Fibrolamellar Hepatocarcinoma: Clinical, Radiologic, and Pathologic Features

Isaac R. Francis,1 Farooq P. Agha,1 Norman W. Thompson,2 and David F. Keren3
Departments of 1 Radiology, 2 Surgery, and 3 Pathology, University of Michigan Hospitals, Ann Arbor, Michigan, USA

Abstract. Three new cases of an unusual subtype of hepatocellular carcinoma (HCC) referred to as fibrolamellar hepatocarcinoma (FLHC) recently seen at our institution are described. This report focuses on the clinical, radiologic, and pathologic features of this rare subset of HCC. All three patients were under 30 years of age with no previous history of hepatitis or cirrhosis. Each had had subacute symptoms for 5 months to 1 year before medical attention was sought and/or diagnosis of FLHC was established. There was no reliable correlation with oral contraceptive use in the 2 female patients. Serum alpha-1-fetoprotein levels were normal with only mild elevation of liver enzymes. The CT features, although not specific, were suggestive of an aggressive tumor with amorphous calcification in 2 of the 3 cases. Angiographically all tumors were hypervascular without any evidence of arterioporal shunting or venous invasion of major vessels. The clinical and radiologic recognition of these tumors is important since the surgical resectibility rate and 2- and 5-year survival rates are higher than those applicable to conventional HCC.

Key words: Liver tumor – Hepatoma – Hepatocellular carcinoma – Fibrolamellar hepatocarcinoma.

Hepatocellular carcinoma (HCC) is a relatively rare hepatic neoplasm in the western hemisphere, occurring with an incidence of 1:100,000, predominantly in men over 50 years of age. The duration of symptoms is short and prognosis usually poor, with mean survival measured in months only. A distinct subtype of HCC has been recently classified as fibrolamellar hepatocarcinoma (FLHC) due to its characteristic clinical and pathologic features [1–9]. We report 3 newly diagnosed cases of FLHC at our institution during the past 2 1/2 years. The analysis of clinical, radiologic, and pathologic features with surgical management and follow-up from 15–33 months in this group of patients constitutes the basis of this report.

Case Reports

Case 1
A 22-year-old white woman presented with a 3-day history of right upper quadrant pain associated with nausea, vomiting, and a 44-kg weight loss. There was no present or past history of oral contraceptive use. Physical examination revealed a tender mass in the right upper quadrant with a venous hum. The serum bilirubin and alpha-1-fetoprotein levels were normal with a mild elevation of liver enzymes. CT scan of the abdomen demonstrated (Fig. 1 A) a large mass involving most of the right lobe of the liver and a separate large mass near the porta hepatis compressing the second part of the duodenum. Areas of amorphous calcifications were present within the tumor mass. Hepatic angiography demonstrated a large hypervascular mass in the right lobe of the liver without arterioporal shunting or venous invasion (Fig. 1 B) and a large hypervascular mass in the region just below the porta hepatis. At surgery, a large inoperable mass confined to the right lobe of the liver and a single large metastatic node posterior to the porta hepatis were found compressing the duodenum. Extrahepatic spread precluded right hepatectomy. A liver biopsy and excisional lymph node biopsy were performed. The histopathologic diagnosis was FLHC. Postoperatively she received chemotherapy consisting of adriamycin, mitomycin C, and 5-fluorouracil. Reevaluation 10 months after initial surgery revealed that tumor was confined to the liver; therefore, she underwent an elective right lobe hepatectomy and excision of porta hepatitis nodes. She did well until a repeat exploratory laparotomy revealed recurrent tumor in the liver and porta hepatis. Thirty-three months after initial diagnosis, the patient is still alive with recurrent disease.
Fig. 1. Case 1. A A contrast-enhanced CT scan shows a large hyperdense mass in the right lobe of the liver (black arrows) with an area of low attenuation (white arrow) corresponding to tumor necrosis and amorphous calcification (arrowhead). B A select subtraction view from celiac arteriogram shows hypervascular mass with no arterioportal shunting. No venous involvement was evident. C Low-power photomicrograph demonstrating the FLHC: note the characteristic prominent bands of collagen separating the neoplastic cells (hematoxylin and eosin stain, × 47). D High-power photomicrograph from resected specimen shows plump, deeply eosinophilic malignant hepatocytes separated by bands of collagen stroma arranged in parallel lamellae (hematoxylin and eosin stain, × 200)