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Fibrolamellar Hepatocellular Carcinoma: a Case Report with Distinct Radiological Features

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Abstract

Introduction—We report a rare case of a 23-year-old male who presented with abdominal discomfort for 15 days. An ultrasound was performed which showed a hypoechoic, heterogenous mass in the left lobe of the liver and distended portal vein, followed by further investigation with computed tomography (CT), MRI, and MRA. Serum alpha-fetoprotein was not elevated and hepatitis B antigen was negative.

Methods—CT scan depicted a nodular mass in left liver lobe with occlusion of both the central part and the two main branches of intrahepatic portal vein.

Result—Biopsy of the liver mass led to a diagnosis of fibrolamellar hepatocellular carcinoma.

Conclusion—Fibrolamellar carcinoma is an uncommon variant of hepatocellular carcinoma. The diagnosis is suggested by radiographic studies and is confirmed by histological examination.

Keywords

fibrolamellar carcinoma; portal vein occlusion; radiology

Introduction

Fibrolamellar hepatocellular carcinoma has long been recognized as an unusual tumor with distinct clinical, histopathologic, and imaging findings, initially described by Edmonson in 1956. In 1980, Craig et al. and Berman et al. established the clinical diagnosis.

Although it is uncommon, it represents a considerable proportion of hepatocellular carcinoma in patients younger than 50 years of age with noncirrhotic liver, usually between the 2nd and 3rd decade of life, with men and women being at equal risk.

Case Report

A 23-year-old man with no previous pathological history was admitted to our hospital, in August 2008, with an abdominal discomfort for the past 15 days.

The patient is a regular blood donor; but, at his last session he could not donate blood because a very low platelet count (lower than normal level) was discovered.

His laboratory tests showed only a mild elevation of SGPT 65 U/l, total bilirubin 2.32 mg/dl (direct bilirubin, 0.65 mg/dl), vitamin B12 880 pmol/l, and PLT count of 152 K/ μ l.

The physical examination revealed a firm nodular mass in the epigastrium on palpation, and the patient was referred to our ultrasound department for further investigation.

Imaging Studies

The ultrasound (US) examination showed an enlarged left liver lobe occupied by a solitary, irregular, heterogeneous mass with mixed echogenicity, measuring 9.5 cm in its largest diameter, with distended and occluded portal vein [Fig. 1]. Color Doppler imaging and power Doppler imaging depicted increased vascularization within the tumor [Fig. 2]. Splenomegaly was also found.

Abdominal computed tomography (CT) was performed the same day before and after intravenous contrast administration in arterial, portal, and delayed phases.

On nonenhanced CT scan, the tumor appeared hypodense with well-defined lobulated margins, with markedly distended portal vein and its two major branches [Fig. 3]. During the arterial phase of dynamic enhanced CT, the tumor showed prominent and heterogeneous enhancement with a more central portion remaining hypodense (central scar), contrast medium enhancement of the thrombus in the portal vein, and abnormal blood flow around the distribution of portal vein. During the portal phase, the central scar remained hypodense, and the blood flow was absent in the left and right branches and the central part of portal vein. The delayed phase showed relative homogeneity of the tumor with mild enhancement of the central scar [Figs. 4, 5].

There was no lymph node involvement, and the chest CT scan was negative for metastases.

Pathology

The following day, a liver biopsy was scheduled to establish the diagnosis. The biopsy sample showed proliferation of large cells containing large nuclei with abundant and eosinophilic cytoplasm. Cytoplasmic paranuclear pale bodies were also found. The cells were arranged in a trabecular pattern and were segregated by deposition of fibrous connective tissue, predominantly collagen, with lamellae formation. In included portal vein parts, neoplastic cells were observed corresponding to neoplastic thrombi. Bile was found in the tumor cells.

Immunohistochemical studies showed that the neoplastic cells were Hep-Par1- and cytokeratin 7-positive, reactive to monoclonal carcinoembryonic antigen but negative for AFP and cytokeratin 19, strongly suggestive of fibrolamellar hepatocellular carcinoma (FL-HCC). The proposed diagnosis was fibrolamellar hepatocellular carcinoma.

Clinical Course

Due to extensive portal vein thrombosis, the tumor was inoperable, and the patient is on the list for liver transplantation. Abdominal ultrasound every 15 days and abdominal Liver MRI every month are scheduled to monitor the progression of the tumor.

Discussion

FL-HCC is a rare primary liver carcinoma occurring predominantly in young adults between the 2nd and 3rd decade of life. On the basis of 182 cases reported in the literature, the age at presentation shows a unimodal distribution with a peak in the late teens and an overall average age of 24.8 ± 13 years. In contrast, an average age of 39 ± 20 years was reported for 68 individuals from the Surveillance, Epidemiology, and End Results (SEERs) database [1].

The etiology of FL-HCC is unknown. FL-HCC does not generally arise in the setting of any known chronic liver disease, hepatitis B infection, or the use of oral contraceptives [1–4]. The serum alpha-fetoprotein is usually not elevated, but elevations in vitamin B12 binding capacity have been reported [5,6].

Histologically, the tumor is made up of large polygonal cells with abundant eosinophilic cytoplasm, large vesiculated nuclei, and large nucleoli in conjunction with the lamellar fibrosis, which are the defining features of FLC [3,7,8].

Clinical findings at presentation of FL-HCC are, in most cases, vague and nonspecific, often with elements of abdominal pain, weight loss, and malaise. The most common physical finding is an abdominal mass or hepatomegaly [6,8–10].

Imaging studies play a major role in diagnosis, identifying, and differentiating FL-HCC from other primary liver tumors. Current US, CT, MR, and FDG PET techniques allow accurate assessment of major hepatic vascular invasion and lymphadenopathy and are useful for both initial assessment and surveillance for recurrence [2,5–8,10–12].

The treatment of choice is surgery and resection of the tumor or liver transplantation for unresectable tumors because FL-HCC does not respond better than typical HCC to chemotherapy.

The prognosis and survival rate of FL-HCC are, until today, very debatable [1,5,11], but, in summary, the weight of the evidence indicates that FL-HCC has a survival advantage over most cases of ordinary HCC largely on account of the absence of cirrhosis. It can be an aggressive tumor with significant mortality at 1 and 5 years, even for those who undergo complete resection, due to high incidence of recurrence.

In conclusion, FL-HCC is a rare histologic variant of common HCC and should be suspected in patients with HCC who are young and who do not have underlying hepatitis or cirrhosis. These tumors have characteristic CT and MRI appearances that often allow their identification before resection.

We reported a rare case of FL-HCC with distinct imaging features. To our knowledge, there are very few cases of FL-HCC in the literature with such an extensive portal vein thrombosis at initial presentation, indicating the severity of the disease despite its mild clinical findings.

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Fig. 1.
Large, irregular, hypoechoic tumor in left liver lobe [a], distended and occluded portal vein [b]

Fig. 2.
Increased tumor vascularity CDI [a], PDI [b]

Fig. 3.
Distended portal vein, left–right branch and central part (*arrows*) [a], hypoattenuating mass in left liver lobe (*arrow*) [b]

Fig. 4. Central scarring in arterial and portal phases, homogeneity of the tumor and mild enhancement of central scar in delayed phase (arrow)

Fig. 5.
Abnormal blood flow and thrombus enhancement in arterial phase and absent blood flow in portal vein during portal phase