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Sarcomatoid Carcinoma of the Distal Common Bile Duct
- A Case Report -

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Sarcomatoid carcinoma of the common bile duct (CBD) is an extremely rare malignant neoplasm, which is characterized by the presence of carcinomatous and sarcomatous components. We report a case of sarcomatoid carcinoma arising in the distal CBD. The patient was a 68-year-old woman who presented with abdominal pain. The computed tomography and endoscopic ultrasonography revealed a polypoid mass in the distal CBD. The resected specimen showed a polypoid mass with a narrow stalk in the distal CBD which was confined to the mucosa. The cut surface revealed a gray-whitish solid mass with focal hemorrhage and necrosis. Microscopically, the tumor was composed of carcinomatous and sarcomatous components without any heterologous elements. The sarcomatous area predominently consisted of pleomorphic spindle cells. The carcinomatous component was an adenocarcinoma. On immunohistochemistry, cytokeratin was coexpressed in the carcinomatous and sarcomatous components but vimentin was expressed exclusively in the sarcomatous component. The patient has been doing well for one year postoperatively.

Key Words : Sarcomatoid Carcinoma; Common bile duct

Sarcomatoid carcinoma (carcinosarcoma) is a very rare lesion in the hepatobiliary pancreatic system.[1] These tumors have been reported in diverse sites including the ampulla of Vater, gallbladder, pancreas and liver.[2-5] There are only very rare reports of sarcomatoid carcinoma arising in the extrahepatic bile ducts including the common hepatic duct and common bile duct. To the best of our knowledge, only two cases of sarcomatoid carcinoma of the extrahepatic bile duct have been reported in the world literature.[1,6] Here we report an additional case of sarcomatoid carcinoma arising in the distal CBD and review the relevant literatures.

CASE REPORT

On June 2004, a 68-year-old woman visited our hospital with symptoms of abdominal pain, anorexia, and weight loss that she had experienced for 2 months. She had a history of percutaneous transluminal coronary angioplasty last year that was due to angina. Her personal and family history were unremarkable. Physical examination revealed epigastric pain, a nontender palpable mass in the right upper quadrant of the abdomen and icteric sclera. Routine laboratory tests including a complete blood count, electrolytes, liver function tests and routine urine analysis were unremarkable except for the slightly increased total and direct bilirubin (1.4 mg/dL and 0.9 mg/dL, respectively). The serum levels of alpha-fetoprotein and carcinoembryonic antigen (CEA) were within the normal reference range, but the carbohydrate antigen 19-9 was increased (48.6 U/mL).

The abdominal computed tomography and endoscopic ultrasonography revealed an intraluminal protruding polypoid mass located in the distal CBD with marked dilatation of the proximal CBD and the common hepatic and intrahepatic ducts (Fig. 1). No lymphadenopathy was noted and the pancreatic duct was

