B-Cell Lymphoma Involving the Biliary Tree

Primary or secondary non-Hodgkin's lymphomas of the extrahepatic bile ducts are rare findings. The diagnosis can be established by means of exfoliative bile cytology, brush cytology, and biopsy, including immunohistochemistry and immunocytochemistry via endoscopic retrograde cholangiography, or percutaneous transhepatic cholangiography.

In an 84-year-old patient with jaundice, known to have a stage-III B-cell chronic lymphatic leukemia (B-CLL), sonography and CT confirmed lymph node enlargement in the porta hepatis, hepatosplenomegaly, and dilated intrahepatic bile ducts. Endoscopic

a, b

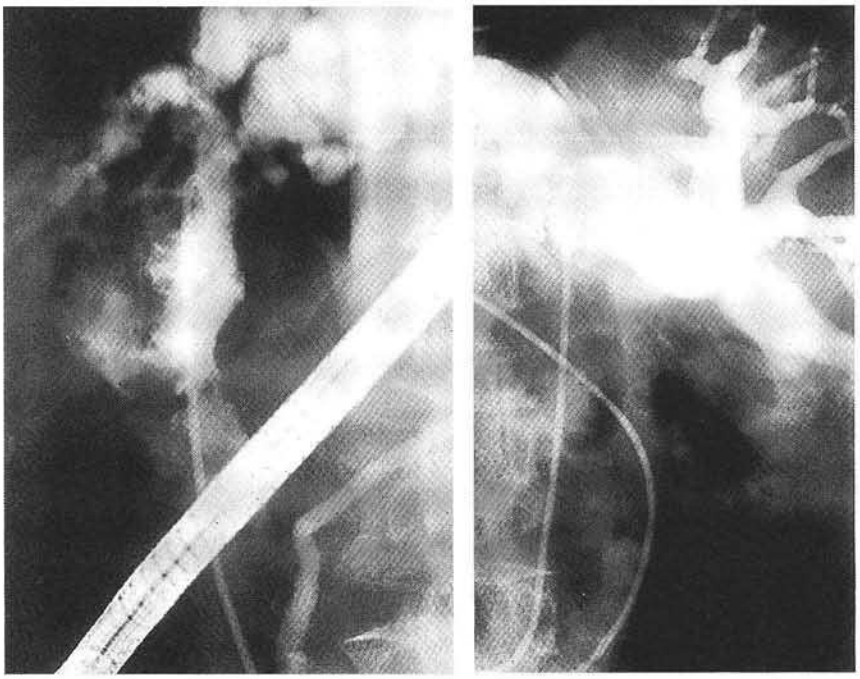

Figure 1 a: Common bile duct with narrowed and dilated segments. **b** Dilated intrahepatic bile ducts, visible after insertion of a nasobiliary tube.

retrograde cholangiopancreatography (ERCP) revealed massive dilation of the intrahepatic bile ducts and the biliary duct showed narrowed and dilated segments (Figure **1**). After endoscopic papillotomy (EPT), cytological and biopsy tests were performed. Brush cytology and exfoliative bile cytology resembled a peripheral blood smear in B-CLL. The histology and immunohistology confirmed a B-CLL involving the common bile duct. The small lymphocytes and lymphoplasmacellular elements expressed CD-19, 20, 22, 24 and CD-5. There was no evidence of cytomegalovirus or mycobacterial infection. Serological tests for cytomegalovirus, anti-neutrophile cytoplasmatic antibodies (ANCA), and human immunodeficiency virus as well as the tuberculin skin reaction, were negative.

The sensitivity of brush cytology via ERCP in the diagnosis of biliary and pancreatic carcinoma is 56.2%; the specificity is 100% (1). In the diagnosis of malignant bile duct strictures, the sensitivity of brush cytology was 59% and that of exfoliative bile cytology was 24% (2).

Non-Hodgkin's lymphoma of the bile ducts is rare. Broulard et al. (3) described a primary high-grade malignant T-cell-rich B-CLL of the common bile duct in a 34-year-old woman; the diagnosis was made by laparotomy. Wegerle et al (4) reported a 28-year-old man with angioimmunoblastic lymphadenopathy or T-zone lymphoma of the bile ducts, initially diagnosed as primary sclerosing cholangitis. Moody reported three patients developing Non-Hodgkin's lymphoma in the porta hepatis after orthotopic liver transplantation (5).

Jaundice in malignant lymphoma can be caused by liver involvement, bile duct obstruction by lymph nodes or malignant bile duct involvement. Another rare cause of jaundice is the "vanishing bile duct syndrome" in Hodgkin's disease, where the diagnosis is confirmed by liver biopsy. Viral, bacterial, and parasitic infections, as well as secondary malignancy, should be considered in an immunocompromised host. Secondary autoimmune diseases of the liver, and toxic effects of chemotherapy and radiotherapy in Non-Hodgin's lymphoma can cause intrahepatic or extrahepatic jaundice.

The diagnosis of bile duct involvement by a Non-Hodgkin's lymphoma can be established by combining brush cytology, exfoliative bile cytology, and biopsy from the bile ducts via endoscopic retrograde cholangiography or percutaneous transhepatic cholangiography. In leukemic non-Hodgkin's lymphoma, biopsy may be the best diagnostic approach, as cytology specimens can be contaminated with blood. Most patients present with secondary bile duct involvement in advanced-stage non-Hodgkin's lymphoma. Although bile duct locations are very rare, they should be included in the differential diagnosis of obstructive jaundice in non-Hodgkin's lymphoma patients.

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