

An Uncommon Presentation of a Common Disease: Periportal Infiltrating Form of Hepatic Lymphoma

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J Gastrointestinal Abdominal Radiol ISGAR

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Hepatic lymphoma with primary or secondary involvement includes three characteristic morphological patterns: focal nodular, diffuse infiltrative, and mixed infiltrative-nodular. However, periportal infiltrating form of hepatic lymphoma is an extremely rare entity, and only few cases are reported in the literature.¹ We reported a histopathologically proven case of diffuse periportal infiltrating hepatic lymphoma and described the ultrasonography (USG), computed tomography (CT), and magnetic resonance imaging (MRI) findings.

A 44-year-old male patient presented with complaints of slowly progressive abdominal discomfort and distension since 6 to 8 months. The patient did not have any history of fever, icterus, night sweats, or weight loss. Physical examination per abdomen revealed hepatosplenomegaly. Laboratory investigations including liver function tests were normal and the patient was negative for hepatitis B surface antigen. Serum alpha-fetoprotein and CA19-9 were within normal limits.

USG abdomen revealed hepatomegaly with periportal linear hypoechoic areas around the portal vein and its branches (►Fig. 1B). On color Doppler no vascularity was noted; however, the right and left portal vein and its branches appear attenuated in caliber but shows normal hepatopetal flow (►Fig. 1C, D). Gallbladder appeared partially distended and shows homogeneously hypoechoic mural thickening with maintained hyperechoic mucosal outline (►Fig. 1A). USG abdomen also revealed splenomegaly, enlarged mesenteric and retroperitoneal lymph nodes, and mild ascites.

Contrast-enhanced CT abdomen revealed hepatomegaly with ill-defined linear mildly enhancing hypodensity along the portal triad surrounding the portal vein and its branches

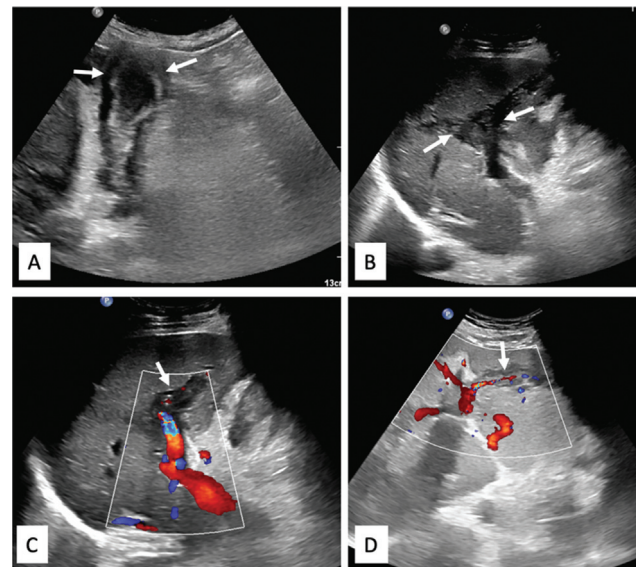


Fig. 1 Grayscale ultrasonography (USG) images (A, B) showed partially distended gallbladder with homogeneously hypoechoic mural thickening with maintained hyperechoic mucosal outline and linear periportal hypoechoic areas. Color Doppler images (C, D) revealed attenuated caliber of the left and right portal vein branches.

which show irregular outline and attenuated caliber, however, normal contrast opacification (►Fig. 2A).

Gallbladder was partially distended and showed diffuse irregular mildly enhancing mural thickening (►Fig. 2B) with maximum wall thickness of 15 mm noted involving the body region. Common hepatic and cystic duct showed hazy outline and hypodense intraluminal contents within with non-visualization of central biliary radicles and mild bilobar

DOI <https://doi.org/10.1055/s-0043-1777297>.
ISSN 2581-9933.

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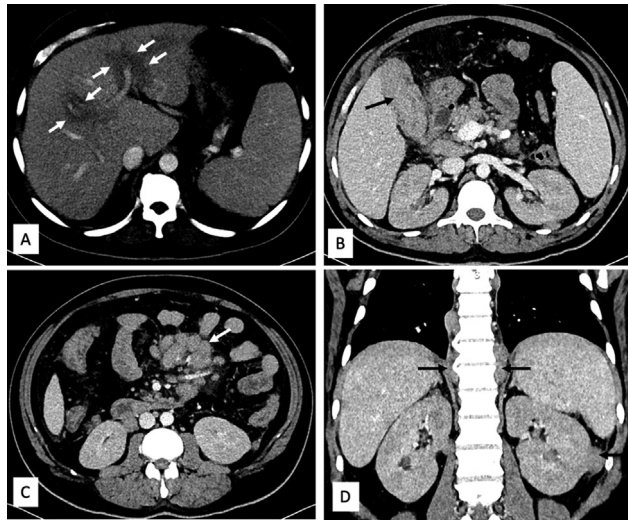


Fig. 2 Axial contrast-enhanced computed tomography (CECT) image at the level of portal vein bifurcation (A) shows linear circumferential periportal hypodensity along the right and left portal vein branches (white arrows). Axial CECT image (B) shows extensive gallbladder mural thickening and pericaval lymph node. Axial CECT image (C) shows enlarged conglomerated mesenteric lymph node encasing the mesenteric vessels. Coronal CECT image (D) shows paravertebral and left perirenal enhancing soft tissue deposits.

peripheral intrahepatic biliary radicles dilatation. Common bile appears normal in caliber and shows no abnormal wall thickening.

Multiple discrete as well as conglomerated enlarged homogeneously enhancing lymph nodes are noted in the mesentery (→ Fig. 2C), aortocaval, along the right anterior perirenal fascia, left common and internal iliac vessels, and bilateral inguinal regions. Multiple well-defined homogeneously enhancing lesions were noted in the bilateral perirenal and prevertebral and paravertebral region (→ Fig. 2D) along lower thoracic spine extending from T8 to T12 vertebrae.

Few homogeneously enhancing peritoneal deposits were also noted abutting the parietal peritoneum along with mild ascites suggestive of peritoneal spread of the disease.

Limited MRI sections were acquired on 3T extending from dome of diaphragm to upper pole of kidneys with axial T2 HASTE, T1 VIBE, diffusion-weighted imaging (DWI) (b50, 400, and 800), and T2 TRUFI sequence.

The lesions in the periportal space, thickened gallbladder wall, bilateral perirenal and paravertebral spaces, and visualized lesions in the mesentery and peritoneum appeared hypointense on T1-weighted imaging (T1WI), mildly hyperintense on T2WI (compared to liver parenchyma), and marked diffusion restriction on DWI with corresponding low apparent diffusion coefficient values (→ Fig. 3).

On histopathology and immunohistochemistry, the mass was diagnosed as non-Hodgkin's hepatic lymphoma—diffuse large B cell type.

Primary hepatic lymphoma (PHL) is defined as a lymphoma that is confined to the liver and perihepatic lymph nodal

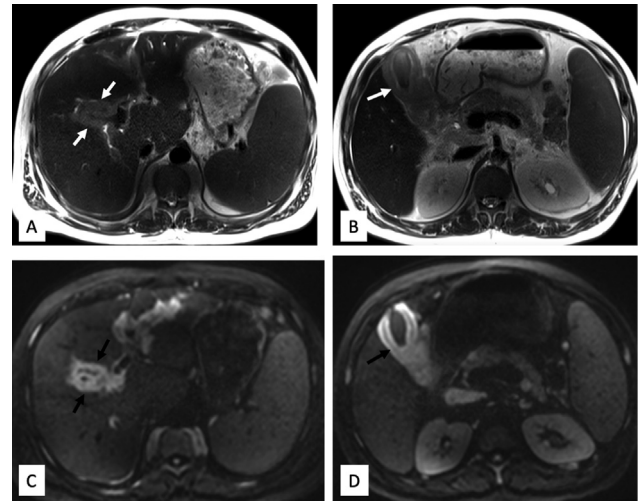


Fig. 3 Axial T2 HASTE sequence (A) shows T2 hyperintensity in the periportal region along the left and right portal veins branches. Axial T2 HASTE sequence (B) shows homogenous hyperintense gallbladder mural thickening. Axial diffusion-weighted imaging (DWI) images (C, D) show corresponding restricted diffusion in the periportal region and gallbladder wall thickening.

sites at initial presentation in the absence of distant lymphadenopathy, splenomegaly, bone marrow disease, and leukemia for at least 6 months after the liver lesion(s) has been detected. It constitute less than 1% of all non-Hodgkin's lymphoma cases.² Diffuse large B-cell lymphoma comprises the majority of cases of PHL. The most common imaging manifestation is a solitary discrete lesion seen in 60% of cases. Multiple lesions are seen in 35 to 40% of patients, with one of the lesions being typically dominant. It can rarely present as ill-defined at the porta hepatis.

Secondary hepatic lymphoma is typically diffusely infiltrating rather than a focal mass, presenting as hepatomegaly, multiple small nodular lesions, or rarely periportal infiltrating form. The latter variant manifests as linear ill-defined or circumscribed mass encasing the portal vein and may cause its attenuation or thrombosis; however, the incidence of venous involvement is significantly lower than in hepatocellular carcinoma.² Persistence of this periportal disease may suggest refractoriness to chemotherapy or disease exacerbation.³

A variety of neoplastic as well as nonneoplastic condition can affect the periportal space, resulting in periportal hypodensity by either impeding the lymphatic drainage or retrograde dissemination of malignant cells from the hilum. Common differentials include extramedullary hematopoiesis, chloroma, neurofibroma, Langerhans cell histiocytosis, periductal cholangiocarcinoma, and malignant lymphadenopathy at the porta hepatis.³

Hepatic lymphoma is a rare disease that generally shows nonspecific imaging findings, such as periportal hypodensity, and leading to diagnostic dilemma on ultrasound and CT. Therefore, the radiologist should be aware of the

spectrum of atypical imaging manifestations of hepatic lymphoma along with its important differential diagnosis.² MRI owing to its excellent soft tissue characterization is the ideal modality of choice for evaluation of periportal infiltrating lymphoma with DWI being the most specific sequence.

Conflict of Interest

None declared.

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