

IgG4-related disease involving the gallbladder and bile duct mimicking a Klatskin tumor

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A 76-year-old man was admitted to our hospital complaining of abdominal pain with fever. Physical examination showed scleral icterus and right upper quadrant tenderness. Laboratory findings revealed leukocytosis, abnormal liver function tests, conjugated hyperbilirubinemia, and elevated carbohydrate antigen 19-9. Abdominal computed tomography demonstrated irregular wall thickening of the gallbladder with multiple stones surrounding a large low-density lesion and an irregular, thickened, enhancing wall at the hilum and common hepatic duct (Figure 1-A, B), Magnetic resonance cholangiopancreatography showed a stricture at the hilum extending into the CHD, highly representing a Klatskin tumor (Figure 1-C). We performed percutaneous catheter drainage for gallbladder empyema and endoscopic retrograde cholangiopancreatography for biliary drainage with intraductal biopsy. No evidence of malignant cells was noted, however, so we performed a peroral cholangioscopy using the SpyGlass™ direct visualization system (Figure 2-A). SpyDS demonstrated a stricture with a hyperemic, smooth, and edematous mucosal surface in the hilum (Figure 2-B). Specimens obtained by the Spybite forceps showed a few inflammatory cells with > 10 IgG4-positive plasma cells (Figure 2-C). A cholecystectomy was performed, and the histology showed xanthogranulomatous inflammation and necrosis with more than 10 IgG4 plasma cells. Therefore, we diagnosed IgG4-related disease (IgG4-RD) involving the gallbladder and bile duct, and we initiated steroid therapy. One month later, MRCP demonstrated decreased extent of luminal irregularity and enhancing wall-thickening of the proximal CHD (Figure 3-A, B). The patient takes a low-dose steroid and continues with follow-up visits. IgG4-RD involving the gallbladder and bile duct is rare, and is difficult to differentiate from malignancy. In the present case, the results of the SpyDS-guided biopsy and visualization played an important role in the definitive diagnosis. Even when biliary cancer is strongly suspected by radiologic studies, IgG4-RD may be considered as a differential diagnosis.

