Primary carcinosarcoma of the liver: an unusual case with clearly separated epithelial and mesenchymal components

Dear Editor,

Primary carcinosarcoma of the liver is a rare and extremely aggressive neoplasm that usually causes the death of the patient in less than a year after it is diagnosed (1). According to WHO, it is a malignant tumor containing a mixture of carcinomatous elements (hepatocellular carcinoma or cholangiocarcinoma) and sarcomatous elements (osteosarcoma, chondrosarcoma, rhabdomyosarcoma, or undifferentiated spindle cell sarcoma) (2). Only a few cases have been reported thus far and in all of them the epithelial and mesenchymal components were intermingled (3,4).

The case we report is most unusual because, unlike all previously reported hepatic carcinosarcomas, its two components—epithelial and mesenchymal—are clearly separated.

Case report

A 70-year-old man with chronic liver disease due to hepatitis B diagnosed 20 years earlier and refractory to the treatment, presented dark urine, pale stools, moderate fatigue and jaundice of the mucoses and skin. Analytical data demonstrated an abnormal liver test.

The abdominal ecography and the CAT scan (Fig. 1A) revealed a heterogeneous, 5 cm intrahepatic mass located in left lobe of the liver (segment IVa-b) with peripheral enhancement during arterial phase and a hypodense central area suggestive of necrosis. The mass compressed the left hepatic duct without affecting the right hepatic via. Other radiological findings included thrombosis of left portal vein and dilated intrahepatic bile ducts. The rest of the parenchyma was characterized by steatosis and two simple cysts in the right lobe.

All clinical and radiological features were suggestive of hepatocarcinoma. A segmentectomy IVb was performed, which included resection of the extrahepatic bile duct, resection of both common hepatic ducts due to the infiltration of their bifurcation, a tumoral thrombectomy of left portal vein, and a cholangiojejunostomy. After overcoming a hydropic decompensation, the patient was discharged from the hospital.

The Pathology Department received a segmentectomy IVb about 9 x 6 cm in size and weighing 210 g. Gross examination revealed a soft, whitish confluent multinodular tumor (6 x 5 cm) close to the deep surgical margin (Fig. 1B). An extensive sampling of the tumor and the surrounding parenchyma was performed.

Microscopically, one third of the neoplasm was made of a conventional, well-differentiated hepatocarcinoma with trabecular pattern, abundant polygonal cells with large eosinophilic cytoplasm and pleomorphic nuclei, and scarce cells with clear cytoplasm (Fig. 2A). Carcinomatous cells were immunopositive for HepPar-1 (hepatocyte) and CK7, but immunonegative for CK20, carcinoembryonic antigen (CEA), and alpha-fetoprotein. The rest of the tumor consisted of a sarcomatous component with pleomorphic-fusocellular, storiform pattern (Fig. 2B), made of abundant spindle cells with elongated nucleus and fine chromatin, and scarce pleomorphic cells. Sarcomatous cells were immunopositive for vimentin (Fig. 2C), alpha-1-antitrypsin, CD68, actin and epithelial membrane antigen (EMA) (the latter three showed focal staining pattern), but immunonegative for all other epithelial markers (Fig. 2D), as well as desmin, CD117, Cd34, S-100, HepPar-1 (hepatocyte), glypican-3 and alpha-fetoprotein.

The two components of this tumor (carcinomatous and sarcoma-
The neoplasm presented also vascular infiltration and extensive necrosis. The non-tumoral parenchyma showed chronic hepatitis with moderate fibrosis.

Even though the neoplasm was successfully resected, the patient died four months after surgery as a consequence of portal vein tumor thrombi, liver insufficiency and sepsis.

Discussion

Hepatic carcinosarcoma must be differentiated from sarcomatoid carcinoma, which is a poorly differentiated carcinoma with fusiform cells. Unlike the carcinosarcoma, sarcomatoid carcinoma contains exclusively cells of epithelial origin (6).

The pathogenesis of carcinosarcoma is unclear. It has been suggested that the neoplastic cells of conventional hepatocarcinoma are capable of transforming into multipotent immature cells, which in turn redifferentiate into sarcomatous components (3,6), an idea supported by the mixture of the two components seen in all cases reported thus far. In our case, in contrast, the two components were clearly separated—adjacent, but not intermingled—, which increases the difficulty of the diagnoses and makes it advisable to perform an extensive and detailed sampling of the tumor.

In summary, primary hepatic carcinosarcoma is clinically similar to conventional hepatic carcinoma; however, the presence of the sarcomatous component increases the aggressiveness and invading capacity of the tumor, thus worsening its prognosis.

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References