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Primary liver leiomyosarcoma: CT appearance

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Abstract
Primary hepatic leiomyosarcomas are exceedingly rare tumors. To the best of our knowledge, only 17 cases have been reported in literature. We report the computed tomographic findings of two cases of primary location in the liver. We also discuss the differential diagnosis of such lesions.

Key words: Liver neoplasms, computed tomography—Liver neoplasms, magnetic resonance—Leiomyoma.

Leiomyosarcoma is a mesenchymal tumor of ubiquitous origin [1], originating from smooth muscle cells. It is among the most frequent soft-tissue sarcomas, being the most frequent beyond the fifth decade of life. It most commonly arises in the retroperitoneum, alimentary tract, genitourinary tract, and soft tissues. Liver, lung, and peritoneal involvement are also reported to be primary sites of origin, along with other rarer locations [1, 2]. Primary hepatic localization of such malignancy is exceedingly rare; in fact, leiomyosarcomas found in the liver are generally metastatic. To the best of our knowledge, only 17 cases have been reported in the literature.

We present computed tomographic (CT) findings of two cases of primary leiomyosarcoma of the liver, with magnetic resonance (MR) correlation in one of them.

Case Reports

Case 1

A 50-year-old male, with a previous history of gastric ulcer, came to our observation because of aspecific digestive complaints, pain in the right upper quadrant, malaise, and recent weight loss. Laboratory tests were unremarkable. CT of the abdomen before contrast medium administration (Fig. 1A) showed a voluminous mass, with heterogeneous hypodensity, massively infiltrating the right hepatic lobe and the medial segments of the left lobe. At contrast-enhanced CT (Fig. 1B), enhancement, although heterogeneous, of the entire lesion was noted. Within the mass, several foci of nonenhancing hypodensity were present, representing areas of pseudocystic necrosis. On spin-echo MR, T1-weighted images of the liver (Fig. 1C) nicely confirmed both the location and the extent of the mass, which exhibited a homogeneous hypointensity, with a discrete hyperintense area located laterally in the right hepatic lobe. In contrast, the mass appeared heterogeneously hyperintense on T2-weighted images (Fig. 1D), suggesting a malignant nature. The hyperintense area on T1 did not remarkably change signal on the long TE image, probably due to the presence of blood catabolites within the focus of necrosis. Multiple scattered vertebral metastases of D11, L1, and L2 were detected on the coronal images (not shown); they appeared hypointense on the T1- and hyperintense on the T2-weighted images. Histologic examination (Fig. 1E) of a needle-core biopsy of the hepatic mass under US guidance showed spindle-shaped cells, with blunt endings of the nuclei (''cigar-shaped'' nuclei), arranged in interlacing bundles. The nuclei had variable shape and dimensions. Several mitotic figures were detected at ×400 magnification. The final diagnosis was primary hepatic leiomyosarcoma.

Case 2

A 35-year-old female had a previous history of Hodgkin's disease treated 5 years previously with radiation therapy and in complete remission at this time and moderate hypertension for 2 years. She came to our observation because of a weight loss of approximately 8 pounds, low-grade fever (38.5°C), malaise, and a recent rise of arterial blood pressure (180–190 mmHg). An abdominal ultrasonogram showed a highly heterogeneous large mass, located in the right hepatic lobe. At unenhanced CT the mass appeared heterogeneously hypodense and well demarcated (Fig. 2A). After intravenous contrast-medium administration, the lesion exhibited marked contrast enhancement of the peripheral solid portion, with a large central necrotic area (Fig. 2B). After administration of a second bolus of iodinated contrast medium, marked enhancement of the pseudocapsule was detected (Fig. 2C). A partial hepatectomy was then performed at a different hospital. Pathologic examination yielded a diagnosis of primary leiomyosarcoma. At follow-up 6 months later, the patient was still free of disease.

