# Active Autoimmune Hepatitis With Minimally Elevated Liver Enzymes: A Case Report

S Sarkar, P Dreyer, N Patel, J Salhab, J Sobrado

**Citation**

S Sarkar, P Dreyer, N Patel, J Salhab, J Sobrado. *Active Autoimmune Hepatitis With Minimally Elevated Liver Enzymes: A Case Report*. The Internet Journal of Gastroenterology. 2017 Volume 17 Number 1.

**DOI:** 10.5580/IJGE.52668

**Abstract**

Autoimmune hepatitis (AIH) is a chronic hepatitis of unknown etiology, which is characterized by the body's own immune cells attacking the liver cells. This results in inflammation, leading to scarring of the liver and ultimately cirrhosis. Diagnosis is made with a combination of laboratory and histological findings. Lab findings, include a striking elevation in amino transferase levels (as compared to bilirubin and alkaline phosphatase ALP), along with increased immunoglobulin G (IgG)/gamma globulin and the specific serological markers (ASMA/ANA/ALKM-1). We present a case of a 25-year-old male with a history of Type 1 AIH who presented with complaints of sharp abdominal pain in the right upper quadrant with distension, fever, chills, nausea, constipation and one episode of melena. The patient's blood work revealed only a modest elevation in serum transaminases and normal gamma globulin levels. He was diagnosed with spontaneous bacterial peritonitis (SBP) and showed evidence of cirrhosis, with a Model for End Stage Liver Disease (MELD) score of 11. A repeat liver biopsy at this time revealed pathological changes suggestive of AIH. This patient's case of autoimmune hepatitis is unique for several reasons, mainly because his liver enzymes were only minimally elevated at a time when his liver biopsy showed signs of active disease. One of the hallmark features of AIH are very high transaminases and elevated globulin, both of which this patient did not have. The case highlights the importance of diagnosing and treating AIH early in the course of the disease. This raises the question as to whether or not all cases of AIH are associated with a gross elevation of amino transferases. It emphasizes on the fact that chronic liver diseases can also present only with mildly elevated or even normal transaminases.

## BACKGROUND

Autoimmune hepatitis (AIH) is a chronic hepatitis, of unknown etiology, characterized by the body's own immune cells attacking the liver cells. This results in inflammation, leading to scarring of the liver and ultimately cirrhosis. It is generally characterized by circulating auto antibodies and high serum globulin concentration[1]. There are two types of autoimmune hepatitis. Type 1 is characterized by circulating auto antibodies including antinuclear antibodies (ANA) and/or anti smooth muscle antibody (ASMA). Type 2 is associated with antibodies to liver/kidneymicrosomes (ALKM-1) or liver cytosol antigen (ALC-1)[2]. The spectrum of presentation can vary from asymptomatic patients to those with signs of acute liver failure. Diagnosis is made with a combination of laboratory and histological findings[3]. Lab findings, include a striking elevation in aminotransferase levels (as compared to bilirubin and alkaline phosphatase ALP), along with increased immunoglobulin G (IgG)/gamma globulin and the specific serological markers (ASMA/ANA/ALKM-1). Histologically it is characterized by portal mononuclear infiltrate (lymphoplasmacytic) often with eosinophils, invading the hepatocyte boundary that surrounds the portal triad, percolating into the surrounding lobule, called as a piece meal necrosis[4]. It should be noted bile duct changes can be seen in up to 25% of cases. A high index of suspicion is required to identify the above findings and distinguish it from other causes of chronic hepatitis, so as to ensure a timely diagnosis and prevent progression to cirrhosis and need for transplant. We present the case of a 25-year-old with recurrent AIH but displaying no significant elevation of liver enzymes in the current flare up.

