Primary Hepatic Leiomyosarcoma: A Case Report

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Primary hepatic leiomyosarcoma is a rare tumor, most frequently occurring in liver parenchyma. We recently encountered an exophytic hepatic leiomyosarcoma; CT scans indicated an indistinct border, with the parenchyme of the liver and parenchymal beaking suggesting a primary hepatic mass. We present an unusual case of primary leiomyosarcoma which showed exophytic growth.

Index word: Liver neoplasm, CT

Primary hepatic leiomyosarcoma is a rare tumor and may arise from hepatic veins or bile ducts (1); it should be distinguished from leiomyosarcoma of the ligamentum teres or inferior vena cava (1-3). To our knowledge, there is no description of exophytic growth of this tumor in the radiologic literature (1-5). We present an unusual case of primary leiomyosarcoma of the liver exhibiting exophytic growth.

Case Report

A 62-year-old woman was admitted to our hospital with an abdominal mass. Abdominal computed tomography (CT) performed at another institution a low-density mass projecting into the porta hepatis. Fine-needle biopsy of the mass indicated leiomyosarcoma. The patient was transferred to our hospital for further evaluation and treatment.

We reviewed the abdominal scans, and these indicated that the mass was bilobulated, with a huge low-density area in the central portion, suggesting tumor necrosis. The border of the mass was relatively well demarcated and partially indistinct in the area adjacent to the left lobe of the liver. The mass itself was in contact with lesser curvature of the stomach, IVC, and caudate and lateral segments of the liver (Figs. 1A, 1B).

UGI showed that the extrinsic mass effect on lesser curvature was without mucosal irregularity, thus suggesting that the lesion might have been submucosal or extragastric lesion. Abdominal US demonstrated a well-marginated hypoechoic mass between the stomach and the left lobe of the liver. Endoscopic US performed to evaluate the origin showed partial obliteration of the muscularis propria of the antrum of the stomach, suggesting a gastric origin rather than a hepatic mass. Selective hepatic angiography, however, showed a mass in the left lobe; peripheral tumor vessels were present, and tumor staining via the left hepatic artery and central avascular area was noted (Fig 1C).

Neither abdominal CT, US, UGI or colon study revealed a mass at the other possible sites of origin, including the alimentary and genitourinary tract.

Surgery revealed that the mass originated from the inferior surface of the lateral segment of the liver and adhered to the stomach, IVC, diaphragm, and omentum. Lateral segmentectomy was performed.

The dumbbell-shaped mass, which consisted of bundles of spindle cells in a myxoid background, and was surrounded by fibrotic tissue, was partially attached to the liver and growth was mainly exophytic (Fig. 1D). Histologic examination showed that the central low density area corresponded to myxoid degeneration. The normal liver capsule continued to that of the outer surface of the mass, and there was parenchymal infiltration at the interface between the liver parenchyme and mass; secondary involvement by the continuous lesion primarily located in the stomach, gall bladder, and ligamentum teres was thus rule out.
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Fig. 1. A. Contrast-enhanced CT scan shows a lobulated low density mass projecting into porta hepatis, which has parenchymal beaking (arrows) with the left lobe of the liver, and also indistinct border (open arrows).
B. On CT scan below than Fig. 1a, the mass is abutting the lesser curvature of the stomach (arrows), and seen in extrahepatic location. CHA (common hepatic artery, open arrow) is displaced by the mass.
C. Selective hepatic arteriography shows a large hypervascular mass, which has central necrotic area and dislocate the stretched left hepatic artery (open arrows) laterally. There are contrast filling in the inferior aspect of the mass suggesting tumor vessel or tumor staining (arrows).
D. Cut gross specimen shows a lobulated mass with peripheral solid portion (arrows) and central myxoid area (not shown). The remnant left lobe of the liver had a triangular shape corresponding to the parenchymal beaking on CT. The capsule of the mass is continuous to the normal capsule of the liver in microscopic examination (not shown).

Discussion

Leiomyosarcoma of the liver may arise within the liver (primary hepatic leiomyosarcoma) or from the ligamentum teres (ligamentum teres leiomyosarcoma) (4). Diagnosis of a smooth muscle tumor arising in the liver requires that certain criteria be met. The tumor should not originate from an adjacent structure, nor represent a single metastasis from another primary origin. The most frequent primary sites, including the alimentary tract (stomach, small bowel, colon), genitourinary tract (uterus, bladder, prostate, kidney), and the retroperitoneum (inferior vena cava), should be evaluated (2—6). Grossly, primary leiomyosarcoma of the liver usually presents as a single large mass, of firm consistency.

Primary hepatic leiomyosarcoma should be differentiated from that of the ligamentum teres; the location of the latter is unique, the prognosis is better, and it is encapsulated and clearly demarcated from the liver (4), which is mainly compressed by the mass in its
ligamentum teres rather than by infiltration(7). To ascertain that the tumor did not originate from an adjacent structure, a diagnosis of primary leiomyosarcoma of the liver thus requires a careful search.

The common signs and symptoms are hepatomegaly, an abdominal or right upper quadrant mass, abdominal distention, and weight loss(3, 4).

The literature contains few descriptions of the CT findings of leiomyosarcoma of the liver(2). Most primary hepatic leiomyosarcomas are located in liver parenchyma(3, 4), and to our knowledge, no description of exophytic growth of this tumor, as in our case, is to be found in the radiologic literature(3-5). The tumor has described as a large well-delineated mass with a predominantly peripherally enhanced wall or a mainly cystic appearance. Findings depend on the tumor’s pathologic presentation; as in our case, central low density corresponds to predominantly central necrosis, hemorrhage or amorphous gelatinous tissue (2).

In our case, the main portion of the mass projected into the porta hepatis, and on CT scans was found to show mainly a mass effect, with displacement of surrounding organs rather than invasion. Selective hepatic arteriography of leiomyosarcoma of the IVC may show a mass supplied by the left hepatic artery(8, 9). Because parasitic tumor supply is possible, the extrahepatic mass may still involve the left lobe.

Leiomyosarcoma of the liver is a slow growing tumor, and the survival period ranges from several months to years from initial diagnosis(3, 4). Excision of the bulk of the primary tumor is the treatment of choice. In the previous literature, consideration of the possibility of primary hepatic leiomyosarcoma was seen as important; in such cases, because of aggressive surgery, the prognosis was good(1, 3, 4).

We have described an exophytic hepatic leiomyosarcoma which was shown by CT scanning to have an indistinct border with the parenchyme of the liver, and parenchymal beaking that suggested primary hepatic mass. It was differentiated by imaging modalities which included abdominal CT and visceral angiography.

References
1999년도 춘계 전공의 연수교육 안내

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