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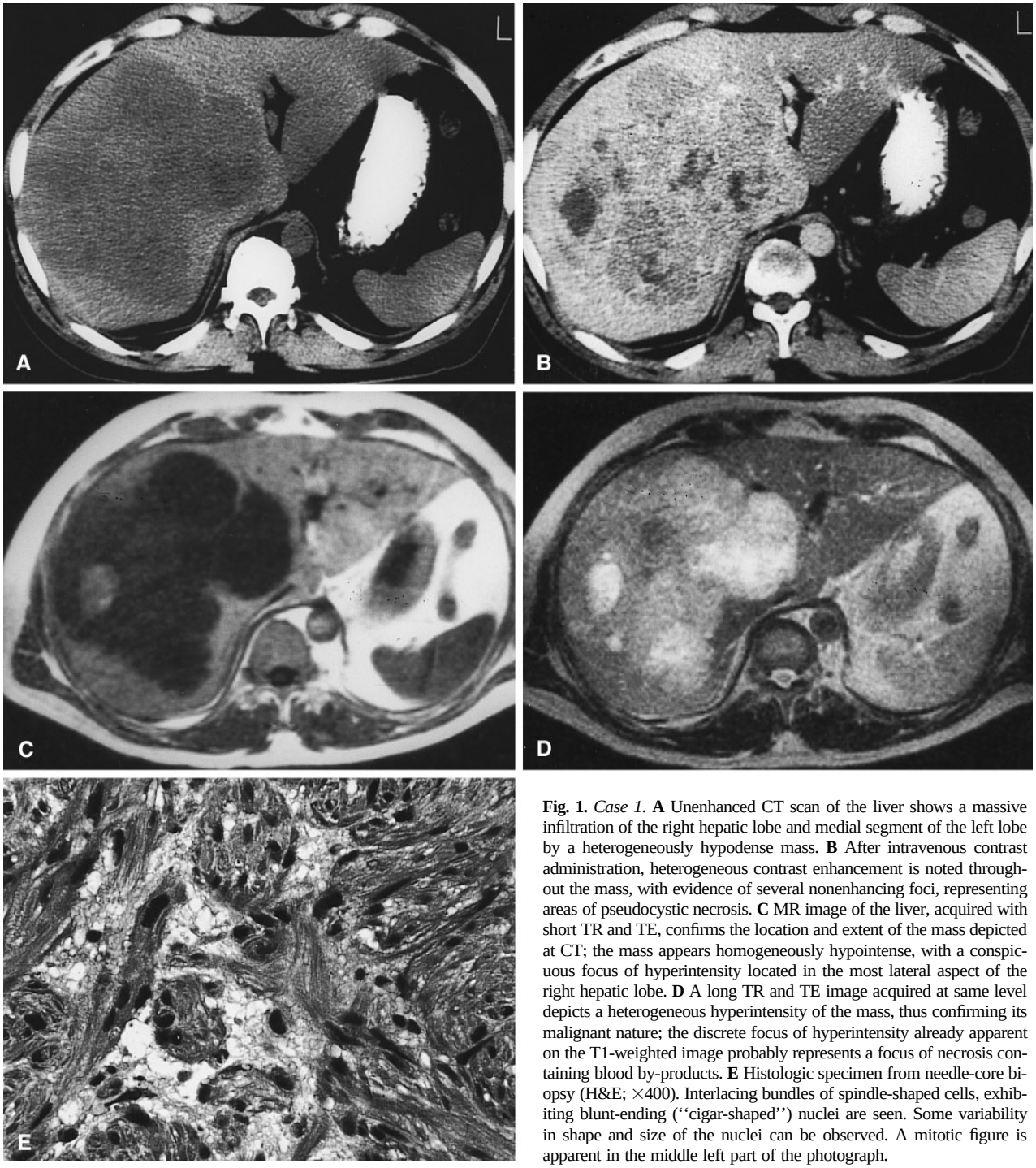

Fig. 1. *Case 1.* **A** Unenhanced CT scan of the liver shows a massive infiltration of the right hepatic lobe and medial segment of the left lobe by a heterogeneously hypodense mass. **B** After intravenous contrast administration, heterogeneous contrast enhancement is noted throughout the mass, with evidence of several nonenhancing foci, representing areas of pseudocystic necrosis. **C** MR image of the liver, acquired with short TR and TE, confirms the location and extent of the mass depicted at CT; the mass appears homogeneously hypointense, with a conspicuous focus of hyperintensity located in the most lateral aspect of the right hepatic lobe. **D** A long TR and TE image acquired at same level depicts a heterogeneous hyperintensity of the mass, thus confirming its malignant nature; the discrete focus of hyperintensity already apparent on the T1-weighted image probably represents a focus of necrosis containing blood by-products. **E** Histologic specimen from needle-core biopsy (H&E; ×400). Interlacing bundles of spindle-shaped cells, exhibiting blunt-ending (''cigar-shaped'') nuclei are seen. Some variability in shape and size of the nuclei can be observed. A mitotic figure is apparent in the middle left part of the photograph.

Discussion and Conclusions

Primary malignant mesenchymal tumors of the liver are much rarer than epithelial neoplasms [3]; they occur ten times more often in children than in adults [4]. Among all soft-tissue sarcomas, primary leiomyosarcoma of the liver is exceedingly rare. As a matter of fact, to assume the diagnosis of primary leiomyosarcoma of the liver, all other possible sites of origin, including the more frequent inferior vena cava and ligamentum teres, must be excluded by careful staging. The possibility that the liver may be secondarily involved by a contiguous lesion primarily located in the stomach, gallbladder, or diaphragm ought to be ruled out [5].

