Fibrolamellar Hepatocellular Carcinoma – An Uncommon Tumor

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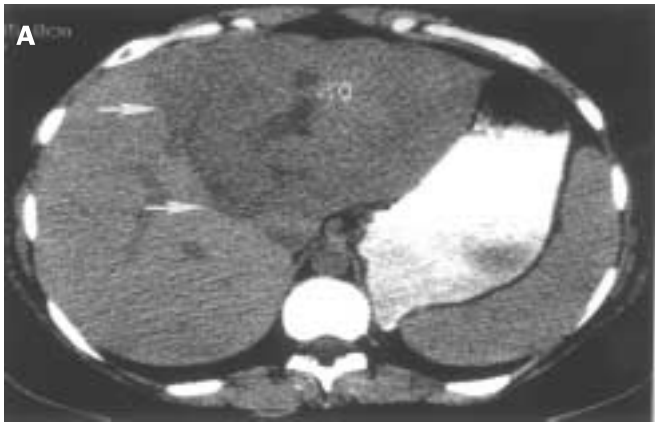
An 18 year-old girl was evaluated for a hepatic mass by axial computed tomography imaging [Figure 1]. Non-contrast-enhanced axial CT image demonstrated a large lobular mass (straight solid arrows) in the left liver lobe that was hypo-attenuating to the liver, with a central hypo-attenuating area consistent with a central scar. Punctate calcifications were present within the scar (open arrow) [Figure 1A]. Axial contrast-enhanced CT section during the hepatic arterial phase showed heterogeneous enhancement of the mass [Figure 1B]. During the portal venous phase the mass was further enhanced and the central scar was better seen [Figure 1C]. A photograph of the resected specimen shows a lobular tumor with a clear central scar and stellate fibrous septa (black arrows) [Figure 1D].

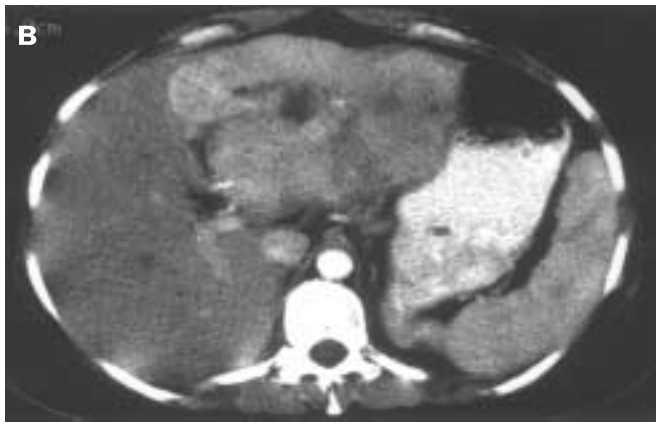
Fibrolamellar hepatocellular carcinoma is an uncommon tumor that is usually seen in adolescents and young adults lacking underlying liver disease. Therefore, although FL-HCC accounts for only 1–9% of hepatocellular cancer cases, it represents up to 35% of HCC cases in patients younger than 50 years of age with a non-cirrhotic liver [1]. It has no reliable serum tumor markers. FL-HCC is usually a large (>10 cm), solitary, hypervascular, heterogeneous mass that demonstrates well-defined, lobulated margins. It lacks large areas of necrosis and hemorrhage that are typical of conventional HCC [2]. FL-HCC is usually hypodense relative to liver on non-contrast-enhanced and portal venous phase images, and demonstrates heterogeneous, marked enhancement on hepatic arterial phase images. A large scar that can be broad or stellate, eccentric or central, is seen on helical CT in up to 71% of patients, with radiating fibrous septa and calcifications present in 68% of these [2]. Histologically, irregular fibrolamellar bands of dense collagen coalesce to form a scar and separate hepatocyte-like tumor cells that are arranged in cords and sheets. The fibrous tissue within the scar and its radiating septa is hypodense on non-contrast, hepatic arterial phase and portal venous phase images, but may demonstrate delayed enhancement when imaged within 10–20 minutes after intravenous contrast administration.

