

Autoimmune hepatitis with cholestasis; An uncommon presentation in elderly

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Abstract

Auto immune Hepatitis (AIH) with cholestatic jaundice is a rare entity. AIH is characterized by inflammatory changes in liver with the presence of circulating auto antibodies and hyperglobulinemia. Here we report a case of 67-years-old lady who presented with Acute autoimmune hepatitis with cholestatic jaundice with early accurate diagnosis and early initiation of corticosteroids recovered successfully. Even though it is very rare AIH should not be overlooked in patients with cholestatic jaundice without viral/drug induced hepatitis.

Keywords: Auto immune hepatitis, cholestatic jaundice

Introduction

Auto immune Hepatitis (AIH) is a rare chronic liver cell disease with interesting variety of clinical manifestations suspected based on the presentation, biochemical findings, specific serological tests and liver histology with exclusion of other causes of chronic liver cell disease. AIH has a female predominance with bimodal age distribution with two peaks occurring in 2nd and 5th decade⁽¹⁻²⁾

Three types of AIH have been recognized. Among them the most common form, type 1, predominantly occurring in females and seropositive for both antinuclear antibody (ANA) and/or anti-smooth muscle antibody (SMA). AIH type 2, male-dominant and seropositive for anti-liver/kidney microsome type 1 (anti-LKM-1) antibodies. Type 3 AIH, antibodies against soluble liver antigen/hepatic pancreatic antigen (SLA-LP).

Case history

A 67 years old lady transferred from DGH-Mullaitivu for further evaluation of progressive painless jaundice, dark urine and decoloured stools for 2 weeks. There was no weight loss but she had anorexia during the illness. She denied nausea, vomiting, abdominal pain, illicit drug use, skin tattooing, blood transfusions, alcohol abuse, use of complimentary medications and alternative medications, recent travel, or high-risk sexual behaviour in the past. She didn't have any features of connective tissue disorders such as oral ulcers, skin rashes hair loss, joint pain, and skin rashes. She has dyslipidaemia for which she was on low dose Atorvastatin with regular follow up.

Physical examination is positive for deep icterus without lymphadenopathy, organomegaly, abdominal masses and features of Acute / chronic liver cell disease noted. Her BMI was within normal range.

Initial blood investigations revealed normal FBC with persistent markedly elevated liver enzymes (AST 1429 U/L, ALT 2458 U/L), conjugated hyperbilirubinemia (total bilirubin 384.8, Direct bilirubin 372.8, ALP 220 (46-116U/l) and altered coagulation profile (PT17.0 INR1.7, APTT 42.1). Abdominal ultrasound revealed hypoechoic liver without evidence of obstruction in biliary system or presence of gall stones. Viral markers like HBsAg, anti-HCV, IgM anti-HEV, IgM anti-HAV, anti-Cytomegalovirus IgM and anti-Epstein bar virus IgM were negative. Blood Glucose values,

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lipid profile Ferritin and ceruloplasmin levels were found to be within normal range. Serological markers such as rheumatoid factor, anti-smooth muscle antibodies (ASMA), and antimitochondrial antibodies (AMA) were negative. Antinuclear antibodies (ANA) were positive in a dilution of 1:1280. Hypergammaglobulinemia (IgG) level of 2437 mg/dl (569-1919 mg/dl) was noted with negative ANCA studies. Liver biopsy performed after 6 days of admission revealed mild to moderate interface hepatitis with mixed inflammatory cell infiltration including lymphocytes, eosinophils and occasional plasma cells with bile plugging supporting the clinical diagnosis of autoimmune hepatitis with cholestatic jaundice.

Prednisolone was initiated with the dose of 1mg/kg soon after the diagnosis. One week later her liver enzymes and bilirubin improved dramatically.

Discussion

Autoimmune hepatitis may present as either an acute or chronic disease with a fluctuating pattern (8, 9). Higher elevation of liver enzymes, milder elevation of ALP and IgG supported the diagnosis AIH in our patient. Though the typical finding of AIH is elevated liver enzymes than the elevation of bilirubin our patient's presenting symptom was deep jaundice with higher elevation of bilirubin, as her imaging study revealed no biliary tract obstruction it excluded the biliary tract destruction by inflammation it was additionally supported by prompt response to prednisolone in our case.

Mild elevation of bilirubin less than 3-fold of upper limit is common but it is very rare to have over 10 fold rise like in our patient. Liver histology typically showed the features of AIH in the patient.

The International Autoimmune Hepatitis Group established criteria for the diagnosis of the disease is shown in table 1.

Table 1 Simplified criteria for the diagnosis of AIH

Variable	Cutoff	points	Our patient
ANA/SMA	≥ 1:40	1	
ANA/SMA or anti-LKM or anti SLA	≥ 1:80 ≥ 1:40 Positive	2	2
IgG	≥ Upper limit of normal	1	
	≥ 1.10 times upper limit of normal	2	2
Liver histology	Compatible with AIH	2	
	Typical of AIH	1	1
Absence of hepatitis	yes	2	2

Autoantibody screening is frequently performed with automated techniques such as ELISAs and coated beads. These tests are reliable for SMA, anti-LKM1, anti-LC1, and anti-SLA antibodies, for which the target antigen is known, but not for ANAs, whose target in AIH has not been identified. The cutoff values for the automated tests vary between assays and laboratories. *A score ≥ 6 indicates probable AIH; a score ≥ 7 indicates definite AIH. †Additional points for all autoantibodies cannot exceed a maximum of 2.

According to this our patient's score was 7 indicating the diagnosis of AIH thus the rare entity of AIH with cholestatic jaundice in her case. Appropriate immunosuppressive therapy at the earliest based on the diagnosis can expedite a good response and it happened to be a practical clue for diagnosis as well.

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