

## Clinical Image

# Immunoglobulin IgG4 Systeem Disease with Colangitic Onset

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A 76 years old male was admitted to our unit for jaundice and progressive weight loss (20 kilos in 3 months). Blood tests showed increased values of bilirubin 1,2 mg/dl (conjugated 0,83mg/dl), Gamma-glutamyltransferase 767mg/dl alkaline phosphatase 509mg/dl and polyclonal hypergammaglobulinemia. Abdominal contrast-enhanced CT scan showed intrahepatic biliary dilation, diffuse pancreas enlargement with a halo of edema surrounding its body and tail, ectasia of Wirsung's collateral ducts, concentric thickening of the sub-renal aortic wall (Figure 1). Colangio-NMR showed segmentary thickening of common bile duct and I-II order biliary ducts causing stricture and upstream dilation (Figure 2). Suspecting IgG4-RD, serum IgG4 level was measured, resulting greatly increased (>1275 mg/dl). Based on typical imaging and lab-test findings IgG4-RD diagnosis was made [1]. The patient was treated with prednisone 30mg/day for 6 weeks, followed by 5mg/week tapering with rapid resolution of cholestasis and weight regain. Typical imaging and prompt response to steroid treatment may favour the diagnosis, even in the absence of biopsy.

## Reference

1. Kamisawa T, Zen Y, Pillai S, Stone JH. "IgG4-related disease". *Lancet*. 2015; 385: 1460-1467.



Figure 1:

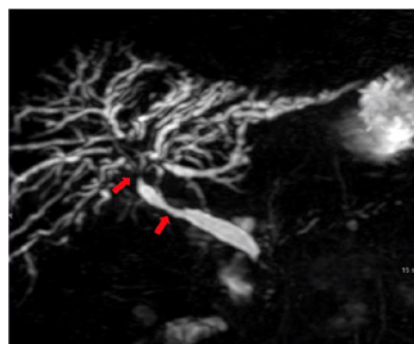


Figure 2: