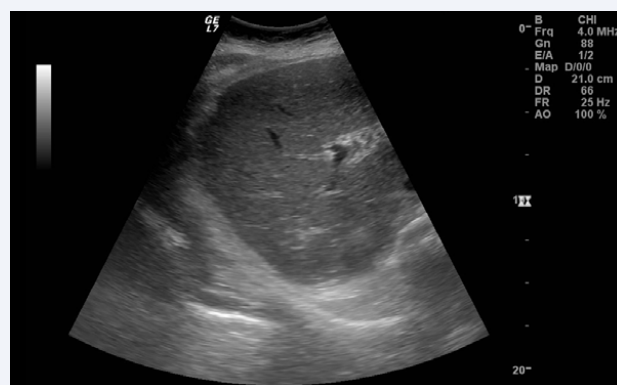


Figure 1 Patient's abdominal ultrasound, no abnormal findings were detected in liver, gallbladder or intra or extrahepatic bile ducts.



Figure 2 Liver biopsy kit. The kit contains a syringe, a sharpened needle without the mandrel for hepatic tissue aspiration, a needle used for the syringe filling with a biocompatible liquid necessary for cleaning the internal needle before the tissue perforation and a scalpel.

Coxiella burnetti, *Bartonella* spp, *Borrelia burgdoferi*, *Rickettsia* spp and *Brucella* spp was also negative). Iron overload was also excluded by normal serum ferritin and transferrin saturation. The autoimmune initial study stood out for positive antinuclear antibody (ANA) (+ 1: 320) and *anti-smooth muscle antibody* (ASMA).

Abdominal ultrasound showed no abnormal findings in liver, gallbladder or intra or extra hepatic bile ducts.

Due to suspicion of AIH because of ANA and ASMA positive antibodies, although normal serum protein levels and subtle, unspecific clinical presentation, investigation was continued with liver biopsy. Microscopic evaluation showed cylinder of hepatic tissue with marked reinforcement of reticulin structure and moderate portal fibrosis with focal septa outline. Gate spaces with ductile proliferation and moderate to intense inflammation with interface activity, rich in lymphocytes, rare plasmocytes and some polymorph nuclear cells, namely eosinophils. Hepatocytes with signs of intracellular cholestasis and multiple foci of intralobular necrosis; undiluted sinusoids containing rare lymphocytes; negative Perl's. The histologic diagnosis was then chronic hepatitis with marked activity that could be compatible with autoimmune or toxic etiology.

Attending to this unclear diagnosis and laboratory and histologic evidence of hepatitis and urgent treatment institution we applied simple diagnostic scores to consolidate and establish a diagnosis. By using the simplified AIH score, the diagnosis of AIH was likely (7 points) and probable by using the revised

original score for AIH (15 points). As soon as the diagnosis was established the patient started immunosuppressive treatment with prednisolone 1mg / kg / day and azathioprine 25mg / day with rapid remission.

After one month, six months and one year of follow-up the patient is asymptomatic, with normal liver biochemical tests, maintaining azathioprine and prednisolone in weaning dose.

DISCUSSION

AIH is a chronic, inflammatory liver disease with a variety of clinical phenotypes, included abnormal liver biochemical tests, acute hepatitis, cirrhosis, or acute liver failure [1] and it might range from asymptomatic or mild, subtle disease to an acute and fulminant hepatic failure. Diagnosis of AIH can be therefore challenging and an early diagnosis is important because immunosuppression is life-saving [2]. In the very acute presentation of AIH, the histological findings may be unclear or similar to drug-induced liver injury with centrilobular necrosis. At the same time, IgG levels may not be yet elevated as in our patient. The simplified score for the diagnosis of AIH, using only four criteria (presence of autoantibodies, immunoglobulin G, histology, and exclusion of viral hepatitis), establishes a probable diagnosis of AIH when the total points are 6 (88% sensitivity and 97% specificity) and a likely or definitive diagnosis when the total points are ≥ 7 (81% sensitivity and 99% specificity) [3]. The use of simple diagnostic scores on our case helped to establish the diagnosis and the decision to start immunosuppressive treatment.

The diagnosis of AIH is often difficult and needs the evaluation of several parameters not always available at the bedside of the patient [4]. The simplified diagnostic criteria, although not yet validated in prospective studies, is easy to apply and have a high sensitivity and specificity for the diagnosis of AIH [3,4]. Its application in clinical practice led to the diagnosis of AIH after the result of a non-definitive liver histology and the initiation of immunosuppressive therapy, in this case with remission of the disease.

REFERENCES

1. Wang Q, Yang F, Miao Q, Krawitt EL, Gershwin ME, Ma X. The clinical phenotypes of autoimmune hepatitis: A comprehensive review. *J Autoimmun.* 2016; 66: 98-107.
2. Hennes EM, Zeniya M, Czaja AJ, Parés A, Dalekos GN, Krawitt EL. Simplified criteria for the diagnosis of autoimmune hepatitis. *Hepatology.* 2008; 48: 169-176.
3. Yeoman AD, Westbrook RH, Al-Chalabi T, Portmann BC, Ma H. Diagnostic value and utility of the simplified International Autoimmune Hepatitis Group (IAIHG) criteria in acute and chronic liver disease. *Hepatology.* 2008; 48: 1094A.
4. Muratori P, Granito A, Pappas G, Muratori L. Validation of simplified diagnostic criteria for autoimmune hepatitis in Italian patients. *Hepatology.* 2009; 49: 1782-1783.

Cite this article

Verdasca I, Franco S, Melo L, Torres J, Peres S, et al. (2020) The Diagnostic Path beyond Liver Biopsy. *JSM Clin Case Rep* 8(1): 1171.