## CASE REPORT

A 25 year old Haitian incarcerated male with a known past medical history of Type 1 AIH(diagnosed in 2006), presented with complaints of sharp abdominal pain in the right upper quadrant. This was accompanied by abdominal distension, fever, chills, nausea, constipation and one

episode of melena. The patient was previously seen at our institution three years prior to the current presentation. During that previous hospitalization, blood work revealed Perinuclear anti-neutrophil cytoplasmic antibodies (P-ANCA), cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA), anti-mitochondrial antibodies (AMA) and antinuclear antibody (ANA) to be negative. ASMA was positive at 1:34. IgG level was normal at 135mg/dl, with aspartate transaminase (AST) at 247 U/L and alanine transaminase (ALT) at 253U/L. He was discharged back to the Department of Corrections but did not follow up until the current hospitalization. During this admission, blood work revealed only a modest elevation in serum transaminases –AST at 45 U/L, ALT at 60 U/L and ALP at 45 U/L. The patient now had evidence of cirrhosis, with a Model for End Stage Liver Disease (MELD) score at 11. He was diagnosed with spontaneous bacterial peritonitis (SBP) with a polymorphonuclear leukocyte (PMN) level of 975 cells/ mm3in ascitic fluid, and was then treated with intravenous ceftriaxone. The patient then underwent a screening esophagogastroduodenoscopy (EGD) which revealed non-bleeding esophageal varices. Colonoscopy did not reveal melena. The patient underwent repeat liver biopsy, which revealed portal areas having mixed inflammatory infiltrate, predominantly of lymphocytes, plasma cells and a few eosinophils, along with evidence of bile duct injury and atrophy, suggestive of activeAIH. The patient was discharged on a prednisone taper and azathioprine, with strong recommendations to follow up at a liver transplant center.

## DISCUSSION

This patient's case of autoimmune hepatitis is unique for several reasons, firstly because his liver enzymes were only minimally elevated, at a time when his liver biopsy showed signs of active disease. One of the hallmark features of AIH are very high transaminases and elevated globulin, both of which this patient did not have. It is possible, that this patient's liver parenchyma evolving to cirrhosis prevented the release of high levels of liver inflammatory markers. This raises the question as to whether or not all cases of AIH are associated with a gross elevation of aminotransferases. Some studies have shown cases of AIH with a cholestatic pattern, but that wasn't seen in our patient either[5]. Secondly, the IgG levels and serum globulin levels were normal which is uncharacteristic of the disease. Again, the patient's advanced cirrhotic liver may preclude a rise in the levels of globulins, which are mostly produced by the liver. Thirdly, the biopsy report revealed bile duct injury that is seen only in 25% of the cases, and can also be seen in overlap syndrome with primary biliary cirrhosis[6]. This case highlights the importance of diagnosing and treating AIH early in the course of the disease.It emphasizes on the fact that chronic liver diseases can also present only with mildly elevated or even normal transaminases [7]. More evidence is needed to evaluate the liver transaminases when the patients present with advanced cirrhosis.

## References

1. Edward, L. and M. Krawitt, Autoimmune hepatitis. N Engl J Med, 2006. 354(1): p. 54-66.
2. Johnson, P. and I.G. McFarlane, Meeting report: international autoimmune hepatitis group. Hepatology, 1993. 18(4): p. 998-1005.
3. Muratori, P., et al., Autoimmune hepatitis in Italy: the Bologna experience. Journal of hepatology, 2009. 50(6): p. 1210-1218.
4. Hofer, H., et al., Centrilobular necrosis in autoimmune hepatitis: a histological feature associated with acute clinical presentation. Journal of Clinical Pathology, 2006. 59(3): p. 246-249.
5. Washington, M.K., Autoimmune liver disease: overlap and outliers. Modern Pathology, 2007. 20(1s): p. S15.
6. Rust, C. and U. Beuers, Overlap syndromes among autoimmune liver diseases. World Journal of Gastroenterology: WJG, 2008. 14(21): p. 3368.
7. Brissos, J., et al., Autoimmune hepatitis: trust in transaminases. BMJ case reports, 2014. 2014: p. bcr2014203869.

## Author Information

**Sauradeep Sarkar, M.B.B.S**

**Patrick Dreyer, D.O.**
Larkin Community Hospital
South Miami, Florida, USA

**Nimal Patel, D.O.**
Larkin Community Hospital
South Miami, Florida, USA

**Joseph Salhab, D.O.**
Larkin Community Hospital
South Miami, Florida, USA

**Javier Sobrado, M.D.**
Larkin Community Hospital
South Miami, Florida, USA