Perihepatic Lesions Mimicking Primary Liver Tumors

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Key words: perihepatic tumor, magnetic resonance imaging, computed tomography

IMAJ 2003;5:204–206

With the increasing use of ultrasonography for evaluation of vague abdominal pain, "incidental" hepatic lesions are more often encountered. Although computed tomography arterial portography is most commonly used and is highly sensitive to assess liver tumors before resection [1], it is not always helpful to distinguish extrahepatic lesions from primary liver tumors, especially when the tumor is hypovascular. Here we describe three patients in whom perihepatic masses were initially misdiagnosed as primary hepatic lesions due to misinterpretation of the preoperative imaging studies.

Patient Descriptions

Patient 1

A 66 year old woman was referred for resection of a "liver mass" found incidentally on ultrasonography 5 years prior to her referral. This mass had not changed in size during the follow-up period and was considered to be a benign liver tumor. Because of its location in proximity to the hilar structures, biopsy was not performed. Indications for surgery included the recent onset of symptoms, including weight loss and fatigue associated with anemia (6.2 g/dl hemoglobin) and elevated serum liver enzymes (alkaline phosphatase 445 IU/L, gamma-glutamyl transpeptidase 235 IU/L) and serum bilirubin (2.8 mg/dl). Serologic tests for hepatic viruses and tumor markers

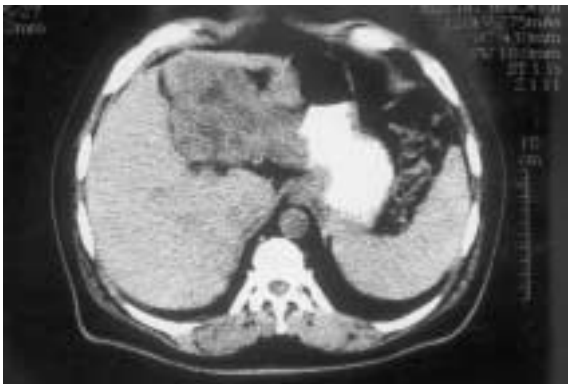
(alpha-fetoprotein, CA 19-9 and carcinoembryonic antigen) were negative. CT scan and MRI showed a large (5 x 6 x 6 cm) caudate lobe mass encasing the vena cava and compressing the hilar structures. At surgery, the mass was found to be a large aggregation of lymph nodes at the hepatoduodenal ligament extending posteriorly to enwrap the vena cava. The mass was removed and histologic examination revealed a malignant B cell lymphoma composed of diffuse anaplastic large cells within lymph node hyperplasia.

Patient 2

A 62 year old man was admitted to the hospital with anemia (9.8 g/dl hemoglobin) associated with weight loss and upper abdominal discomfort. The patient was aware of a "benign" liver mass that had been present for at least 3 years. On physical examination, a large irregular mass below the sternum could be palpated. Laboratory studies, including serum liver enzymes and αFP levels, were normal. CT with contrast [Figure] and MRI demonstrated a large vascular liver mass at the left lateral segment compressing the stomach at the lesser curvature. Core needle biopsy of the tumor showed histologic features consistent with epithelioid hemangioepithelioma. At operation the mass was found to originate from the gastric wall at the lesser curvature. The tumor compressed the adjacent liver, and the left lateral segment seemed atrophic. A subtotal gastrectomy was performed, and histologic studies revealed a malignant gastric stromal tumor in the submucosa invading the muscularis mucosa. Immunohistochemical stainings were positive for α-1 actin and negative for s-100, neuron-specific enolase, synaptophysin and chromogranin.

Patient 3

A 6 year old girl presented with a huge abdominal mass. Imaging studies, including ultrasonography and CT, suggested a mass involving most of the right lobe of the liver, with multiple small nodules studding the hepatic parenchyma. Serum αFP levels were markedly elevated (276,000 ng/ml), and open biopsy findings were consistent with a germ cell tumor. Combination chemotherapy was instituted, with a marked drop in αFP serum levels (63 ng/ml). On repeated CT and MRI studies, the tumor seemed to have shrunk and was confined to the right lobe of the liver.



Patient 2. CT scan with contrast showing an exophytic mass of a gastric tumor, which was mistakenly diagnosed as an epithelioid hemangioendothelioma in the left lateral segment of the liver.

At operation, the tumor was a large (10 x 8 x 7 cm) retroperitoneal germ cell tumor that compressed the vena cava. The right lobe of the liver appeared smaller in size than the left lobe. The tumor was completely resected and the child made an uneventful recovery.

Comment

Perihepatic tissue at the porta hepatis or the portocaval space may be the origin of lymphatic tumors. Because of their location and enhancing characteristics on dynamic CT, these hypervascular lesions may be difficult to differentiate from benign liver tumors such as focal nodular hyperplasia and hepatocellular adenoma [2]. In its benign form, Castleman disease, a localized lymph node hyperplasia with marked capillary proliferation, has been reported to mimic liver tumor [2]. In our first case, the perihepatic mass was similar to masses seen with Castelman disease. Unlike the benign condition in the localized form of Castelman disease, in our case the histology from the enlarged lymph node revealed a B cell lymphoma. The 5 year history of a persistent mass may suggest transformation of a benign lymph node hyperplasia into malignant lymphoma.

Extrahepatic lesions may originate from different structures surrounding the liver, including muscle, nerve, lymphatic, fat or vascular tissue. Adrenal and gastric tumors may also simulate primary liver lesions. In our second patient, the gastric origin of the tumor was clear during exploration, while preoperative radiology studies and histologic findings of the biopsy were misleading. Gastric stromal tumors are unusual malignancies characterized by poorly differentiated cells showing features of either smooth muscle or neural elements [3]. Diagnosis is established by findings of cytoplasmic neural processes and dense core secretory granules by electron microscopy and positive neuron-specific enolase, synaptophysin, and chromogranin A by immunohistochemistry [3].

Extragonadal germ cell tumors are found in the retroperitoneum and, like hepatoblastomas, are frequently associated with elevated serum αFP level. On CT, these tumors are large, lobulated and of mixed density. Fat plane obliteration against adjacent structures is frequent, and both the aorta and the vena cava can be embedded within the tumor. Although rare, germ cell tumors can be found within the liver [4]. In our third patient, despite the distinct histologic characteristics of the tumor, we could not precisely define its origin by imaging techniques. Despite our uncertainty regarding the origin of the tumor, we employed currently recommended chemotherapy protocol for stage VI germ cell tumor and continued with resection after reduction of tumor mass.

Interestingly, in two of our three cases we noted atrophy of the adjacent liver segments. The left lobe in one case and the right lobe in the other were compressed but not invaded by the tumor and were much smaller than their expected volume. We speculated that compression of the liver was associated with a reduction of portal inflow, leading to hepatic atrophy, as described in experimental model of Eck fistula [5].

In summary, extrahepatic tumors may

αFP = alpha-fetoprotein

mimic primary hepatic lesions. Care must be taken when interpreting CT and MRI studies, with much attention paid to the tumor/normal liver tissue transition zone. Recognition of specific characteristics of these rare tumors on CT and MRI is important to establish the correct diagnosis preoperatively. Newly developed magnetic resonance technologies such as use of liver-specific MRI contrast agents and methods for increasing the speed and improving the resolution of three-dimensional acquisition are promising.

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