

# Autoimmune Hepatitis: Varied Clinical Manifestations in a Tertiary Care Centre.

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## ABSTRACT

Autoimmune hepatitis is defined as chronic liver disease of unknown aetiology with aberrant auto-reactivity and genetic predisposition, characterized by female predominance, circulating auto-antibodies, hypergammaglobulinemia and association with HLA DR3 and HLA DR4. We present two patients with autoimmune hepatitis. The first patient is a young lady who was diagnosed with autoimmune chronic hepatitis. The second patient, on the other hand, is an elderly gentleman who presented to us with autoimmune hepatitis related decompensated cirrhosis of liver.

**Keywords:** Autoantibody, Autoimmune hepatitis, HLA DR3.

## INTRODUCTION

Autoimmune hepatitis is defined as chronic liver disease of unknown aetiology with aberrant auto-reactivity and genetic predisposition, characterized by female predominance, circulating auto-antibodies, hypergammaglobulinemia and association with HLA DR3 and HLA DR4.<sup>[1]</sup>

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## CASE REPORT

The heart specimen for this study are obtained from The first patient was a 20 year old house wife from an upper middle class family. She presented to medicine OPD with unexplained raised serum ALT, detected incidentally on routine investigation. Her investigations revealed serum ALT 74 U/L, serum AST 62 U/L, fasting blood sugar 4.6 mmol/L, triglyceride 170 mg/dl, HDL 39 mg/dl, TSH 1.44 µmol/L, and urinary copper <0.03 mg/L. There was no KF ring on slit lamp ophthalmic examination. She tested negative for HBs Ag, HBV DNA by PCR, anti-HCV, ASMA and AMA, but was positive for ANA at 1:120 dilution. Her endoscopy of upper GIT was normal and so was the abdominal ultrasound. CT scan of upper abdomen was also done and revealed inflammatory or focal fatty change in right lobe of liver. A liver

biopsy was done which revealed necro-inflammation (HAI-NI) score of 7 and fibrosis (HAI-F) score 1. The diagnosis of type 1 autoimmune hepatitis was done made.

The second patient, a 72 year old gentleman, coming from upper middle class socio-economic background presented to the GI clinic with ascites. He was a known diabetic for 20 years.

On examination, he had stigmata of cirrhosis of liver. His liver panel showed serum bilirubin 16 µmol/L, ALT 66 U/L, AST 114 U/L, alkaline phosphatase 291 U/L, albumin 27 gm/L, prothrombin time 12 sec. (control 12 sec.) and AFP 1.5 ng/ml. He tested negative for HBs Ag, anti-HBs, anti-HBc total, HBV DNA by PCR, anti-HCV and HCV RNA by PCR. ANA was positive at 1:640 dilution, but tests for AMA, anti-M2, anti-LKM 1, anti-SLA and anti-LP antibodies were all negative. Ascitic fluid was examined and SAAG was calculated at 2.1 gm/L. Endoscopy of upper GIT revealed grade II oesophageal varices.

The patient's serum creatinine was 82 µmol/L, serum sodium 133 mmol/L, potassium 3.2 mmol/L, chloride 99 mmol/L and bicarbonate 27 mmol/L. His IgG was 74 gm/L. Ultrasonography of abdomen revealed coarse ecotexture of liver with irregular, nodular surface. Percutaneous liver biopsy was done and histopathology revealed moderate inflammatory cellular reaction in portal tract with lymphocytes, histiocytes and plasma cells. There was moderate interface hepatitis and portal to central bridging. Liver parenchyma showed mild inflammation, aggregates of chronic inflammatory cells and severe portal fibrosis. We

diagnosed him as cirrhosis of liver due to sequel of type 1 autoimmune hepatitis.

## DISCUSSION

The entity of chronic autoimmune hepatitis was first described by Waldenstrom in 1950.<sup>[2]</sup> The disease has a female predominance with female: male ration being 8:1. It can be familial.<sup>[3]</sup>

Autoimmune hepatitis is of three types, namely type 1, 2 and 3. Most patients with autoimmune hepatitis have type 1 disease characterized by anti-DNA and anti-smooth muscle antibodies. In type 2, there is association with anti-liver kidney microsomal(LKM) type 1 antibody. It is divided into types 2a and 2b. In type 2a, liver kidney cytochrome mono-oxygenase P450 2D6 is the target antigen.<sup>[4]</sup> Patients often have associated extra-hepatic immune-mediated diseases like diabetes and show good response to corticosteroids. In type 2b on the other hand, anti-LKM-1 antibody is present in upto 70% cases. There is association with chronic hepatitis C virus infection and these patients respond to anti-viral better than to immuno-suppressive treatment. This type shows a male predominance. Type 3 autoimmune hepatitis shows association with anti-soluble liver antigen (SLA) and anti-liver and pancreas antigen (LPA) antibodies.<sup>[5]</sup>

Liver histology shows cellular infiltrates in zone 1 comprising mainly of lymphocytes and plasma cells. There is aggressive septa formation and steatosis in the liver. The condition rapidly progresses to macronodular cirrhosis, usually within two years.<sup>[6]</sup> During remissions, although the disease remains inactive, restoration of normal hepatic architecture does not occur.

The condition characteristically has two peaks, once in pre-pubertal period and then again between 4th to 6th decades. Onset is insidious with patients complaining of feeling unwell and jaundice. Chronic autoimmune hepatitis may remain quiescent for years. Amenorrhoea is common. On examination vascular spiders are seen almost in every case. Acne, hirsutism and cutaneous striae are present. Patients usually have hepatomegaly.

Extra-hepatic manifestations are common and include purpura, erythemas, arthralgia, lymphadenopathy, pulmonary infiltrates, pleurisy, rheumatic heart disease, ulcerative colitis, diabetes, thyroiditis, renal tubular defects and haemolytic anaemia.<sup>[6]</sup>

On investigation, there is hyperbilirubinemia, raised IgG, markedly elevated serum transaminases and pancytopenia. Liver biopsy remains an important tool for diagnosis.

Treatment is with oral corticosteroid *i.e.* prednisolone, which is initially given at a higher dose and then gradually tapered over a week to

maintenance dose. Treatment is at least for 2-3 years and at times for life with disease relapse following premature withdrawal. Azathioprine is added to prednisolone if there is no response. The drug is also used to substitute prednisolone if there are prednisolone induced complication.<sup>[7]</sup> Cyclosporine can be given to patients resistant to prednisolone.<sup>[8]</sup> Liver transplantation remains an option for patients with advanced liver disease, although disease recurrence post-transplant has been reported.<sup>[9]</sup> Mean survival with autoimmune hepatitis is 12.5 years and the 10 year survival is 63%.

## CONCLUSION

Autoimmune hepatitis is not infrequent and careful clinical observation is needed to identify cases. It is very important as the disease is potentially treatable, but otherwise it progresses rapidly to end stage liver disease.

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