Pseudo-Tumor of the Caudate Lobe in Primary Sclerosing Cholangitis

Arye Blachar MD

Department of Diagnostic Radiology, Tel Aviv Sourasky Medical Center, Tel Aviv, Israel

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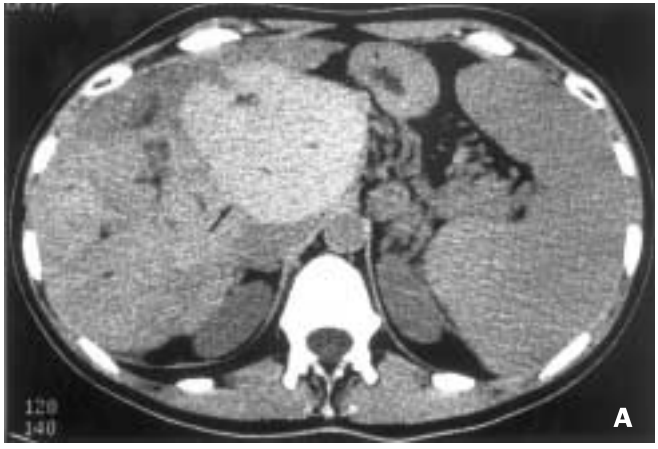

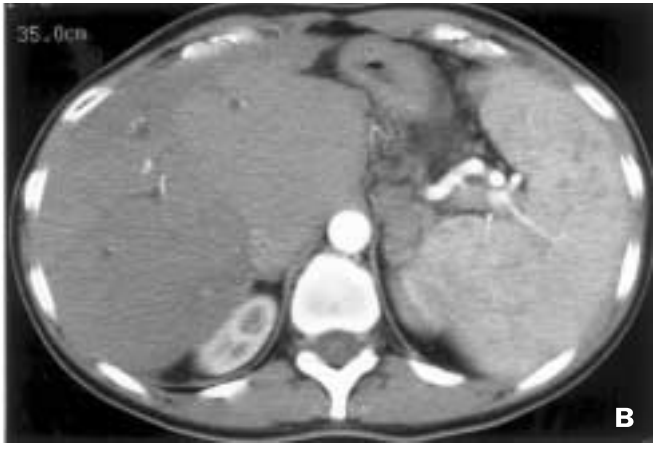

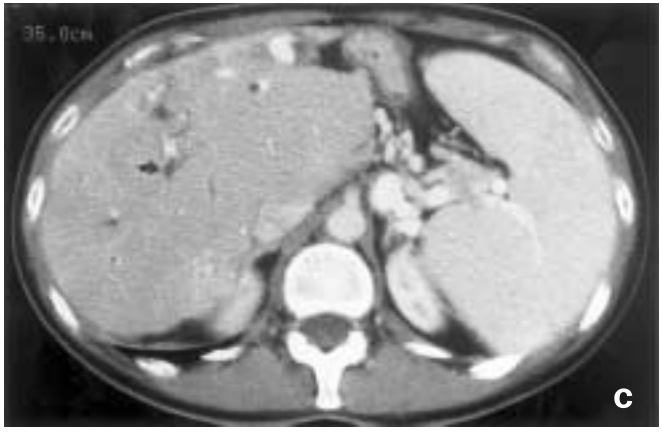

In a 56 year old woman with primary sclerosing cholangitis, non-contrast computerized tomography imaging demonstrated marked hypertrophy of the caudate lobe (arrow) which, combined with the low attenuation of the adjacent liver, manifested as a "pseudo-tumor" [Figure A]. Following rapid intravenous administration of contrast medium and during the arterial phase of enhancement, the caudate remained slightly hyper-attenuated to the adjacent liver [Figure B]. The attenuation difference between the caudate and the liver was lost during the portal venous phase of enhancement [Figure C]. Also note the irregular dilatation of the intrahepatic bile ducts (curved arrow).

Primary sclerosing cholangitis is an idiopathic, chronic, fibros-

ing, inflammatory disease. It occurs in association with inflammatory bowel disease in up to 75% of cases. Approximately 87% of these patients have ulcerative colitis and 13% have Crohn's disease [1]. Although the pathogenesis of the disease is unknown, evidence suggests an autoimmune etiology. Multifocal strictures develop in the biliary tree and eventually lead to obliteration of the bile ducts and to cholestasis and biliary cirrhosis. The rate of progression of the disease varies, and it may progress slowly for 5–15 years from an asymptomatic stage to cirrhosis in up to 50% of patients [2,3]. The most common cause of death is hepatic failure, followed by development of cholangiocarcinoma seen in up to 20% of patients [2]. At autopsy, cholangiocarcinoma is detected in 30–40% of patients. The tumors originate most frequently from the common hepatic duct and its bifurcation, and less frequently from the common duct, cystic duct and intrahepatic bile ducts. Medical treatment is palliative, and orthotopic liver transplantation is the only curative therapy. PSC cholangitis is today the fourth leading indication for liver transplantation in adults in the USA [2].

Patients with PSC are evaluated with CT, cholangiography and endoscopic retrograde cholangiopancreatography. Recently, magnetic resonance imaging and especially MR cholangiopancreatography were shown to be useful in the evaluation of these patients [2,3].

The liver of patients with chronic PSC and cirrhosis demonstrates a lobular contour with parenchymal atrophy involving the peripheral portions of the liver, especially the lateral and posterior segments [4]. The caudate lobe is the most common region of compensatory hypertrophy seen in nearly all patients with PSC. These morphologic changes appear to be related to the chronic multifocal obstruction of the intrahepatic bile ducts, with relative sparing of the bile ducts of the caudate. Other CT findings reported with PSC include: scattered focal areas of irregular peripheral ductal dilatation separate from the hilar ducts, sometimes with a "beaded" appearance; bile duct wall thickening and nodularity; increased enhancement of the bile duct walls; and the radiographic manifestations of portal hypertension such as ascites, splenomegaly and varices [4,5].

PSC = primary sclerosing cholangitis
MR = magnetic resonance

The PSC-induced end-stage liver demonstrates CT findings that are different from those observed in other types of cirrhosis and may suggest the diagnosis. The pseudo-tumor of the caudate lobe should be recognized to avoid misdiagnosis of a neoplasm.

References

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Correspondence: Dr. A. Blachar, Dept. of Diagnostic Radiology, Tel Aviv Sourasky Medical Center, 6 Weizmann St., Tel Aviv 64239, Israel.
Phone: (972-3) 697-3504
Fax: (972-8) 947-1380
email: ablachar@hotmail.com