

IgG4-related cholangitis mimicking Klatskin's tumor in patient with normal serum IgG4 level

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A 66-year-old female patient who was diagnosed with IgG4-related sialoadenitis of right submandibular gland two years ago visited outpatient clinic with painless jaundice. The patient was taking the prednisolone 5mg daily for maintenance therapy after remission of sialoadenitis. The patient had no complaints other than jaundice, and vital sign were stable. Blood tests performed after admission showed cholestatic pattern of liver dysfunction. Considering the patient's history, the serum IgG4 level was measured, but it was within the normal range (3.9-86.4mg/dL) at 52mg/dL. A CT scan was performed to identify the cause of jaundice. On the CT scan, a 2.5cm segmental stricture of the common hepatic duct and dilation of both intrahepatic bile ducts were observed (Fig 1A). Because the patient was in a state of complete remission of IgG4-related sialoadenitis in the past with normal serum IgG4 level and had been taking steroids, it was necessary to make a differential diagnosis for Klatskin's tumor. Thus, endoscopic retrograde cholangiopancreatography (ERCP) was planned for biopsy and decompression of the bile duct. On the next day, ERCP was performed, and stricture of common hepatic duct was shown on cholangiogram (Fig. 1B). After biopsy at the stricture site, two plastic stents were placed in the both intrahepatic bile ducts (Fig. 1C). Subsequently, the patient's symptom and liver function began to improve, and final pathologic report showed IgG4-related cholangitis with more than ten IgG4 positive lympho-plasma cells infiltrate of the bile duct on high power field in microscopic exam. High dose steroid therapy (Prednisolone 30mg/day) was started again for complete remission and ERCP for follow-up examination was planned to check the lesion. On the cholangiogram performed three months later, the stricture of the bile duct was completely resolved (Fig. 1D). Discussion IgG4 related disease can involve systemic organs, and especially when it occurs in the bile duct as in this case, it can be confused with malignancy. Therefore, thoroughly checking the patient's medical history and performing various tests including biopsy are very important.

