Primary Liposarcoma of the Liver
A Case Report and Review of the Literature

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- Liposarcoma is a rare mesenchymal malignant tumor, which usually originates in the retroperitoneum and the extremities. Seven cases of primary liposarcoma of the liver have been previously reported. We present the eighth case, which occurred in an adult female patient. Primary liposarcoma of the liver, although extremely rare, must be considered in the differential diagnosis of a hepatic mass that develops in a noncirrhotic liver, especially in patients who are potential candidates for orthotopic liver transplantation. Liposarcoma is an absolute contraindication for liver transplantation.

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REPORT OF A CASE

A 54-year-old African American woman presented 1 year before admission with abdominal distention, pain, nausea, vomiting, and a 10-lb weight loss. Her only risk factor for liver disease was ingestion of 150 g of alcohol per day, which she discontinued when the abdominal symptoms began. She did not seek medical attention until 3 months before the admission, when her symptoms worsened. On examination, she was not icteric, and there were no stigmata of chronic liver disease. Careful examination of the limbs and buttocks did not reveal lymphadenopathy or soft tissue masses. The abdomen was distended, firm, and tender. A large mass was palpable in the epigastrium and right upper quadrant. There was no ascites, edema, or splenomegaly. Complete blood cell count and standard serum liver test values, including albumin, prothrombin time, and α-fetoprotein, were all within the reference range. A computed tomographic scan of the abdomen revealed a 16-cm solid mass that involved the left lobe and a 3-cm lesion in the right lobe of the liver. An ultrasound-guided liver biopsy failed to provide sufficient diagnostic material. The patient was subsequently transferred to Cedars-Sinai Medical Center for further evaluation and possible liver resection.

A second computed tomographic scan showed that the left lobe mass had increased to 24 cm, with extensive hemorrhage. At that point, exploratory laparotomy and hepatic resection were considered. Laparotomy performed for tissue diagnosis revealed an extensive liver tumor with no obvious extrahepatic extension or distant metastases. The tumor was deemed to be unresectable, and a wedge liver biopsy specimen was obtained. The postoperative course was complicated by continuous bleeding and eventual cardiac arrest and death.

PATHOLOGIC FINDINGS

Permission for autopsy, limited to the abdomen, was obtained. External examination of the body did not demonstrate any palpable masses in the extremities or in the buttocks. The liver weighed 2400 g and the spleen 250 g. The capsule of the superior portion of the right and left liver lobes was infiltrated by an 27 × 15 × 15-cm tumor. Cut section of the tumor revealed a yellow-tan, fleshy, partially lobulated mass that showed scattered, irregular areas of hemorrhage, alternating with areas of necrosis. Several small 2- to 3-cm satellite tumor nodules were present in both lobes of the liver and in the omentum. Surrounding nonneoplastic liver parenchyma was grossly unremarkable and noncirrhotic.

HISTOPATHOLOGIC EXAMINATION, IMMUNOHISTOCHEMISTRY, AND ELECTRON MICROSCOPY

The tumor exhibited a plexiform vascular pattern and myxoid stroma. Lipoblasts were seen in varying stages of differentiation (Figure 1, A). Abundant myxoid stroma was present between tumor cells. The cytoplasm of the lipoblasts was for the most part granular, with nuclei indented by fat vacuoles. Some tumor cells were spindled, and others showed bizarre cell nuclei with occasional giant cell formation (Figure 1, B). Antibody to vimentin (mesenchymal cell marker) was demonstrable in many tumor cells and anti-α-smooth muscle actin (smooth muscle marker) was focally reactive. Immunostains for S100 (neuroepithelial marker), keratin, HMB-45 (marker for melanoma), and CD34 (endothelial cell marker) were nonreactive. Electron microscopy showed typical lipoblasts with antineuronal formation (Figure 1, B). Antibody to vimentin (mesenchymal cell marker) was demonstrable in many tumor cells and anti-α-smooth muscle actin (smooth muscle marker) was focally reactive.

COMMENT

Virchow, in 1857, first described a malignant tumor of fatty tissue arising in the lower extremity.1 Since then, liposarcomas have been identified in a number of tissue and organs, but only rarely in the liver.2,3 The most common sites are the deep soft tissue of the trunk and retroperitoneum and the subcutaneous fat of the upper and
lower extremities. Liposarcomas account for 15% of all sarcomas and are most prevalent in the fifth decade. The prognosis of liposarcoma is dismal without treatment. The 5-year survival rate of patients who have undergone curative resection or radiation therapy for liposarcoma is approximately 50%. Metastatic spread of soft tissue liposarcomas is relatively common, but the liver is involved in only 10% of cases. Metastases are usually found in the brain, pleura, thyroid gland, pancreas, and spinal cord. There have been many attempts to classify liposarcomas, all reflecting a combination of 2 basic histological aspects of the tumor: (1) the stage of the differentiation of lipoblasts, based on relative amounts of lipid in the cells and myxoid material in the extracellular spaces, and (2) the overall degree of cellularity and cellular pleomorphism. Five major histological categories of liposarcomas are recognized: (1) myxoid, (2) round cell, (3) well-differentiated, (4) dedifferentiated, and (5) pleomorphic.

Four cases of primary liposarcoma of the liver have been previously reported in adults. Wolloch et al in 1973 reviewed 16 cases of malignant liver tumors, including one patient with myxoid liposarcoma who had undergone right hepatic lobectomy with only 46 days postoperative survival. Kim et al described a 14 × 10 × 5-cm liposarcoma of the right lobe of the liver. The patient subsequently underwent a hepatic resection and remained tumor free for 10 months.

Primary liposarcomas of the liver have also been described in children. Soares et al described a case of a liposarcoma of the hepatic hilum that presented as obstructive jaundice in a 28-month-old infant. Wright et al described a 3-year-old boy who presented with abdominal pain, later proved to be due to liposarcoma in the hilum. Chen et al evaluated primary malignant tumors in the pediatric population. They described one child with liposarcoma, but sex, age, and exact location within the liver were not described. To our knowledge, there are no re-

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ports of hepatic liposarcoma in principal reference sources of rare hepatic tumors.\textsuperscript{13–15}

We report a case of primary liposarcoma of the liver in an adult, perhaps the largest such tumor yet reported (Table). Although primary liposarcoma of the liver is exceedingly rare, this tumor should be considered in the differential diagnosis, especially in those patients who are potential candidates for liver resection or orthotopic liver transplantation.

References