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# A case of spindle cell sarcomatous change of hepatic ducts manifesting as obstructive jaundice

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**Abstract:** Spindle cell carcinoma is a rare tumor commonly occurring in the upper aerodigestive tract. We report a 62-year-old male with spindle cell sarcomatous change located at the hepatic hilum, resulting in obstructive jaundice. The patient died after an extended resective operation. The rare disease and its histogenesis is discussed.

**Key words:** spindle cell sarcomatous change, hepatic ducts, cholangiocarcinoma, sarcoma

## Introduction

Spindle cell carcinoma is usually a polypoid tumor occurring in the upper aerodigestive tract,[1–4] skin,[5] breast,[6] the uterine cervix,[7] and lung.[8–10] Adenosquamous carcinoma with spindle cell features[11] and another small cell carcinoma with squamous and adenocarcinomatous components[12] have recently been reported in the gallbladder. However, to our knowledge, spindle cell sarcomatous change originating from the cholangiolar cells of common hepatic ducts has not been described. We report a case manifesting as obstructive jaundice in a 62-year-old male.

## Case report

A 62-year-old Taiwanese male was admitted to Kaohsiumg Yuan's General Hospital, Taiwan, on October 16, 1993 due to jaundice and pain over the right upper quadrant of 2 days, duration. There was no past history of liver and biliary disease. The patient had no history of either cigarette smoking or alcohol intake. Two days prior to admission, he had begun to suffer from dull pain in the right upper quadrant, without radiating pain. Yellowish discoloration of the skin and tea-colored urine were also noted. There was neither clay-colored stool nor body weight loss. No fever or chills were experienced.

On physical examination, he was moderately developed and well-nourished. The conjunctiva was not pale but the sclera was icteric. The chest and heart were negative for pathologic findings. The abdomen was slightly distended with hepatomegaly about 4 cm below the right costal margin. The spleen was not enlarged. The gallbladder was not palpable. No shifting dullness was demonstrated. There was neither pitting edema nor cyanosis of the extremities. Laboratory examination revealed elevation of serum transaminase (AST 145 U/ml, normal <35 U/ml; ALT 251 U/ml, normal <30 U/ml), and bilirubin (total bilirubin 9.5 mg/dl, direct bilirubin 6.4 mg/dl). Serum alkaline phosphatase was 1131 U/ml (normal <272 U/ml). Albumin was 4.3 gm/dl and globulin was 3.6 gm/dl. Results of renal function tests, hemogram, electrolytes, coagulation profiles, and other biochemical tests were within normal limits. Serum alpha-fetoprotein (AFP) was 0.8 ng/ml (normal <20 ng/ml). Serum carcinoembryonic antigen (CEA) and carbohydrate antigen (CA19-9) were 8.3 ng/ml (normal <5 ng/ml) and 482 μg/ml (normal <37 μg/ml), respectively.

Abdominal ultrasonography of the liver disclosed a small atrophic left lobe and an enlarged right lobe. The intrahepatic ducts in the right lobe (especially at segments S6 and S7) were dilated. Hepatic hilar lesion was suspected. Computed tomography showed similar features with a suspicious infiltrative lesion at the porta hepatis (Fig. 1). Percutaneous transhepatic cholangiography revealed irregular dilatation of the right intrahepatic ducts, ending at the porta hepatis. The left intrahepatic ducts were not shown (Fig. 2).

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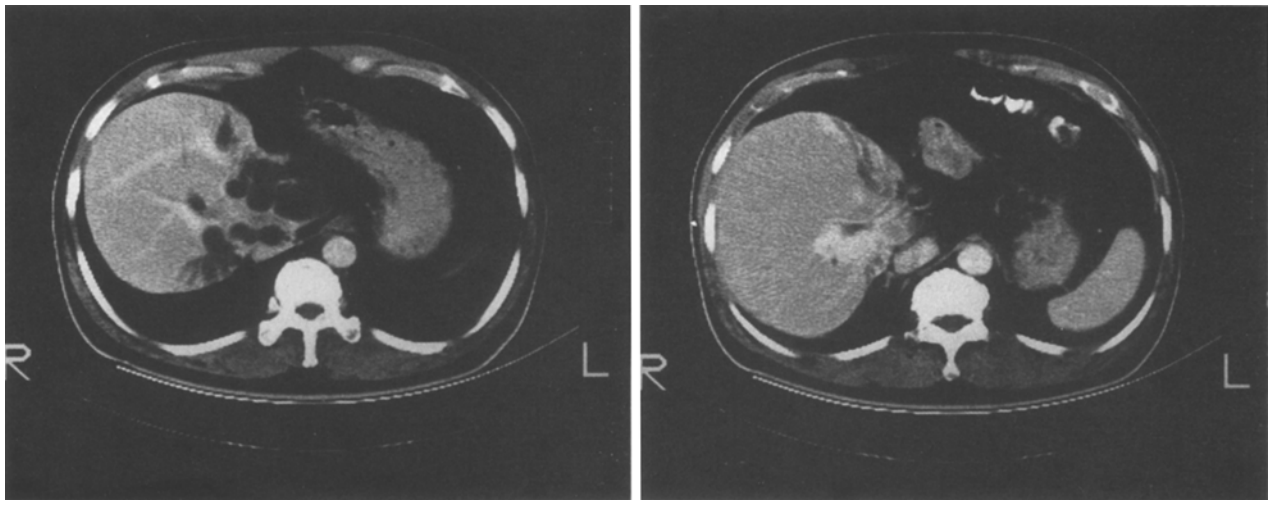


a b

**Fig. 1a,b.** Contrast enhanced computed tomography shows the atrophied left lobe and remarkable dilatation of the intrahepatic ducts. An infiltrative lesion at the porta hepatis is suspected. **a**, Dilatation of intrahepatic ducts; **b**, suspicious lesion at porta hepatis after enhancement