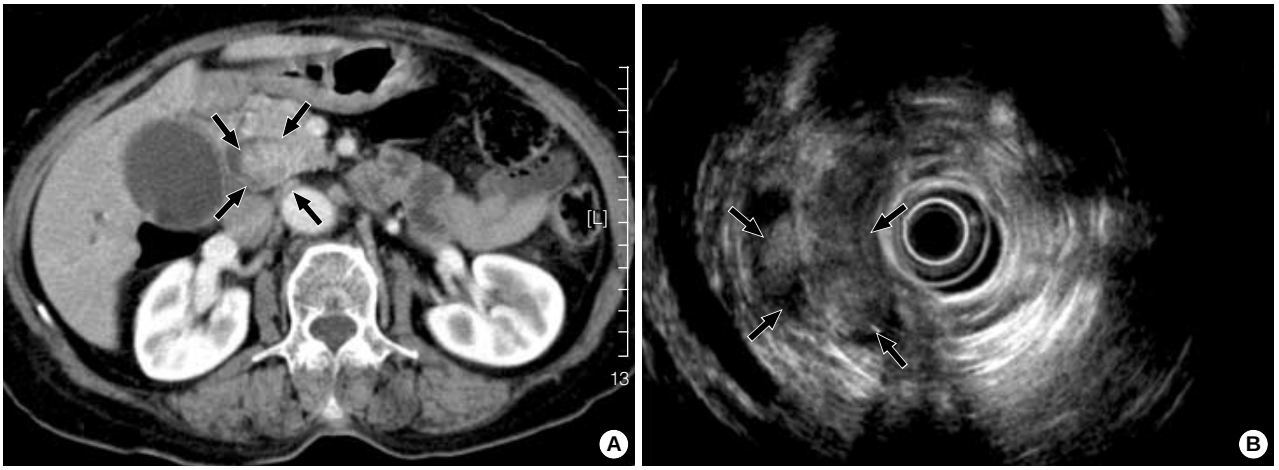

Fig. 1. Abdominal computed tomography (A) and endoscopic ultrasonography (B) show an intraluminal polypoid mass (arrows) in the dis-

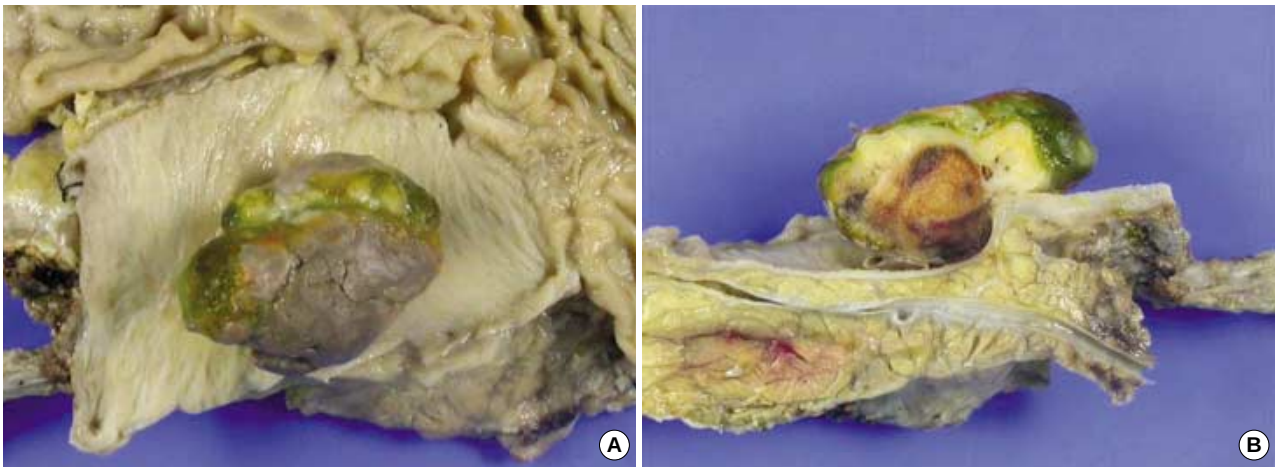

Fig. 2. A polypoid mass with a narrow stalk is present in the distal common bile duct (A). The cut surface shows a grayish white solid mass with focal hemorrhage and necrosis, which is confined to the mucosa (B).

unremarkable. Whipple's operation was performed under the impression of distal CBD cancer. The resected specimen showed a movable polypoid mass that measured $3.5 \times 2.5 \times 2.5$ cm in dimension. On the section along the CBD, the cut surface revealed a grayish white, solid and rubbery to firm mass with areas of hemorrhage and necrosis (Fig. 2). The mass was confined to the mucosa. Microscopically, the tumor showed a biphasic pattern of predominantly sarcomatous and focally carcinomatous components (Fig. 3). The sarcomatous area was highly cellular and it consisted of mostly spindle-shaped cells, some neutrophils and lymphocytes intermixed among the tumor cells. The tumor cells showed predominantly spindle-shaped nuclei with severe pleomorphism; mitotic figures were occasionally seen. The carcinomatous component was seen in a focal area and it was composed of irregular glands and tubules of adenocarcinoma. Any heterologous sarcomatous elements were not identified. On the immunohistochemical staining, cytokeratin was strongly positive in the carcinomatous area and weakly positive in the sarcomatous area. CEA was strongly positive in the carcinomatous area and negative in the sarcomatous area. Vimentin was diffusely positive exclusively in the sarcomatous area (Fig. 4). The tumor cells showed negative immunoreactivity for smooth muscle actin (SMA), desmin, myoglobin, S-100 protein and c-kit.

