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Carcinosarcoma of the ampulla of Vater

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Dear Editor,

Carcinosarcomas, also known as spindle cell carcinomas or sarcomatoid carcinomas (1), are very rare tumors in the gastrointestinal tract. They have an aggressive clinical course with frequent metastasis and those located at the ampulla of Vater location are extremely uncommon.

Case report

The case was a 64-year-old male with a two-month history of intermittent, painless jaundice and weight loss of 8 kg. The abdominal examination was normal. Magnetic resonance cholangiopancreatography (MRCP) showed an abrupt termination of the distal bile duct and a hypodense soft tissue lesion at the ampulla of Vater, dilatation of the intra and extrahepatic bile ducts (20 mm) and dilatation of the Wirsung duct.

Laboratory test showed 15.2 mg/dl total bilirubin, 12.4 mg/dl direct bilirubin, 60 IU/l AST, 65 IU/l ALT, 456 IU/l alkaline phosphatase and 174 IU/l GGT. Endoscopic retrograde cholangiopancreatography (ERCP) revealed a proliferative, prominent, purple, bleeding, friable and ulcerated tumor at the ampulla of Vater that measured 25 x 25 mm in size (Fig. 1A). Histological analysis revealed a poorly differentiated tubular adenocarcinoma and sarcomatoid

tissue composed of spindle tumor cells with large, bizarre nuclei. The border between the carcinomatous and sarcomatoid elements was relatively well demarcated and the immunohistochemical staining were positive for vimentin and keratin (Fig. 1B).

The majority of cases of digestive carcinosarcomas have sarcomatous elements composed of anaplastic spindle cells. These tumors are called sarcomatoid carcinoma, spindle cell carcinoma, pseudosarcoma or malignant mixed tumor (1,2). Survival ranges from five to 36 months (3). These tumors of the gastrointestinal tract are most commonly located in the esophagus. In the biliary tract, the gallbladder is the most common site, with < 30 cases described. Keratin was positive in our case, which indicates that this tumor has epithelial characteristics. Some studies (4) have suggested that carcinosarcomas in various organs arise from epithelial cells with sarcomatous differentiation. This is also a possibility in our case.

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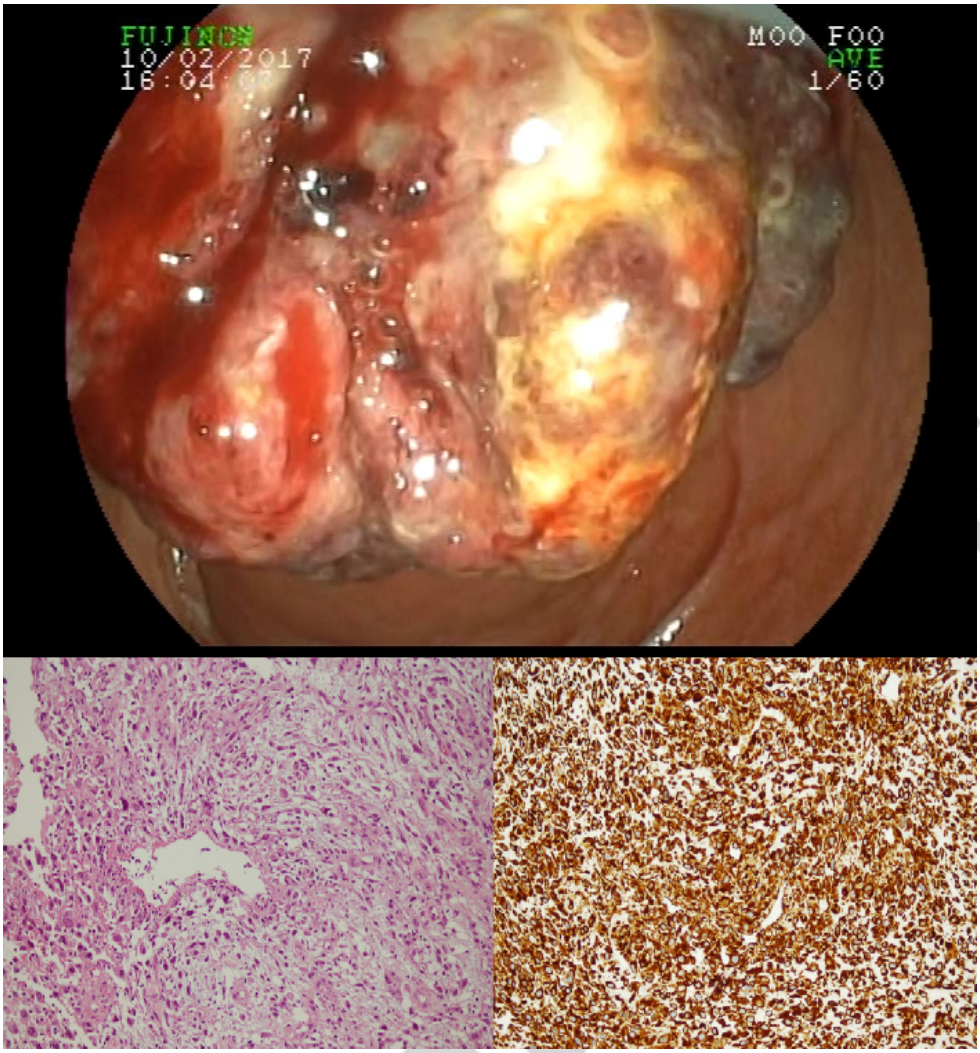

Fig. 1. A. ERCP showed a proliferative, prominent, purple, bleeding, friable and ulcerated tumor at the ampulla of Vater, measuring approximately 25 x 25 mm in size. B. A poorly differentiated tubular adenocarcinoma and sarcomatoid tissue composed of spindle tumor cells with large, bizarre nuclei. The border between the carcinomatous and sarcomatoid elements was relatively well demarcated. Immunohistochemical staining was positive for vimentin.