Hepatic Angiosarcoma and Liver Transplant: A Report of 2 Cases With Diagnostic Difficulties

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Abstract

Angiosarcoma is a rare primary malignant mesenchymal tumor of the liver. The prognosis of hepatic angiosarcoma is poor with an average life expectancy of 6 months after diagnosis. Diagnosing hepatic angiosarcoma is challenging because of nondiagnostic liver biopsy or spurious history and radiologic presentation. We report 2 cases with hepatic angiosarcoma which were diagnosed histopathologically in the native liver after liver transplant. One of 2 patients was lost to follow-up, and another patient died of relapsing hepatic angiosarcoma 18 months after the liver transplant.

Key words: Hepatic angiosarcoma, Liver transplant, Diagnosis, Liver cirrhosis

Introduction

Angiosarcoma is a rare disease, accounting for only 2% of primary tumors of the liver.¹ The prognosis of hepatic angiosarcoma is poor, with an average life expectancy of 6 months after diagnosis. The reason for the poor prognosis is because of difficulty in diagnosing hepatic angiosarcoma in the early stage, rapid progression of the disease, high recurrence rate, and resistance to traditional chemotherapy and radiotherapy.² Diagnosing hepatic angiosarcoma is challenging because of nondiagnostic liver biopsy or spurious history and radiologic presentation. The use of a liver transplant has been abandoned as treatment for hepatic angiosarcoma because of the high recurrence rate and poor survival posttransplant.³ Partial liver resection to remove tumors radically is still the cornerstone of treatment options.⁴ We report 2 cases of hepatic angiosarcoma that were diagnosed in the native liver after liver transplant.

Case 1

A 38-year-old man with progressive liver failure owing to hepatitis B viral infection was referred to our hospital with hepatosplenomegaly, ascites, and jaundice. The tumor markers—especially the serum alpha-fetoprotein levels of the patient—were within normal limits. The results of an abdominal ultrasound revealed significant hepatomegaly with a homogeneous echo nodular structure, as well as massive ascites and splenomegaly indicating cirrhosis. A computed tomography revealed heterogeneity throughout the liver parenchyma. During the examination, the patient showed a rapidly worsening clinical course owing to decompensated cirrhotic liver disease. Therefore, the patient underwent a living-donor liver transplant. After the heptectomy, the native material revealed diffuse multinodularity and a spongelike appearance with blood-filled cystic spaces. Finally, diffuse angiosarcoma was diagnosed in the explanted liver. The patient was lost to follow-up after being discharged from the hospital, 14 days postoperatively.

Case 2

A 43-year-old man with complaints of abdominal pain and jaundice was referred to our hospital. The abdominal ultrasonographic examination showed a massive liver with multiple nodules throughout the

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parenchyma, as well as massive ascites. The patient underwent ultrasound-guided, Tru-Cut biopsy. Epithelioid hemangioendothelioma was diagnosed on biopsy. The serum levels of tumor markers such as carcinoembryonic antigen and alpha-fetoprotein were within the normal limits. High serum levels of CA 19-9 were noted. Three days after the liver biopsy was performed, the patient’s clinical course worsened and he underwent a liver transplant. Gross examination of the explanted liver showed diffuse nodularity on the liver parenchyma. The final diagnosis of the explanted liver was reported as hepatic angiosarcoma. Distinct from the previous biopsy was vascular invasion and necrosis, as noted in the hepatectomy specimen. Eight months after liver transplant, a recurrence of the hepatic angiosarcoma was noted in the allograft liver by Tru-Cut biopsy. The patient died because of hepatic angiosarcoma 18 months after the liver transplant.

Discussion

Malignant tumors of the liver that stem from mesenchymal origins are rare neoplasms. Hepatic angiosarcoma is the most common (36%) of hepatic mesenchymal tumors, accounting for up to 2% of all primary liver tumors. Approximately 10 to 20 new cases are diagnosed yearly in the United States, and the prevalence varies from 0.14 to 0.25 per million. In an autopsy series that took place in Chicago, 1 hepatic angiosarcoma was noted for every 30 cases of hepatocellular carcinoma.

Hepatic angiosarcoma is a vascular tumor that is classically associated with polyvinyl chloride, thorium dioxide colloid (thorotrast), arsenicals, radium, possible copper exposure, and chronic idiopathic hemochromatosis, although the cause of most primary hepatic sarcomas is unknown. Our patients had no experience with these risk factors of angiosarcoma, as far as we know.

There is no sex predilection, and unlike the more common hepatocellular carcinoma, primary liver sarcoma is not associated with liver cirrhosis or a history of hepatitis. Contrary to these findings, 40% of patients have been reported as having had hepatic fibrosis or cirrhosis in autopsy studies; thus, the nature of the association between chronic liver disease and hepatic angiosarcoma is a dilemma. One of our 2 cases also showed a cirrhotic background with hepatic angiosarcoma. Further studies are required to discover the cause of hepatic angiosarcoma in 60% of the cases that have no definite epidemiologic association.

Treatment of primary sarcoma of the liver can be delayed for months owing to obscure abdominal symptoms and equivocal radiologic imaging findings. Patients with primary liver sarcomas that present clinically and radiographically as a liver abscess or as an infected echinococcal cyst have been described. The clinical presentation of hepatic angiosarcoma is nonspecific and includes abdominal pain, weakness, and weight loss with hepatomegaly, ascites, and jaundice. Liver function tests are usually abnormal, but there is no specific test for hepatic angiosarcoma. The occurrence of thrombocytopenia and disseminated intravascular coagulation is characteristic and may be related to local consumption of clotting factors and formed blood elements in the tumor. Therefore, the risk of intra-abdominal bleeding is high.

The appearance of the primary sarcoma of the liver on computed tomography was nonspecific. Hepatic angiosarcoma appeared as a hypodense mass on an unenhanced computed tomography scan, with hyperdense areas representing fresh hemorrhage. A contrast-enhanced computed tomography scan showed avid enhancement owing to the vascular nature of the tumor, with cystic areas due to hemorrhage or necrosis. Selective hepatic arteriogram and open liver biopsy are the basis of diagnostic evaluation. Percutaneous liver biopsy should be avoided because of high hemorrhagic risk and the tendency of diagnostic difficulties.

Orthotopic liver transplant has been abandoned as a treatment for hepatic angiosarcoma because the recurrence rate is 64%. Metastatic disease is found in up to 40% of patients on presentation. Hepatic resection is rarely feasible, but should be considered if the disease is limited and the remainder of the liver is functioning normally. The prognosis of patients with this malignancy is poor, with a median survival of 6 months from diagnosis; even after treatment, only 3% of the patients live longer than 2 years.

In our hospital, there were 408 liver transplant cases between January 1990 and December 2012, and two of the patients with hepatic angiosarcoma underwent transplant. One of the 2 patients was lost to follow-up, and another patient died of relapsing hepatic angiosarcoma 18 months after the liver transplant.
Primary hepatic angiosarcoma is a rare disease with a poor life expectancy. Liver transplant offers the only potential chance of extending survival in the patients with end-stage liver failure due to massive hepatic angiosarcoma. On the other hand, the effectiveness of a liver transplant on these patients remains controversial.

References