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IMAGE OF THE MONTH

IgG4-Related Disease with Hypophysitis and Cholangitis as the Main Manifestations

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Case Presentation

A 53-year-old man presented with progressively worsening yellowish skin and sclera, upper abdominal discomfort, and fever for 1 year. Fatigue, anorexia, dry mouth, polydipsia, and difficulty swallowing were also obvious. Prior to presentation, he had been treated with transarterial chemoembolization (TACE) for suspected cholangiocarcinoma. After admission, laboratory examination showed total bilirubin (TBIL) 287.7 μmol/L, direct bilirubin (DBIL) 204.7 μmol/L, serum immunoglobulin G subtype 42.5 g/l (normal range 0.03–2.01 g/l), carbohydrate antigen 19-9 (CA19-9) 144.63 μ/mL, testosterone 30.56 ng/mL, prolactin 30.30 ng/mL, adrenocorticotropic hormone 116.98 pg/mL, and cortisol (8 am) 27.25 μg/mL. Tear film break-up time and Schirmer test of the patient were shorter than normal. Abdominal and pituitary magnetic resonance imaging suggested common bile duct disease, with enlarged pancreas and pituitary gland (Figs. 1, 2). Liver pathological biopsy demonstrated concentric collagenous deposition around interlobular bile ducts with numerous IgG4-positive plasma cells (Fig. 3). According to The 2019 American College of Rheumatology–European Union against Rheumatism Classification criteria for IgG4-RD of rheumatism and relevant diagnostic criteria [1, 2], the patient obtained the total score of 67 points [3] and was finally diagnosed with IgG4-related sclerosing cholangitis, hypophysitis, pancreatitis, sialadenitis, and lacrimal adenitis, belonging to the group of classic Mikulicz disease complicated by systemic organ involvement. After the standard regimen of methylprednisolone 40 mg/day, the symptoms and serum IgG4 were significantly improved. Currently, the patient is receiving methylprednisolone 4 mg/day maintenance, and regular follow-up.

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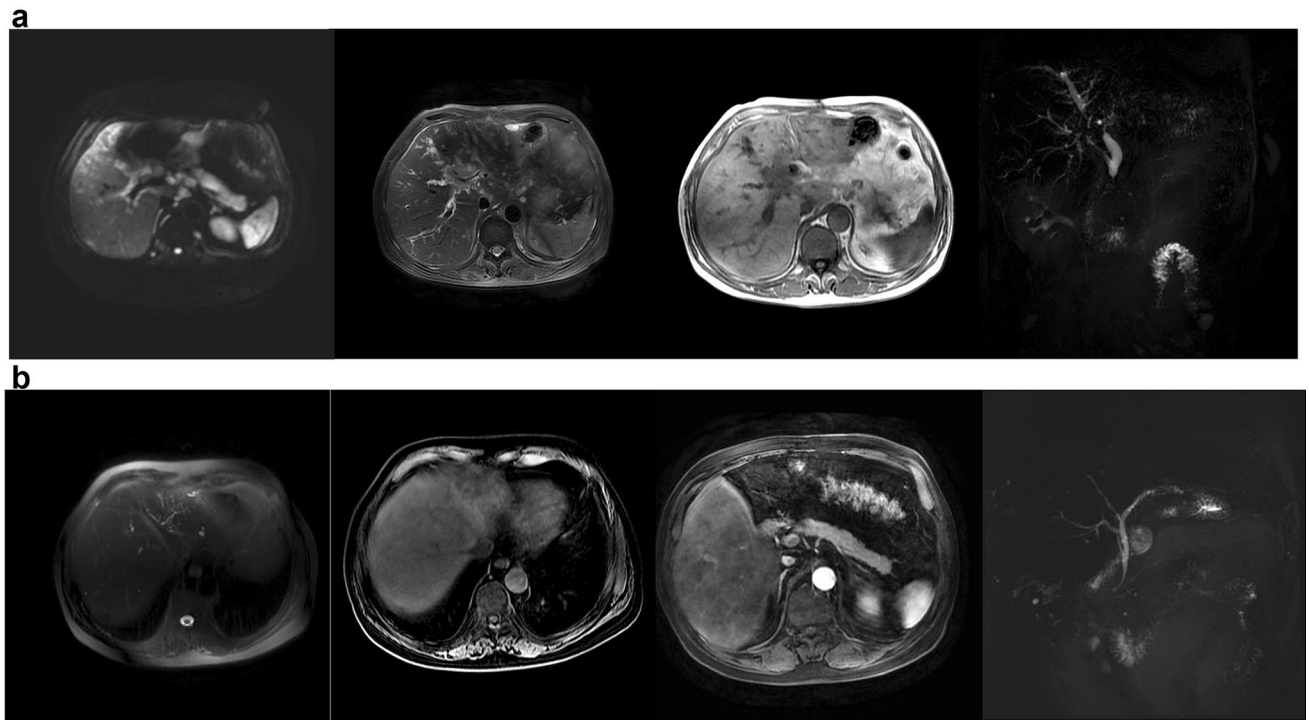

Fig. 1 Abdominal magnetic resonance imaging (MRI) + magnetic resonance cholangiopancreatography (MRCP). **a** Intrahepatic dilatation of bile duct with left hepatic pericholangitis, stenosis of lower common bile duct with dilatation of upper duct, and the head of the pancreas augment and lymph nodes around the head, in the abdominal cavity, and under the right diaphragm were enlarged. **b** One year after treatment, the volume of the left lobe of the liver was reduced, and the dilated left bile duct and the bile duct in the left lobe of liver were significantly improved. No other abnormality was found

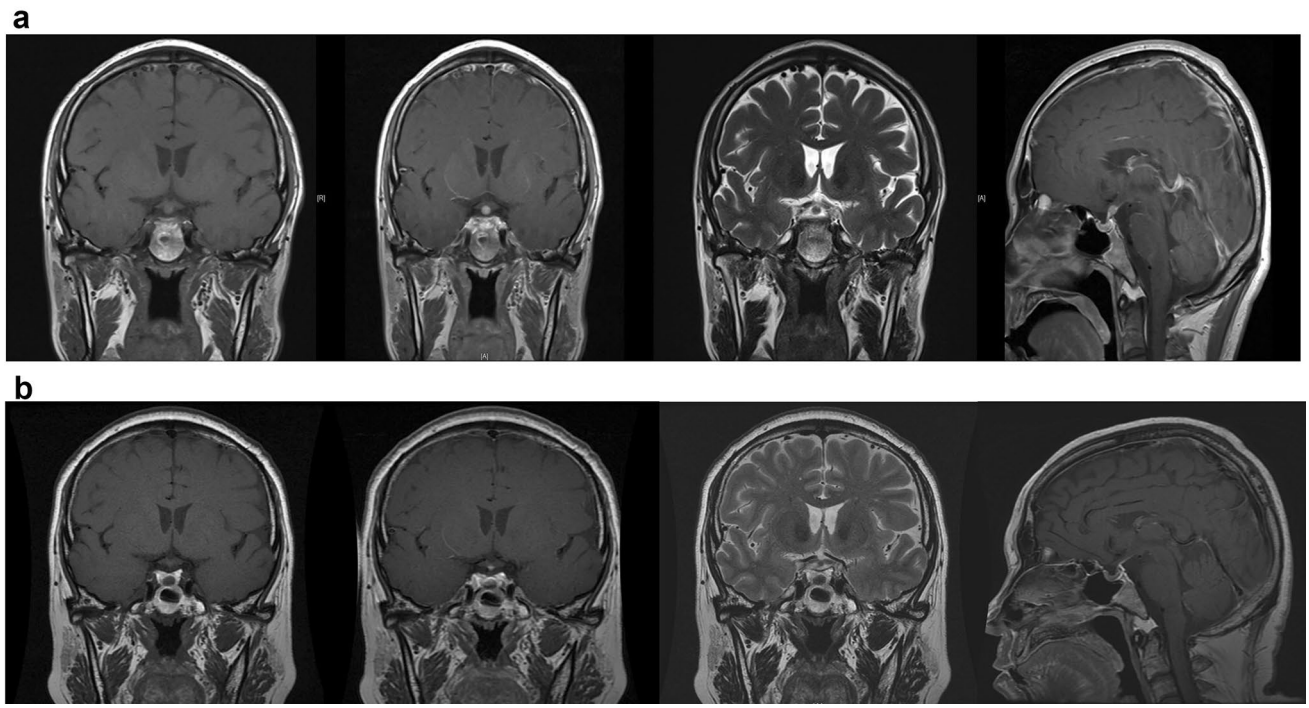

Fig. 2 Pituitary MRI. **a** The upper end of the pituitary stalk was thickened, the high signal in the posterior pituitary disappeared, and the enhancement degree of the pituitary decreased. **b** One year after treatment, the pituitary stalk was thinner than before, and the signal of other pituitary glands was similar

