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Obstructive Jaundice Is Not Always Surgical!

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CASE REPORT

A 60-year-old diabetic man presented with a 1-month history of obstructive jaundice, weight loss of 16 kg, and fever and chills for more than 7 days. On examination, the patient was found to have deep icterus, tenderness in the right hypochondrium, and a palpable gall bladder. His laboratory parameters were hemoglobin 10.4 g/dL, total white blood cells 18.3 $\times$ $10^3/\mu L$, serum total bilirubin 25.4 mg/dL, direct bilirubin 21.0 mg/dL, aspartate aminotransferase 66 U/L, alanine aminotransferase 73 U/L, and alkaline phosphatase 407 U/L. The serological markers for hepatitis B, hepatitis C, and human immunodeficiency virus were negative.

An abdominal ultrasound revealed dilated biliary radicles and proximal common bile duct (CBD). A contrast enhanced computed tomography of abdomen and magnetic resonance cholangiopancreatography revealed a short-segment distal CBD stricture with proximal biliary dilation (Figure 1). An endoscopic ultrasonography showed a hypoechoic lesion involving the ampulla and the distal CBD (Figure 2). CA 19-9 was elevated to 1,026 U/mL, and the serum IgG4 (immunoglobulin G, subclass 4) level was measured to be 236 mg/dL. Under the cover of intravenous ceftriaxone for cholangitis, endoscopic retrograde cholangiopancreatography was

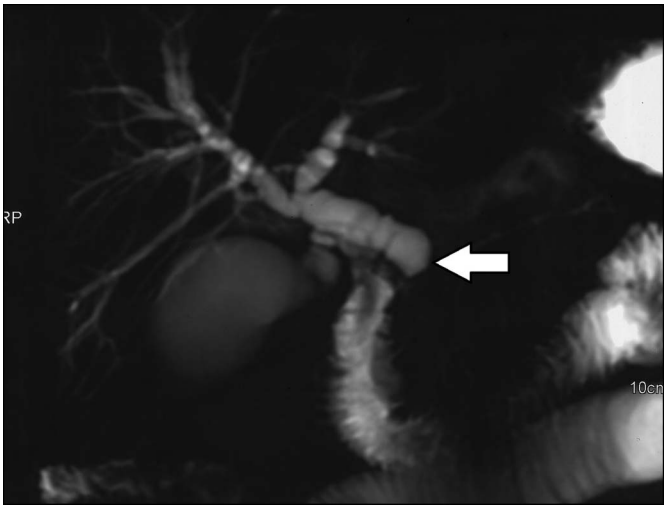

Figure 1. Magnetic resonance cholangiopancreatography revealing short segment distal common bile duct stricture (arrow).

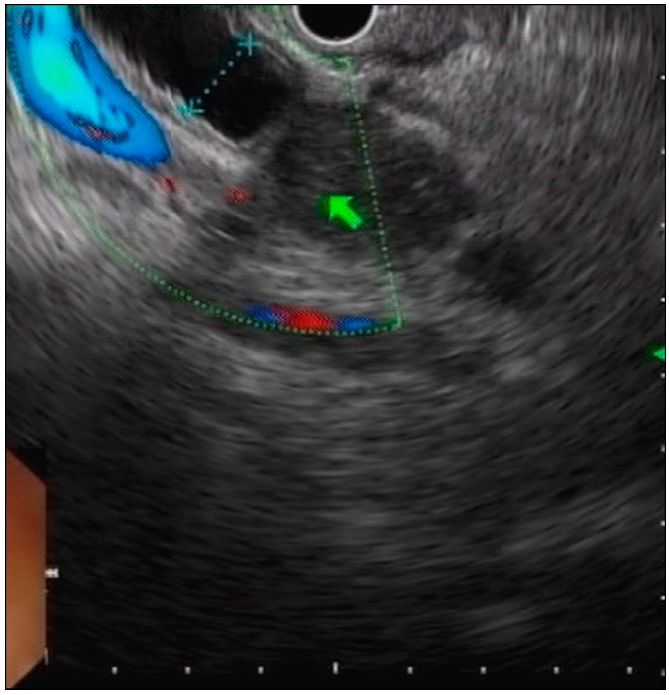

Figure 2. Endoscopic ultrasound showing a hypoechoic lesion involving the distal common bile duct (arrow).

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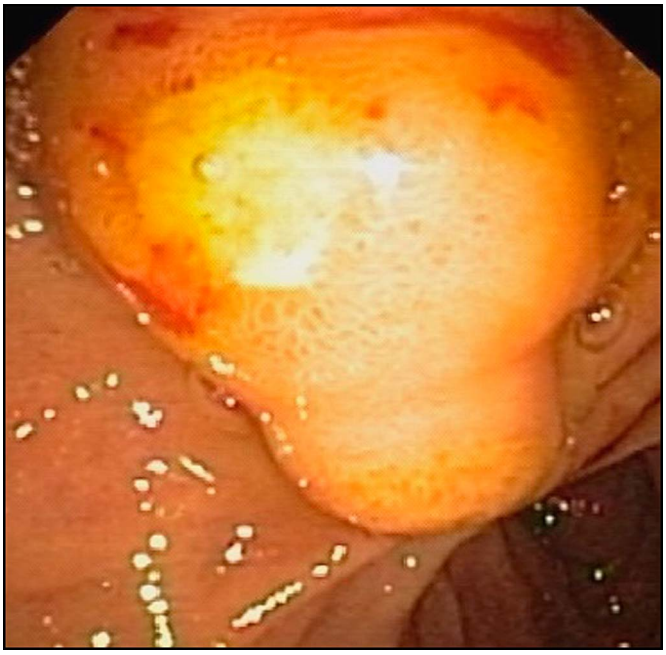