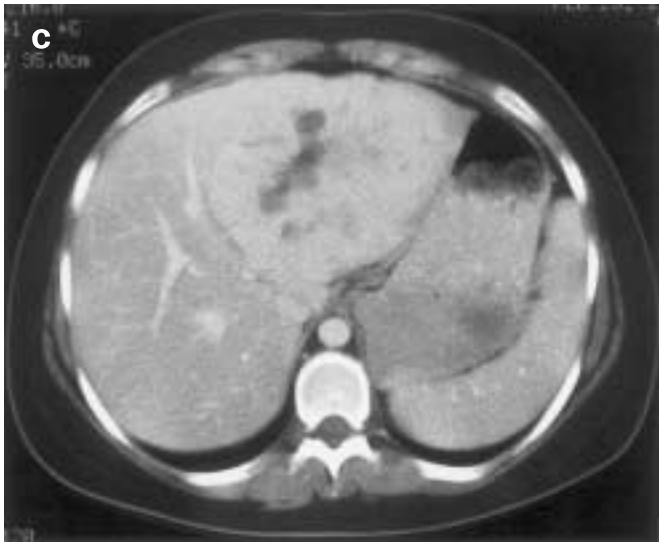
Metastatic abdominal lymphadenopathy occurs frequently (65% of patients), most commonly involving the porta hepatis and the hepatoduodenal ligament, and sometimes shows intense enhancement on the hepatic arterial phase. Pulmonary and peritoneal metastases develop in most patients despite attempts at curative resection.

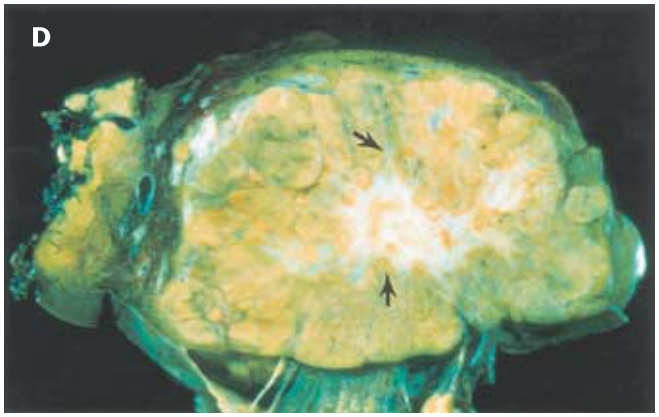
The main differential diagnosis of FL-HCC includes other liver tumors with a central scar. A central area of scarring has been described in several liver tumors, but most commonly with FL-HCC, large liver hemangioma and focal nodular hyperplasia. Large hepatic hemangiomas are characteristically diagnosed in older patients; they demonstrate nodular peripheral enhancement that is iso-attenuating with the blood vessels and rarely have intratumoral

FL-HCC = fibrolamellar hepatocellular carcinoma









calcifications. Cleft-like central or eccentric areas of scarring are often seen and usually do not show delayed contrast enhancement. Focal nodular hyperplasia is seen in young women. It is usually small in size, tends to be peripheral, has a smooth border and demonstrates homogeneous intense enhancement during the hepatic arterial phase. It is usually iso-attenuating to the liver on the portal venous phase. The scar is small and shows delayed contrast enhancement. Intratumoral or scar calcifications are exceedingly rare. A recently published study [3] aimed at determining the performance of the radiologist in the diagnosis of these tumors showed that CT allowed accurate differentiation of these tumors, with FL-HCC diagnosed most accurately.

References

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in Gratitude

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Errata

In the case report "Critical myocardial ischemia: minor electrocardiographic changes – Wellen's syndrome" by Y. Goor, R. Magal, O. Goor, A. Frimerman and S. Cabili (February issue, 2003;5:129–30), the following changes should be noted: a) In the first sentence of the Patient Description, the phrase within parentheses should read: (treated with lovastatin 20 mg a day) and not (..... lovastatin 20 mg 4 times a day). b) The first sentence in the second paragraph on page 130 should read as follows: While the variant with the deeply inverted T waves, which occurs in 76% of patients, is well recognized and will usually prompt aggressive therapy, the variant with biphasic T waves may be overlooked as "non-specific T wave changes," as described in the 2000 ACC/AHA guidelines for the management of patients with unstable angina [1]. [In the printed article, the words "inverted" and "biphasic" were erroneously transposed]

In the Letters section (February issue, 2003;5:151), the letter "The reemergence of sexually transmitted infections in Israel," was written by Michael Dan MD, and not Dan Michaeli MD.