Diagnostic and Interventional Endoscopy

Clinical Image

IgG4-Related Inflammatory Pseudotumor of the Bile Duct Mimicking Cholangiocarcinoma

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IgG4-related disease (IgG4-RD) is a chronic fibroinflammatory condition that can affect various organs, including the pancreas, biliary tree, salivary glands, kidneys, and lymph nodes.[1,2] Histopathologically, it is characterized by dense infiltration of IgG4-positive plasma cells, storiform fibrosis, and obliterative phlebitis.[2] Inflammatory pseudotumors (IPTs) associated with IgG4-RD are rare, most often reported in the lungs, peripancreatic area, and retroperitoneum. Development of IPTs in the bile ducts is exceedingly rare; however, when present, they may clinically and radiologically mimic cholangiocarcinoma, posing significant diagnostic challenges.[1]

A 66-year-old male initially presented to the emergency department with worsening abdominal pain and was diagnosed with acute pancreatitis. Imaging revealed a 6 mm pancreatic duct and a markedly dilated common bile duct (CBD) measuring 21 mm. Endoscopic retrograde cholangiopancreatography (ERCP) showed irregular narrowing of the distal 3 cm of the CBD. Biopsies from the papilla revealed IgG4-related papillitis, and corticosteroid therapy was initiated. Over the years, the patient underwent multiple ERCP procedures for recurrent sludge and stones in the CBD.

Twelve years later, the patient again presented with abdominal pain. Computed tomography imaging demonstrated a soft-tissue density lesion within the CBD. Cholangiography revealed a 20 mm polypoid lesion fixed to the wall of the common hepatic duct (Figure 1). The mass was removed endoscopically using a basket and sent for histopathological examination (Figure 2). The lesion demonstrated diffuse IgG and IgG4 positivity and was diagnosed as an IgG4-related polypoid lesion consistent with a sclerosing inflammatory pseudotumor.The patient provided informed consent for the procedure.

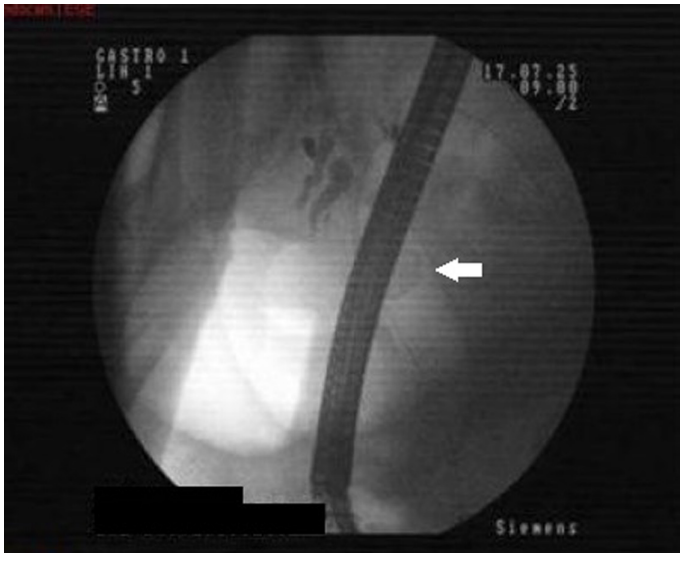

Figure 1. Cholangiographic image obtained during ERCP demonstrating a polypoid lesion (white arrow) adherent to the wall of the common hepatic duct. The lesion caused partial obstruction and radiologically mimicked a malignant stricture.

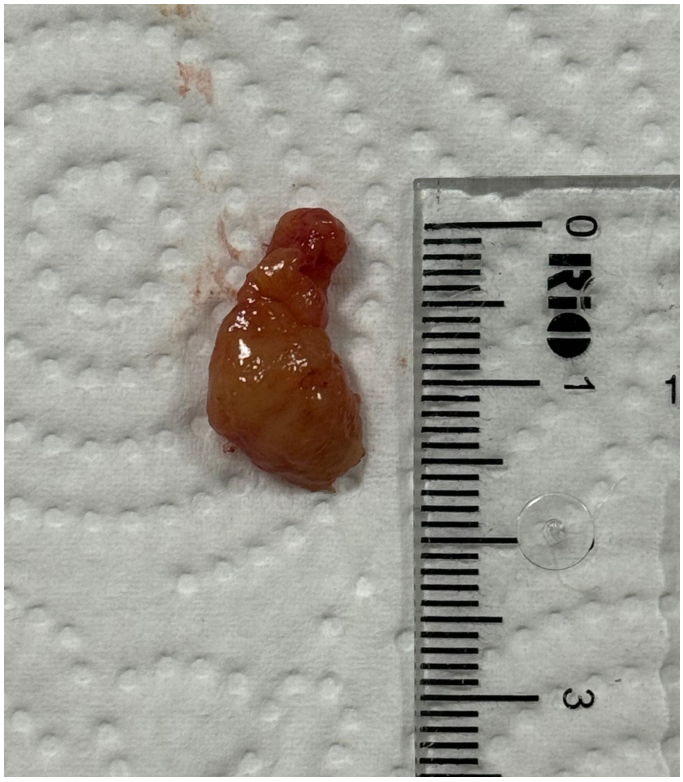

Figure 2. Gross view of the excised polypoid lesion following endoscopic retrieval. The specimen measured approximately 15 mm in length and displayed a lobulated and smooth surface.

Data Availability Statement: The data that support the findings of this study are available on request from the corresponding author.

Informed Consent: Written informed consent was obtained from the patient who agreed to take part in the study.

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