

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.

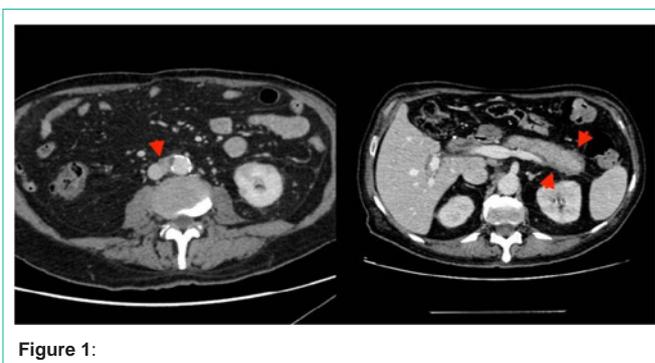


Figure 1:

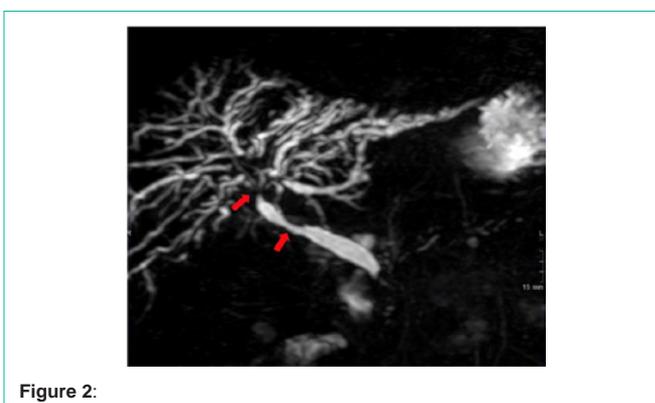


Figure 2: