

# Autoimmune Hepatitis Presenting without Autoantibodies: A Diagnostic Challenge

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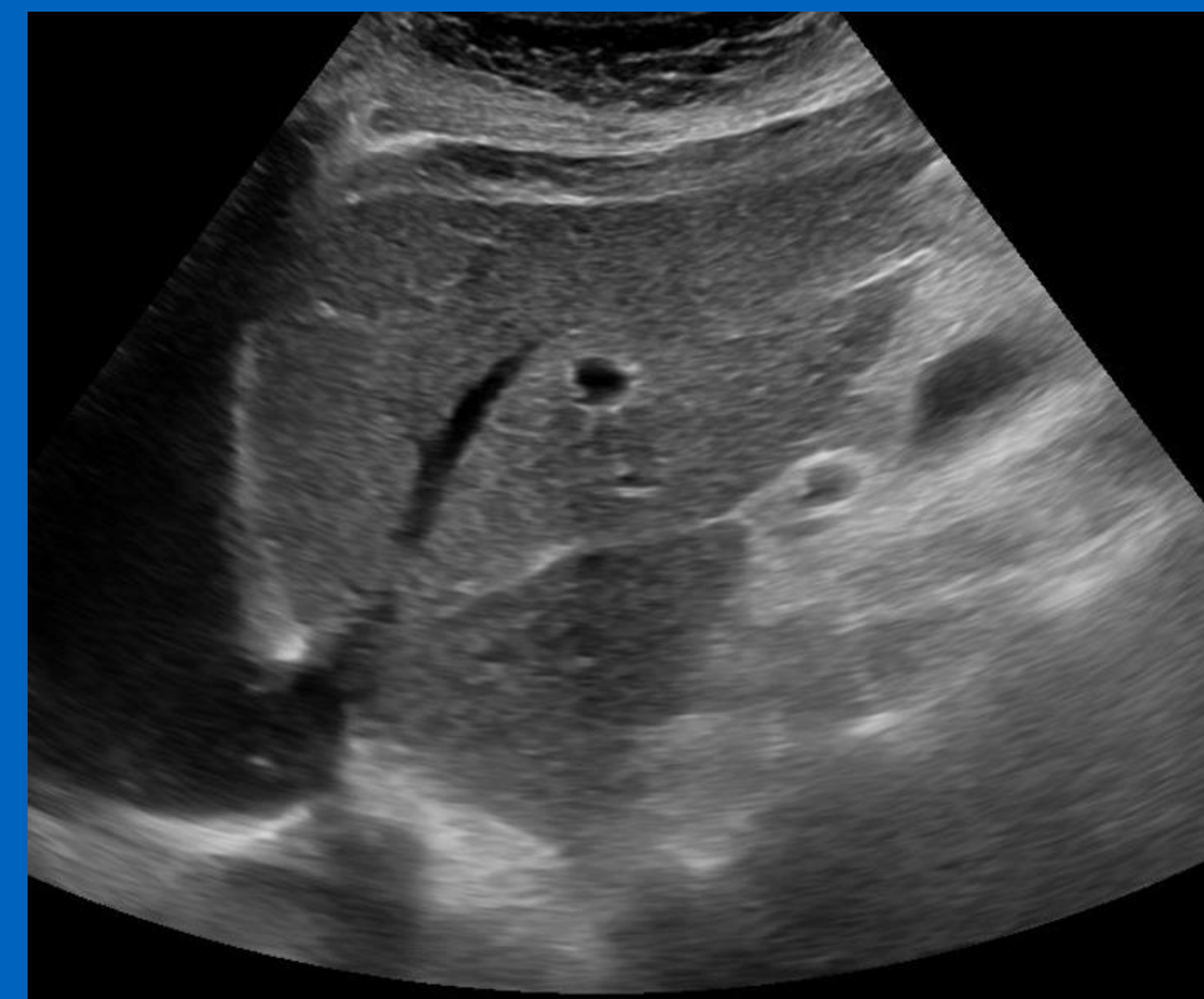


## INTRODUCTION

- Autoimmune hepatitis (AIH) refers to chronic inflammatory liver disease characterized by loss of tolerance to hepatocyte antigens.
- Approximately 70 to 80% of cases present with detectable autoantibodies, such as antinuclear antibody (ANA), anti-smooth muscle antibody (ASMA), and anti-liver-kidney microsomal-1 antibodies (anti-LKM-1).
- We report a unique case of seronegative AIH in a patient whose above mentioned antibodies were negative, but liver biopsy results were consistent with a diagnosis of autoimmune hepatitis.

## DISCUSSION

- Seronegative AIH follows a similar clinical course as seropositive AIH.
- When symptomatic, patients can present with abdominal pain, weight loss, and fatigue as well as laboratory workup with elevated liver enzymes.
- Atypical autoantibodies that may assist in the diagnosis of AIH when ANA, ASMA, and anti-LKM-1 are negative include atypical perinuclear anti-neutrophil cytoplasmic antibodies (p-ANCA) and anti-soluble liver antigen/liver pancreas antibody (anti-SLA/LP).
- *Clinicians with a high degree of suspicion for AIH should be aware that it can present without autoantibodies because when absent, appropriate diagnosis and treatment can be delayed.*



RUQ US: Liver normal in size and echotexture, smooth contour.



CT abdomen/pelvis: Normal liver size, no masses.

## CASE DESCRIPTION/METHODS

- A 54-year-old female with no significant past medical history presented with weeks of fever, night sweats, fatigue, and weight loss. She was vitally stable.
- Labs were significant for an aspartate aminotransferase (AST) of 362, alanine aminotransferase (ALT) of 448, alkaline phosphatase of 496, and normal total bilirubin.
- Imaging was significant for a normal right upper quadrant ultrasound and a computed tomography abdomen that was unremarkable for liver pathology.
- Further lab work-up included negative viral hepatitis serologies and negative ANA, ASMA, and anti-LKM-1.
- Due to concern for liver toxicity, patient's home tylenol and duloxetine were held without significant improvement in her transaminitis or symptoms.
- Liver biopsy was performed and showed marked lobular inflammation of hepatic architecture with predominantly intrasinusoidal pattern and portal tracts demonstrating interface hepatitis, consistent with a diagnosis of AIH.
- Patient was started on budesonide with resolution of her transaminitis and symptomatic improvement.