

Immunoglobulin G4-related cholangitis

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This recently recognised entity presents with diffuse or segmental narrowing of the intra- or extrahepatic bile ducts. Its features may make differentiation from PSC, cholangiocarcinoma and pancreatic cancer difficult. However, patients often have elevated serum IgG4 levels and concomitant autoimmune pancreatitis, IgG4-related sialadenitis or retroperitoneal fibrosis. Biliary biopsies show lymphoplasmacytic sclerosing cholangitis. Treatment is with systemic steroids. Failure to respond to steroid therapy should make one reconsider the diagnosis and exclude an underlying malignancy .

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