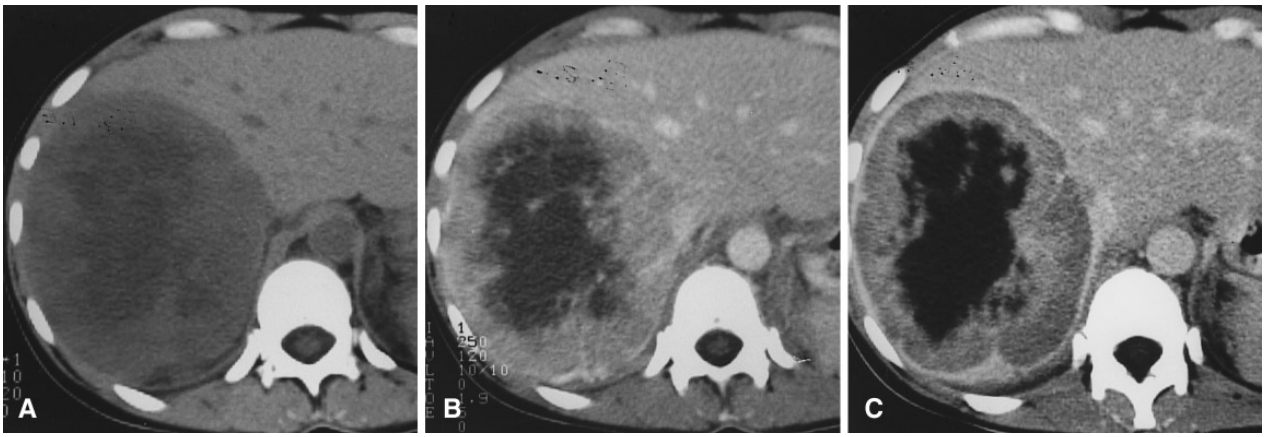

Fig. 2. *Case 2.* **A** Unenhanced CT scan of the liver shows a large mass of the right hepatic lobe, heterogeneously hypodense, with well-defined regular contours. **B** After iodinated contrast-medium administration, definite contrast enhancement of the tumoral solid portion is detected, with evidence of a large central area of necrosis. **C** After a second bolus of contrast is administered, in the early arterial phase, marked enhancement of the pseudocapsule is apparent.

Hepatic smooth muscle cells tumors probably arise from hepatic veins or bile ducts. The differentiation between benign leiomyoma and low-grade leiomyosarcoma still remains problematic, even in modern surgical pathology. Evaluation of the number of mitotic figures per high-power microscopic field has been suggested as the best criterion for diagnosis of malignancy of smooth muscle cell tumors, although the required number of mitoses itself varies according to the site of origin [6].

From the epidemiological standpoint, it is interesting that an increased incidence of both benign and malignant smooth cell neoplasms have been reported in patients with AIDS, particularly in children, and in patients with other forms of immunodeficiencies [6, 7]. Moreover, among the few scattered cases of primary hepatic leiomyosarcomas, several have been detected in patients with another synchronous or metachronous malignancy, such as acute myeloid leukemia, splenic hemangiosarcoma, sigmoid cancer, or cholangiocarcinoma [4]. Our second case was interestingly associated with a previous Hodgkin's disease.

From the clinical point of view, hepatomegaly, a palpable abdominal or right upper quadrant mass, and upper abdominal quadrant pain are the usual presenting findings, due to the generally conspicuous dimensions of the tumor at the time of diagnosis [5, 6]. In both of our patients, tumors developed in the right hepatic lobe, just as in the majority of other reported cases [4].

Although the findings of a large tumor mass with extensive necrosis and/or pseudocystic changes at CT are not specific, inclusion of primary hepatic leiomyosarcoma in the list of differential pathologies is suggested [4]. In fact, much more common lesions may, from time to time, appear as either one of the two we reported. In Europe, 80% of hepatomas occur in cirrhotic patients, and they may show satellite nodules in about 25% of cases but exhibit enhancement only in the most precocious arterial phase and often show a hyperdense rim around the main lesion; further diagnostic aid is offered by their ''geographic'' appearance and the frequent portal vein thrombosis. Fibrolamellar hepatocarcinomas occur in young patients without any underlying risk factors or predisposing pathologies, as in our case 1, but the presence of central calcifications in 40% of cases and the detection of a prominent central stellate scar, rather than colliquative necrosis, are generally helpful hints for the differential diagnosis. A biliary cystadenocarcinoma appears as a complex cystic mass, with variable enhancement of the solid portions, irregular borders, and with occasional satellite nodules. Hepatocellular adenomas are generally suggested by their prevalence in young women with history of oral contraceptive use and their frequent hemorrhagic complications. Metastases may occasionally exhibit features analogous to the lesions described earlier. The differential diagnosis is particularly arduous in respect to other hepatic sarcomas, which are, however, rare lesions.

Among nontumoral conditions, echinococcus alveolaris may offer some diagnostic dilemmas, given its more ''aggressive'' appearance at CT; once again, laboratory tests can lead toward the right diagnosis, supported by the occasional presence of punctate calcifications.

In conclusion, in the presence of a voluminous hypervascular lesion of the liver, suspect for a malig-

nancy, especially with hemorrhagic complications and/or massive liquescent necrosis, a primitive leiomyosarcoma should be considered in the list of the possible differential diagnoses, despite the rarity of the tumor.

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