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Anaplastic Large Cell Lymphoma and Vanishing Bile Duct Syndrome

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Case Report

A 39-year-old healthy man noticed a painful left chest wall mass associated with daily fevers, generalized pruritus, and jaundice. A hepatic panel showed a total bilirubin of 24.9 mg/dL, AST of 224 U/L, ALT of 506 U/L, and alkaline phosphatase of 1947 U/L. A chest computerized tomography (CT) showed a 9.6 x 5.2 cm chest wall mass extending to the mediastinum, and biopsy revealed anaplastic large cell lymphoma (ALCL). An abdominal CT showed normal liver parenchyma without ductal obstruction. Liver biopsy revealed bile duct damage, cholestasis, and hepatocyte injury (Figure 1).

He underwent chemotherapy with gemcitabine, dexamethasone, and platinum. However, his jaundice and pruritus worsened, and a repeat liver biopsy demonstrated the complete loss of intrahepatic bile ducts, compatible with vanishing bile duct syndrome (VBDS; Figure 2). Eight weeks after chemotherapy, he is free of malignancy with complete resolution of the chest wall mass. He is fully active but remains jaundiced with a total bilirubin of 29.7 mg/dL.

VBDS is an acquired disorder resulting in progressive destruction and disappearance of the intrahepatic bile ducts. It can be the manifestation of congenital disease, neoplastic disorders, or drug-induced liver injury[1,2] VBDS is a rare cause of jaundice in patients with Hodgkin's and non-Hodgkin's diseases such as ALCL, a sub-type of lymphoma primarily involving CD30-positive T cells.[3,4] The overall prognosis remains poor, and treatment is dependent on the underlying etiology, with the emphasis on supportive care. The loss of bile ducts is often an irreversible process; however, the resolution following successful chemotherapy in lymphoma patients has been reported.[5] Liver transplantation should be considered for those with decompensation if the underlying disease is under control.[1]

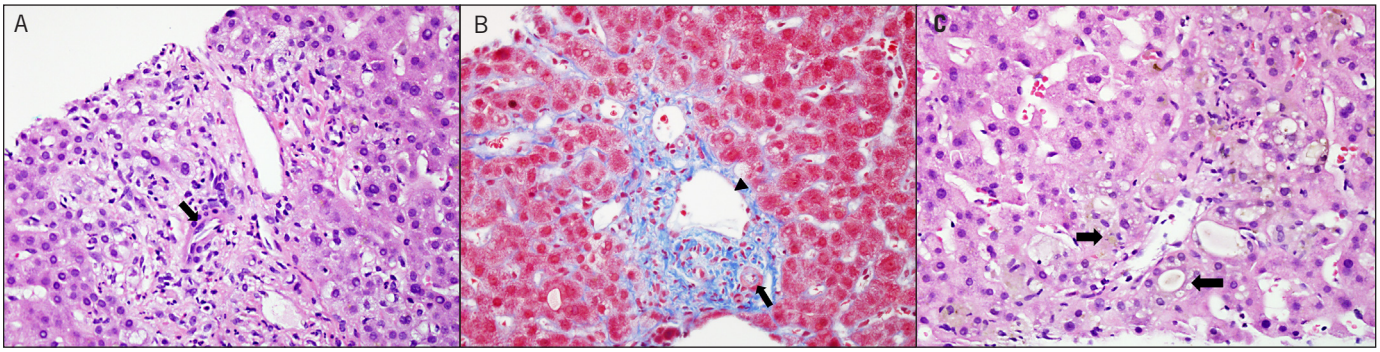

Figure 1. Liver biopsies before diagnosis of ALCL. (A) Bile duct damage is appreciated in a background of minimal inflammatory infiltration (arrow). (B) Arrow indicates hepatic arteriole and arrowhead points to portal venule without the presence of interlobular bile duct. (C) Cholestasis and hepatocyte injury are present predominately in the perivenular region (arrow).

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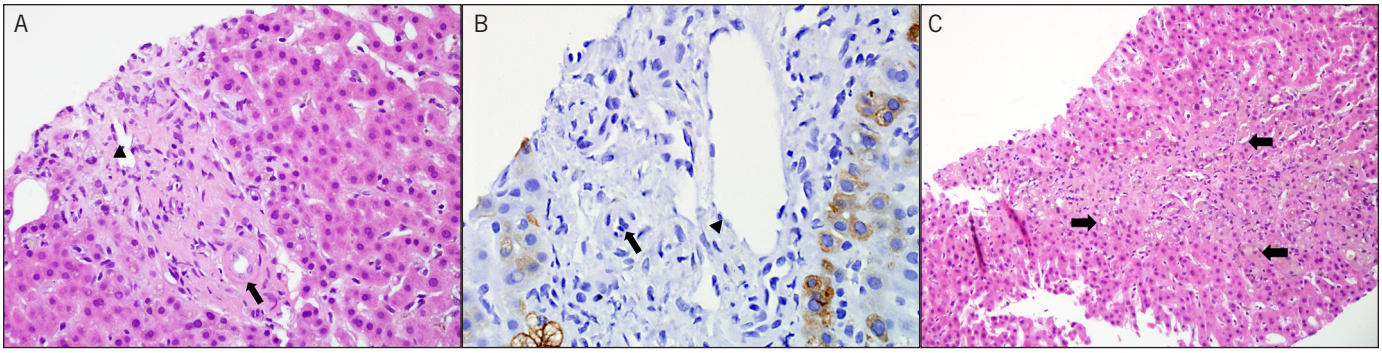

Figure 2. Liver biopsies 10 weeks after diagnosis of ALCL and chemotherapy. Arrow indicates hepatic arteriole and arrowhead indicates portal venule. (A) Complete loss of bile duct resulting in ductopenia. (B) CK7 staining confirms the complete absence of bile duct. (C) Arrow indicates severe cholestasis and perivenular hepatocyte injury.

Disclosures

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