Carcinosarcoma of the Liver: A Case Report

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ABSTRACT

Primary hepatic carcinosarcoma is a rare malignant hepatic tumor containing both carcinomatous and sarcomatous elements. A 40-year-old man referred to our liver transplant team because of hepatic cirrhosis was on the waiting list, having undergone all liver tests, ultrasonography, and with normal serum alpha fetoprotein markers every 6 months to search for a tumor. He underwent a liver transplantation without complication. The pathologic findings of the original liver indicated carcinosarcoma. We have reviewed the literature on this subject.

CARCINOSARCOMA of the liver is a rare tumor containing an intimate mixture of carcinomatous and sarcomatous elements that may occur in any organ. Primary hepatic carcinosarcoma is rare, with fewer than 11 adequately documented cases reported, but none transplanted. We present a case of carcinosarcoma of the liver undergoing transplantation, as well as a review of the literature.

CASE REPORT

A 40-year-old man was referred because of hepatic cirrhosis after failure of hepatitis C treatment. Because he displayed ascites and gastrointestinal variceal bleeding, he was on the waiting list for liver transplantation. He went to the outpatient ward frequently for all liver tests, tumor serum alphafetoprotein marker, and ultrasonography, which were always normal.

In September 2004 he underwent a liver transplant; after 11 postoperative days he was discharged from the hospital. The macroscopic pathologic findings of the explanted liver showed five tumors (segments III, IV, V, VIII). The cut surface revealed a solid, grayish-white, and relatively well-circumscribed tumor mass, which was 2.6 × 2.0 cm and 0.4 × 0.4 cm, respectively.

Immunohistochemically, they were positive for vimentin, epithelial membrane antigen, cytokeratin AE1 and AE3 as well as negative for S-100 protein, factor VIII, CEA polyclonal, CAM 5,2; CK and CK20. These immunohistochemical findings associated with the histological findings provided a diagnosis of carcinosarcoma.

The patient showed a good evolution with normal hepatic enzymes and hematology tests until 3 months postoperatively when he experienced the onset of abdominal and right lumbar pain that intensified each day. The patient showed an intestinal partial occlusion; computerized tomography showed peritoneal carcinomatosis with many hepatic tumors. Quickly the patient displayed renal, pulmonary, and cardiac failure, succumbing at 5 months after the liver transplantation.

DISCUSSION

Carcinosarcoma is a rare tumor with carcinomatous and sarcomatous elements. It is more frequent in the ovary, uterus, and urinary bladder. Primary hepatic carcinosarcoma is extremely rare and aggressive. Graig et al[10] recommend that the term "carcinosarcoma" be reserved for tumors with both carcinomatous and nonsimple cell sarcomatous elements.[1] In contrast, the World Health Organization defined carcinosarcomas as tumors containing both carcinomatous (either hepatocellular or cholangiocellular) and sarcomatous elements, including malignant mixed tumors in this category.

Controversy has surrounded the histogenesis of carcinosarcomas of the liver. Fayyazi et al have speculated that the tumor originates from a single tutipotential stem cell that differentiates in separate epithelial and mesenchymal directions.[2] In contrast, Kubosawa et al proposed that elements of hepatocellular carcinoma were transformed into sarcomatous cells.[3]

In recent years, increasing evidence has supported the theory that carcinosarcoma is monoclonal in origin. The carcinomatous element is the driving force; the sarcomatous component is derived from the carcinoma or from a stem cell that undergoes divergent differentiation.

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A literature review shows there were few reported cases of this type of primary hepatic tumor. Most cases have been observed in men of mean age 60 years (range 46–84). The clinical characteristics of these tumors include abdominal pain, fever, and jaundice. These symptoms can be followed by liver cirrhosis with negative or low serum alphafetoprotein levels. The tumor size ranges from 4 to 23 cm, averaging 12.1 cm. As sarcomatous cell proliferation is rapid, the associated prognosis is generally poor. Hepatocellular carcinoma rarely disseminates to the peritoneal cavity; however, the sarcomatoid variant shows a greater incidence of both dissemination and metastases as in our patient.

Usually early tumor stages are associated with improved survival, and surgical resection is a treatment option. However, the majority of patients present with advanced disease; even those patients with early-stage disease show high recurrence rates following surgical resection.

We have reported a case of a man with hepatic cirrhosis after hepatitis C treatment failure who was on the waiting list for liver transplantation according to established criteria. During the follow-up he never had clinical symptoms nor did he have any examinations suggesting a hepatic tumor, so the histopathology of the explanted liver surprised the authors.

Some cases of primary liver carcinosarcoma have been reported in the literature, but none of them has been transplanted.[2–4,6–8] The aggressive characteristics and poor prognosis observed in our patient, make the authors recommend not to transplant this type of liver tumor.[5,9]

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