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Intrahepatic sarcomatoid cholangiocarcinoma: a rare cholangiocarcinoma subtype

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Informed consent: The patient described in this letter and his family provided informed consent for the publication of this letter and for the use of his medical data.

Conflict of interest: The authors declare no conflicts of interest.

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Dear Editor,

A 71-year-old man with a history of cholecystectomy presented to our hospital with icteric sclera, darker urine, and recurrent fever. Excluding the mild icteric sclera and upper-middle abdominal tenderness, the patient's physical examination was unremarkable. Laboratory data revealed the following: neutrophilic percentage, 76.8% (normal, 60%–70%); CA19-9, 29.15 U/mL (normal, 0–27 U/mL); direct bilirubin, 14.3 µmol/L (normal, 1.71–7.0 µmol/L); total bilirubin, 27.1 µmol/L (normal, 1.71–17.1 µmol/L); aspartate aminotransferase,78 U/L (normal, 0–40 U/L); alkaline phosphatase,225 U/L (normal, 25–150 U/L); glutamyl transpeptidase,138 U/L (normal, 0–50 U/L); and negative results for hepatitis B- and C-associated antigens and antibodies, alpha-fetoprotein, carcinoembryonic antigen, and alanine

transaminase.After administration of an iodine-contrast agent, contrast-enhanced abdominal computed tomography (CT) revealed a soft-tissue density mass shadow in the left lobe of the liver with circular heterogeneous enhancement. Magnetic resonance cholangiopancreatography revealed a heterogeneous hypointense mass on T1-weighted imaging, but hyperintense on T2-weighted imaging and hyperintense on diffusion-weighted imaging in the left lobe of the liver. Moreover, the left intrahepatic bile duct was dilated with calculi (Fig. 1A–1C).The surgical specimen was hard, fishlike, and without capsule. Histological examination revealed that the cells were spindle shaped with coarse chromatin, irregular nuclear contours, pleomorphism, and prominent nucleoli and were arranged in nests. Immunohistochemical stains were positive for CKpan, CAM5.2, CK19, CK7, EMA, CK8/18, and vimentin. These findings confirmed the diagnosis of intrahepatic sarcomatoid cholangiocarcinoma (ISCC; Fig. 1E, F).Tumor recurrence and metastasis occurred one month after resection(Fig. 1D). Consequently, the patient received chemoradiotherapy. However, four months after the operation, the patient succumbed to cachexia.

Discussion

ISCC is a rare subtype of cholangiocarcinoma. It is histopathologically composed of different amounts of cholangiocarcinoma and sarcomatoid components (1). Primary sarcomatoid hepatocellular carcinoma degeneration is common after anticancer chemotherapy or hepatic artery embolization; however, intrahepatic sarcomatoid cholangiocarcinoma degeneration is relatively rare (2). The pathogenesis of ISCC remains unclear, and repeated cholangiolithiasis and infection may play a role in sarcomatoid cholangiocarcinoma (3). ISCC is more common among older individuals. Patients typically present with abdominal pain and weight loss. ISCC is highly invasive, prone to recurrence, and has a poor prognosis (4). Although, patient survival can be improved with radical surgical resection (5). Gemcitabine and cisplatin combined with chemotherapy are known to be beneficial for ISCC treatment (5).