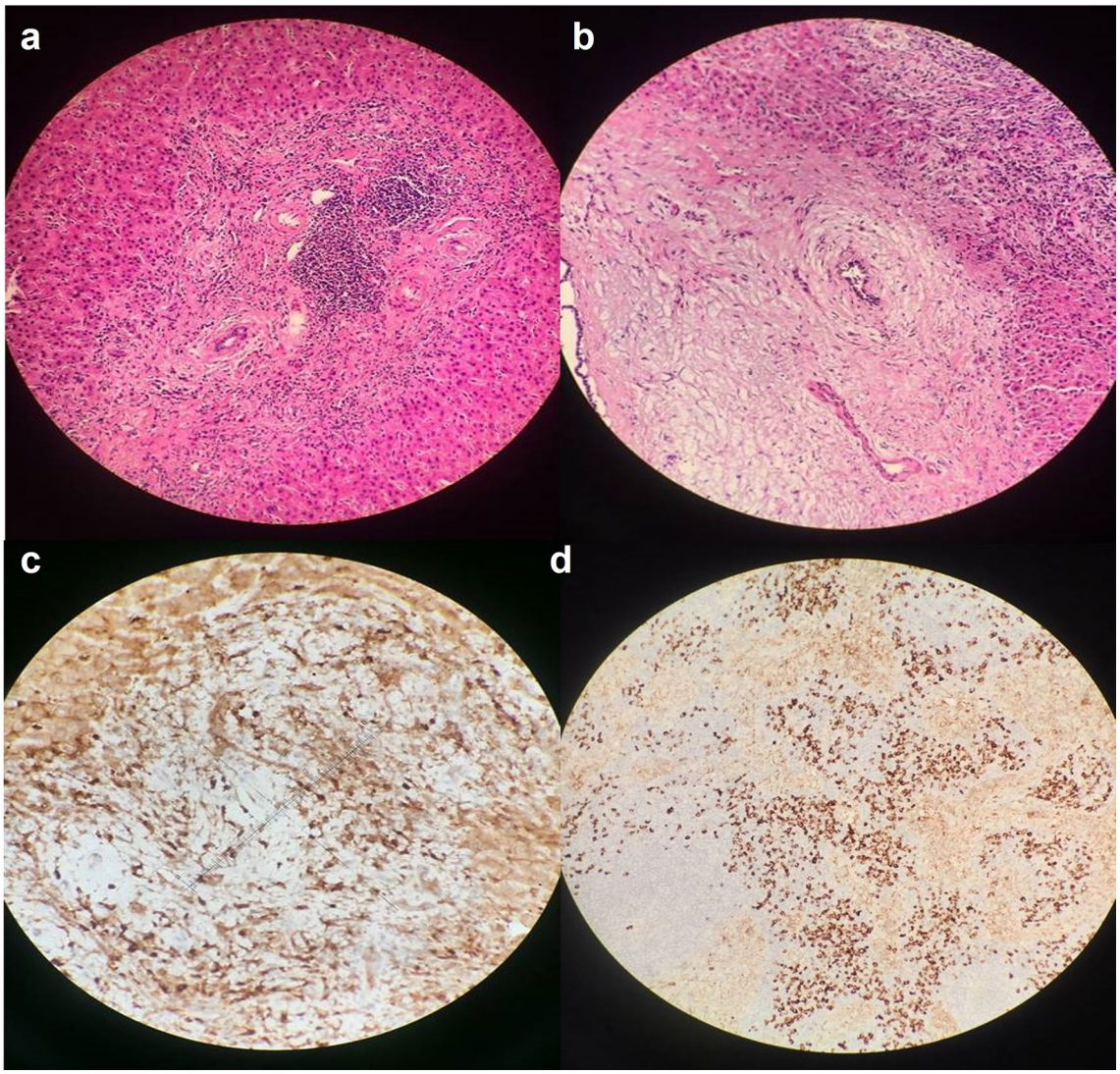

Fig. 3 Liver biopsy pathology. **a** The inflammation in the portal tract was more severe than that in the hepatic lobule, and abundant infiltration of plasma cells and bile duct injury were found. **b** There were concentric collagenous deposition around interlobular bile duct, and obvious interlobular hepatitis. **c** A large number of plasma cells and IgG4-positive plasma cells > 50 per high-power field; **d** Portal lymph nodes showed a large amount of plasma cell infiltration on MUM1 immunohistochemistry and IgG4-positive plasma cells > 50 per high-power field

Author's contribution Yan Huang and Yu Meng Liu contributed equally to this manuscript. H.Y. designed the work and revised it critically for important intellectual content. L.Y.M. acquired and analyzed data for the work and drafted the manuscript. L.Z.H. acquired and analyzed pathological data. G.J.H. substantively revised the manuscript. Z.W.F. contributed ideas to the work. H.X.P. critically revised the manuscript.

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Declarations

Conflict of interest The authors declare that they have no competing interests and approved the final manuscript.

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