

A RARE CASE OF AUTOIMMUNE HEPATITIS-PRIMARY BILIARY CHOLANGITIS AND IGG4- RELATED DISEASE OVERLAP SYNDROME IN A MALE PATIENT

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Poster

Hepatobiliary

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A 56-year-old male with chronic alcohol use developed concurrent IgG4-related disease and autoimmune hepatitis-primary biliary cholangitis overlap syndrome, diagnosed over two years through serial biopsies and evolving laboratory findings.

KEY POINTS

- Initial presentation showed mixed liver function test derangement, pancreatic swelling on MRI, positive antinuclear antibodies (>1:640), anti-M2 antibody (103 u/mL), and normal IgG4 (53.9 mg/dL), with biopsies consistent with primary biliary cholangitis but not diagnostic of autoimmune hepatitis.
- Two years later, IgG4 levels rose dramatically to 7051 mg/dL, proptosis developed, and orbital nerve biopsy confirmed IgG4-related disease with thickened supraorbital nerves on imaging.
- Despite initial response to ursodeoxycholic acid and later steroid therapy, liver function tests remained abnormal; repeat liver biopsy showed progression of interface hepatitis and bile duct damage.
- Multidisciplinary review concluded histology was more consistent with autoimmune hepatitis-primary biliary cholangitis than IgG4-related disease, leading to final diagnosis of concurrent IgG4-related disease with autoimmune hepatitis-primary biliary cholangitis overlap syndrome, managed with azathioprine, fenofibrate, and ursodeoxycholic acid with subsequent improvement.
- This case demonstrates the diagnostic complexity of overlapping autoimmune conditions with multisystem involvement and evolving presentations.

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