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IgG4-related disease mimicking cholangiocarcinoma

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Immunoglobulin G4-related disease (IgG4-RD) is an immune mediated fibro inflammatory disease that consists of a collection of disorders that share particular pathologic, serologic, and clinical features. These disorders were previously thought to be unrelated. The most characteristic features include tumor like swelling of involved organs, a lymphoplasmacytic infiltrate enriched in IgG4 positive plasma cells and a variable degree of fibrosis that has a characteristic storiform pattern. In addition, elevated serum concentrations of IgG4 are found in 60 to 70 percent of patients with IgG4-RD. IgG4 related sclerosing cholangitis (IgG4-SC) is a characteristic type of sclerosing cholangitis, with an unknown pathogenic mechanism. Patients with IgG4-SC display increased serum IgG4 levels and dense infiltration of IgG4-positive plasma cells with extensive fibrosis in the bile duct wall. Circular and symmetrical thickening of the bile duct wall is observed in the areas without stenosis that appear to be normal on cholangiography, as well as in the stenotic areas. IgG4-SC has been recently recognized as an IgG4-related disease. IgG4-SC is frequently associated with autoimmune pancreatitis (AIP). IgG4-related dacryoadenitis/sialadenitis and IgG4-related retroperitoneal fibrosis are also occasionally present with IgG4-SC. However, some IgG4-SC cases do not involve other organs. IgG4-SC is most common in elderly men. Obstructive jaundice is frequently observed in IgG4-SC. A number of diseases such as cystic fibrosis, chronic obstructive choledocholithiasis, biliary structures (secondary to surgical trauma, chronic pancreatitis), anastomotic structures in liver graft, neoplasms (benign, malignant, metastatic), infections, hypertonic saline instillation in the bile ducts, posttraumatic sclerosing cholangitis, systemic vasculitis, amyloidosis, radiation injury, sarcoidosis, systemic mastocytosis, hypereosinophilic syndrome and Hodgkin's disease may easily be confused with IgG4-related sclerosing cholangitis, or coexist in a patient. In this case report a 57 years male patient presented with jaundice, fatigue, weight loss and oral moniliasis and right sided neck swelling. He was misdiagnosed as Cholangiocarcinoma.

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