DISCUSSION

Sarcomatoid carcinoma of the CBD is an extremely rare malignant neoplasm in the hepatobiliary system and it has been found in the liver, pancreas, gallbladder, intrahepatic and extrahepatic

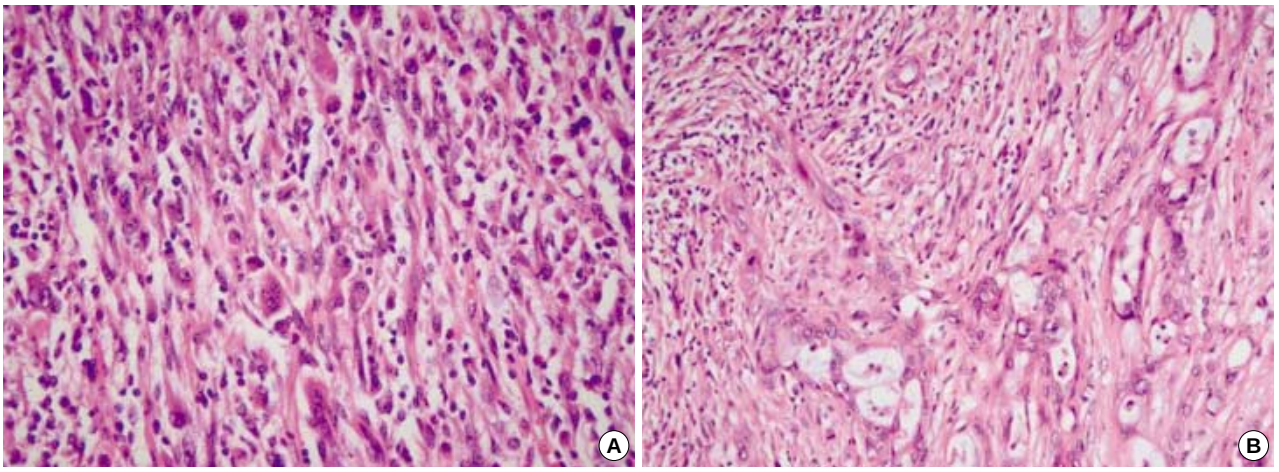

Fig. 3. Microscopically, the tumor is composed of sarcomatous (A) and carcinomatous (B) components. (H&E, ×200).

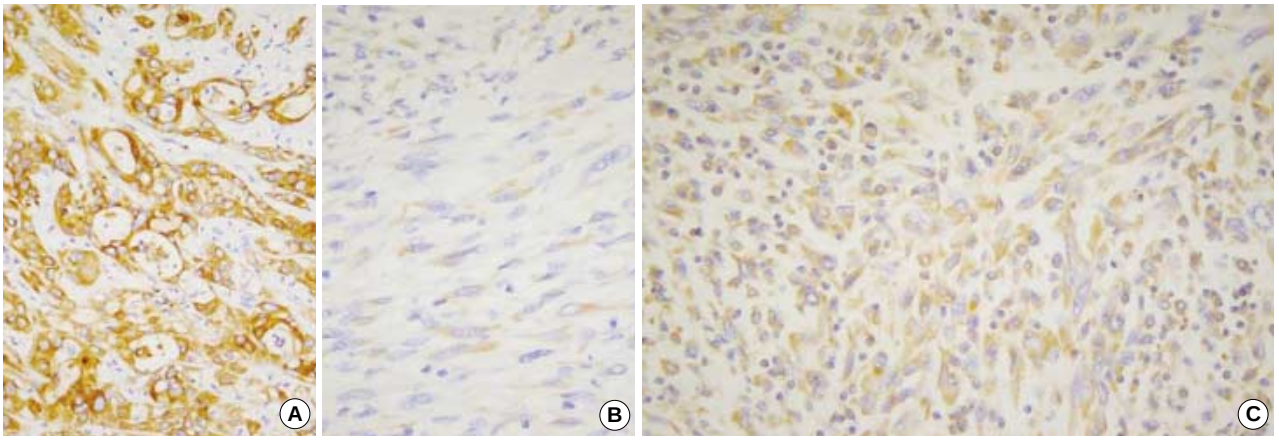

Fig. 4. On immunohistochemical staining, cytokeratin is strongly expressed in carcinomatous component (A) and weakly positive in sarcomatous component (B). Vimentin is expressed exclusively in sarcomatous component (C). (PAP, ×200).