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Figure 1

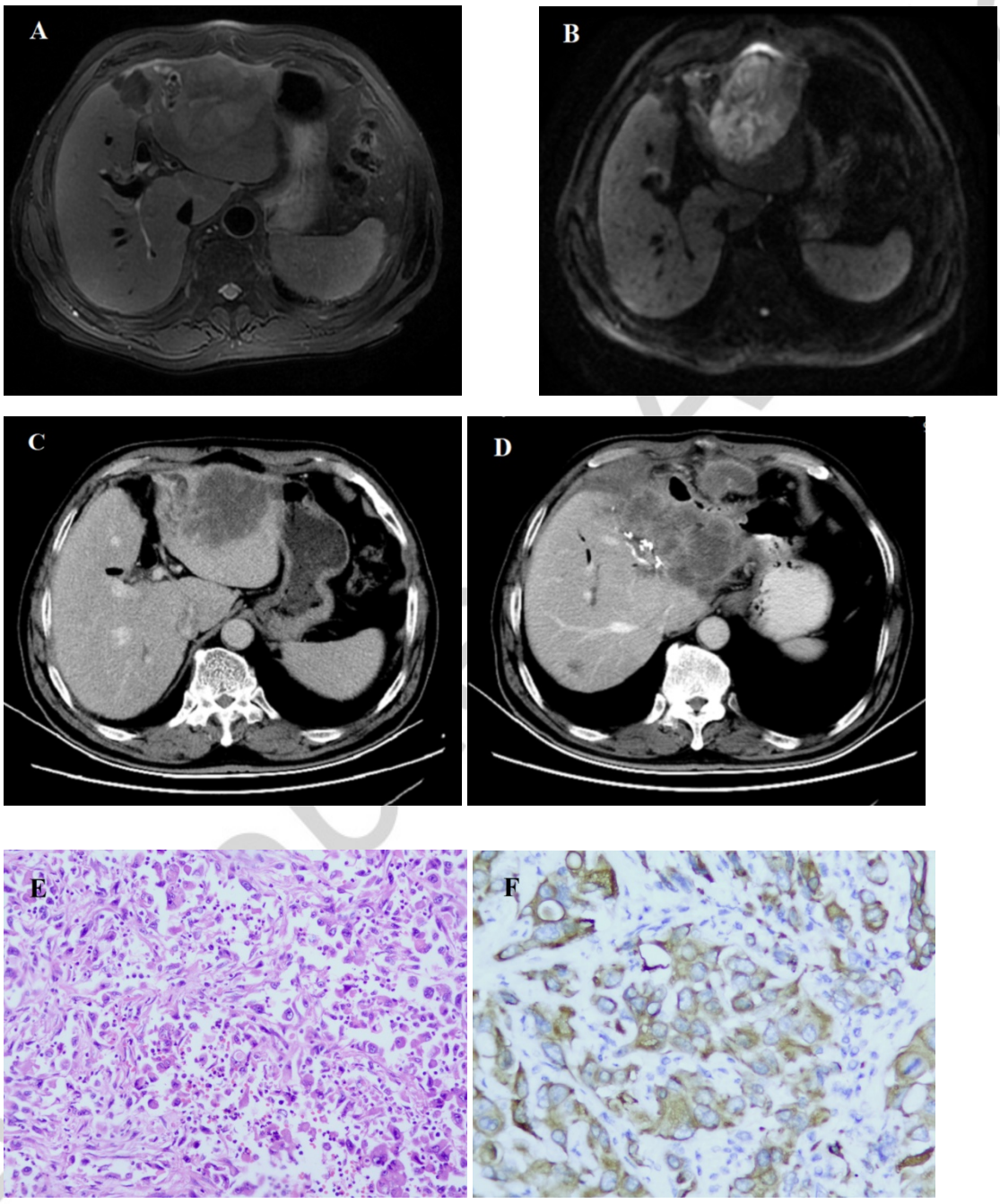

Fig. 1. *A-B. Magnetic resonance imaging showing heterogeneous hyperintense lesions on T2-weighted images and hyperintense on diffusion-weighted images in the left lobe of the liver. C. Contrast-enhanced abdominal computed tomography (CT)*

revealed a soft-tissue density mass shadow in the left lobe of the liver with circular heterogeneous enhancement. D. The tumor recurred and metastasized after surgery. Upper abdominal CT revealing multiple low-density mass shadows with unclear boundaries and ring enhancement. Local lesions involved the gastric wall. E. Sarcomatous areas composed of spindle cells arranged in sheets or bundles and interspersed with pleomorphic giant cells and necrotic tissue (hematoxylin and eosin, ×200). F. Immunohistochemical study of carcinomatous component revealed positive staining for CAM5.2.