A case of IgG4-related disease complicated by duodenal bulbitis with IgG4-positive plasma cell infiltration

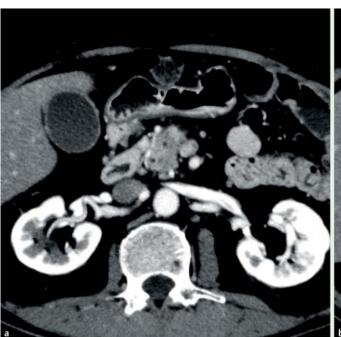




Fig. 1 Computed tomographic findings in the pancreas: a diffuse pancreatic enlargement and a peripancreatic capsule, b tumor with lower contrast enhancement in the pancreatic head.



Fig. 2 Magnetic resonance imaging scan showing diffuse pancreatic ductal stenosis.

Duodenal papillitis is a gastrointestinal lesion commonly complicating IgG4-related diseases. Lesions of the esophagus, stomach, and colon with IgG4-positive plasma cell infiltration also complicate

IgG4-related diseases. However, few reports have been published on complicating duodenal bulbitis. We encountered a patient with an IgG4-related disease complicated by duodenal bulbitis.

A 72-year-old man attended our hospital with a chief complaint of urine discoloration. A blood test showed increased hepatobiliary enzymes (aspartate aminotransferase 401 U/L, alanine aminotransferase 1148 U/L, total bilirubin 1.4 mg/dL, direct bilirubin 0.8 mg/dL, alkaline phosphatase 2527 U/L, γ-glutyl transpeptidase 1792 U/L). All tumor markers were slightly raised. Contrast computed tomography showed a tumor with lower contrast enhancement in the pancreatic head (Fig. 1b). Bile duct stenosis was also noted in the pancreatic head. Biliary duct dilatation was noted on the hepatic side. No pancreatic duct dilatation was observed. Diffuse pancreatic enlargement and a peripancreatic capsule were identified (Fig. 1 a). Magnetic resonance imaging showed diffuse pancreatic ductal stenosis (> Fig.2). An additional blood test showed a high IgG4 level (608 mg/dL) and the presence of antinuclear antibodies (1: 40, homogeneous). Cytological and histological biopsies taken from the common bile duct during endoscopic retrograde cholangiopancreatography showed neither malignancy nor pathological findings

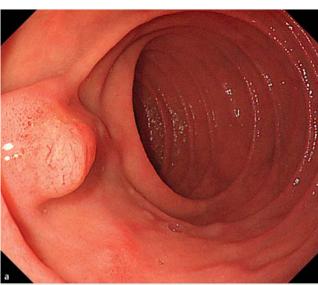


Fig. 3 Endoscopic views showing diffuse redness and edematous lesions: a duodenal papilla, b duodenal bulb.



characteristic of IgG4-related diseases. Upper gastrointestinal endoscopy showed diffuse redness and edematous lesions in the duodenal papilla and bulb (Fig. 3). Histological biopsies from these sites showed IgG4-positive plasma cell infiltration (Fig. 4). The patient was diagnosed with an IgG4-related disease. Steroid therapy was started, improving the hematological findings, pancreatic enlargement, and bile duct stricture and markedly alleviating the duodenal papillitis and bulbitis (Fig. 5).

Endoscopy showed no continuity in the images of the duodenal papillitis and bulbitis. The redness of the duodenal mucous membrane was macular with the mucous membrane pattern retained. Pathological analysis showed erythrocytes in the lamina propria, but showed neither longitudinal ulcer nor luminal contraction, common lesions due to extrinsic inflammation. Thus, the duodenal bulb lesion consisted mainly of mucous membrane inflammation, and may have been a lesion of IgG4-related disease, which developed independently of pancreatitis.

Endoscopy_UCTN_Code_CCL_1AF_2AF_3AZ

Competing interests: None

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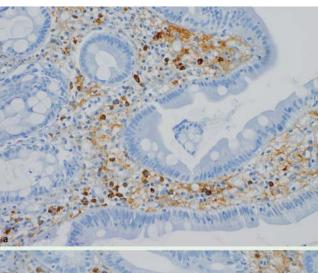


Fig. 4 Histopathologic sections showing IgG4positive plasma cell infiltration: **a** duodenal papilla, **b** duodenal bulb.

Bibliography

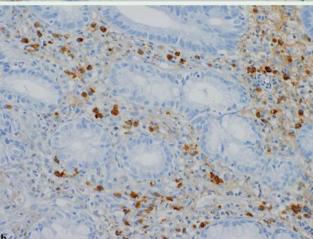
DOI http://dx.doi.org/ 10.1055/s-0034-1377397 Endoscopy 2014; 46: E408–E410 © Georg Thieme Verlag KG Stuttgart · New York ISSN 0013-726X

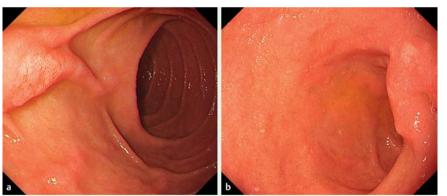
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Endoscopic views showing alleviation of inflammation: **a** duodenal papilla, **b** duodenal bulb.