Table 1. Clinicopathological features of three cases sarcomatoid carcinoma arising in the common bile duct

Source of the cases	Sex/ Age	Site	Tumor size (cm)	Growth pattern	Microscopic findings
Yoon *et al.*[1]	M/78	mid	4×3×3	infiltrative	biphasic pattern with focal smooth muscle differentiation
Sodergren *et al.*[6]	F/64	proximal	2×1.2×1.2	polypoid	biphasic pattern with focal chondroid and smooth muscle differentiation
Present case	F/68	distal	3.5×2.5×2.5	polypoid	biphasic pattern without heterolo differentiation

bile duct. The gallbladder is the most frequently affected site of sarcomatoid carcinoma in the biliary system and there have been about thirty cases previously reported.[7,8] This tumor is characterized by the presence of its unique biphasic histologic nature, the intermixed carcinomatous and sarcomatous components and sometimes there are zones of transition.[1] Due to the rarity of sarcomatoid carcinoma in the CBD, the clinicopathologic features of this tumor are uncertain. To the best of our knowledge, only two cases of sarcomatoid carcinoma arising in the CBD have been previously reported in the english literature.[1,6] The clinicopathologic features of the reported cases and the present case are summarized in Table 1.

The patients were a 78-year-old man and a 64-year-old female. The tumor origin was at the mid and proximal CBD. The tumor sizes of the two cases were 4 cm and 2 cm, respectively, of the greatest dimension. The gross appearance of the tumors was infiltrative or polypoid and the histological features were characterized by a biphasic pattern of carcinomatous and sarcomatous components. Yoon *et al.*[1] reported a smooth muscle differentiation

in the focal sarcomatoid area with a positive result for α-SMA. Sodergren *et al*.[6] reported that the mesenchymal areas included foci resembling smooth muscle and there were other foci that had a chondroid appearance. Our case was a 68-year-old woman with a polypoid growing mass in the distal CBD. The present case also displayed the typical biphasic pattern of sarcomatous and carcinomatous components merging into one another. However, any heterologous sarcomatous components were not found.

The underlying causes or predisposing factors of sarcomatoid carcinoma in the CBD are not so clear. Three major theories have been suggested.[2] These include the possibilities of a metaplastic change of carcinomatous cells to sarcomatous cells, the existence of a single multipotential uncommitted reserve cell capable of divergent differentiation, and the collision tumor proposal that suggests the separate, but concurrent proliferation of malignant epithelium and mesenchyme in the same organ. In our case, the sarcomatoid cells were regarded as metaplastic carcinoma cells or carcinoma cells with sarcomatoid differentiation because of their expression pattern of cytokeratin and vimentin. Cytokeratin was coexpressed both in the carcinomatous and sarcomatous components. The merging that was seen between the carcinomatous and sarcomatous components also supports this theory. Therefore we prefer the term of "sarcomatoid carcinoma" rather than "carcinosarcoma".

Based on the current data on sarcomatoid carcinoma of the gallbladder, the behavior and prognosis of this tumor seems to resemble carcinoma of the gallbladder, which is considered dismal at best.[6] Yoon *et al*.[1] suggested a poor prognosis because their case revealed diffuse thickening and narrowing of the CBD due to diffuse tumor infiltration, direct extension to the pancreas and regional lymph node metastasis. Sodergren *et al*.[6] suggested a better prognosis because of the polypoid nature of the tumor. The polypoid tumor may have led to the early bile duct obstruction, the symptoms of jaundice and the diagnosis of the lesion. This may have resulted in the resection, achieving complete tumor clearance before spreading as in the present case. Our case has been uneventful for one year postoperatively.

Although sarcomatoid carcinoma in the CBD is extremely rare, it should be considered in the differential diagnosis of a polypoid tumor of the CBD.

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