Figure 3. Endoscopic retrograde cholangiopancreatography showing bulky and prominent papilla with surface ulceration.

performed. The papilla appeared bulky and prominent with surface ulceration. After precut biliary access and sphincterotomy, a plastic stent (10 Fr × 10 cm, Cotton-Leung; Cook Medical, Bloomington, IN) was deployed (Figure 3). Biopsies were taken from the cut surface of the ampulla.

The histopathologic examination revealed storiform fibrosis around vessels and glandular structures surrounded by lymphocytes, plasma cells, and eosinophils. Most of the plasma cells were positive for IgG4 (50%) on immunohistochemistry, consistent with IgG4-related diseases (Figure 4). He was treated with oral prednisolone at 40 mg/day, tapered over 12 weeks. His serum bilirubin, transaminases, and CA19-9 gradually normalized after treatment. After biliary stent retrieval, a repeat magnetic resonance cholangiopancreatography imaging revealed a resolution of the distal CBD stricture (Figure 5).

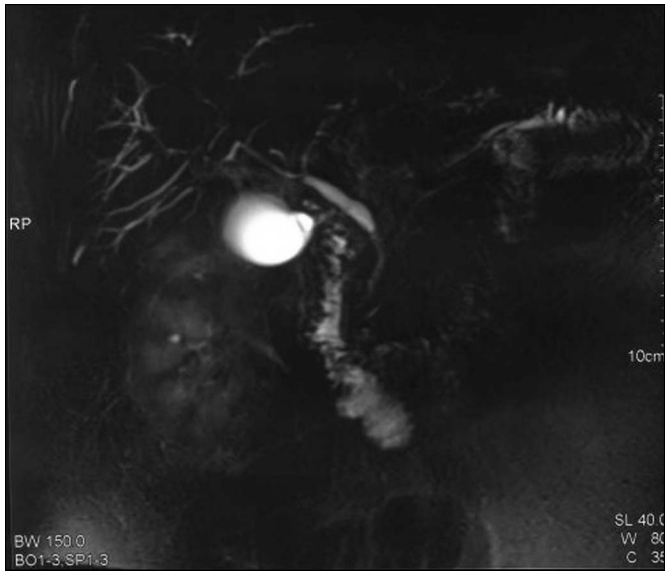

Figure 5. Repeat magnetic resonance cholangiopancreatography imaging showing a resolution of the distal common bile duct stricture.

The IgG4-related sclerosing cholangitis (SC) can involve any part of the biliary tree, commonly presenting as SC or a pseudotumorous hilar lesion.[1] It is associated with autoimmune pancreatitis in nearly 80%–90%.[2] Hence, IgG4-SC, presenting as an ampullary pseudotumor with biliary stricture without autoimmune pancreatitis, as in the present case, is exceptional. Because IgG4-SC closely mimics cholangiocarcinoma, many such cases have been treated surgically.[3] IgG4-SC responds dramatically to steroid therapy. Hence, IgG4-SC should be strongly considered among the differential diagnoses of indeterminate biliary obstruction to avoid an unwarranted surgery.

DISCLOSURES

Author contributions: All authors contributed equally to the manuscript creation. C. Thoguluva Seshadri is the article guarantor.

Financial disclosure: None to report.

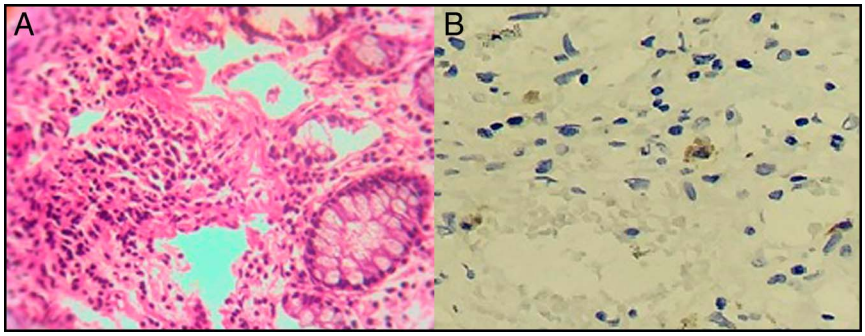

Figure 4. (A) Hematoxylin & eosin stain of ampulla biopsy showing storiform fibrosis around vessels and glandular structures surrounded by lymphocytes, plasma cells, and eosinophils (400×). (B) Most of the plasma cells were positive for IgG4 (50%) on immunohistochemistry (400×).

Informed consent was obtained for this case report.

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