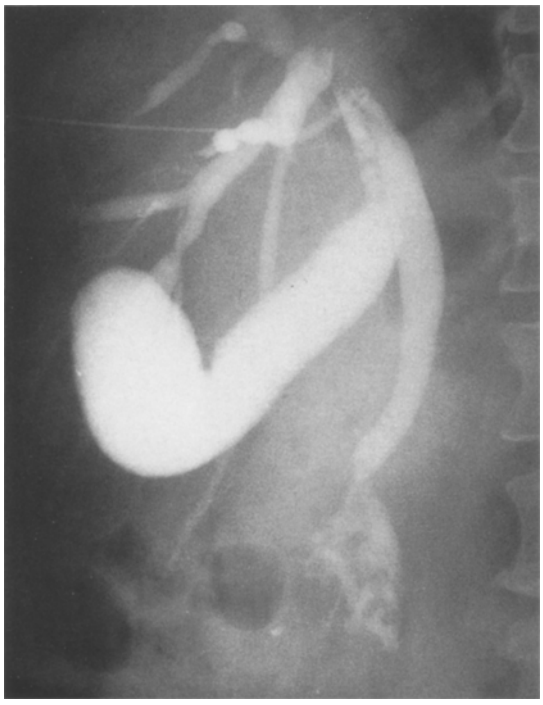

**Fig. 2.** Percutaneous transhepatic cholangiography discloses irregular dilation of the right intrahepatic ducts ending at the porta hepatis. The left intrahepatic ducts are not shown

The patient underwent operation under a diagnosis of hilar cholangiocarcinoma on November 13, 1993. During operation, a whitish nodule, measuring 3 × 3 cm, was noted at the hepatic hilum, obstructing the left and right intrahepatic ducts. Extended left hepatic lobectomey, cholecystectomy, choledochectomy, right hepato-jejunostomy, and jejuno-jejunostomy were performed. The left hepatic lobe and part of the adjacent right lobe were resected. On gross examination, the liver was green in color and the hepatic ducts were markedly dilated. A nodular tumor occluding the common hepatic duct, at the junction of the left and right intrahepatic ducts, was noted. The tumor measured 3.5 × 2.0 × 1.6 cm and was grayish-white, with an elastic consistency. Marked cholestasis was noted in the hepatic parenchyma (Fig. 3). There was neither gross cirrhotic change nor a satellite tumor nodule within the liver. Intrahepatic stones were not detected. Sections from the tumor involving the liver exhibited a malignant neoplasm with spindle cell as well as glandular and epithelial elements. Histopathology revealed tumor nodules of various sizes separated by loose collageneous tissue. Within the nodules, tumor cells were arranged compactly, with a glandular formation, surrounded by elongated spindle cells (Fig. 4). These gland-lining cells tended to lose their polarity. Foci of vascular permeation by tumor embolus formation and areas of inspissated bile pigment with cellular tumor sheet were found. An immunohistochemical study with CEA (Fig. 5a), epithelial membrane antigen (Fig. 5b), and low molecular weight keratin (Fig. 5c) demonstrated epithelial cells lining glandular elements and duct-like luminal surfaces, as well as a few spindle cells focally. There was no immunoreactivity for alpha-fetoprotein, desmin, or s-100-protein. Vimentin staining was weakly positive for the spindle elements. Many nerves displayed considerable reactive hypertrophy. The adjacent hepatic parenchyma disclosed marked cholestasis without evidence of cirrhotic change, as shown by Masson staining.

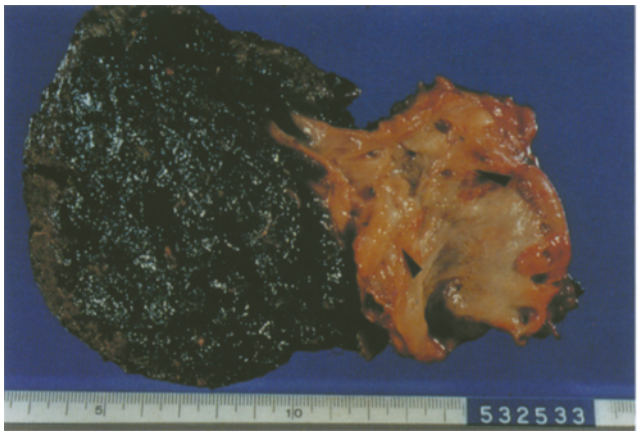


**Fig. 3.** Gross pathology of the resected tumor (*right*) in the hepatic duct. Marked cholestasis is noted in the adjacent liver parenchyma (*left*). *Arrows* indicate the tumor lesion

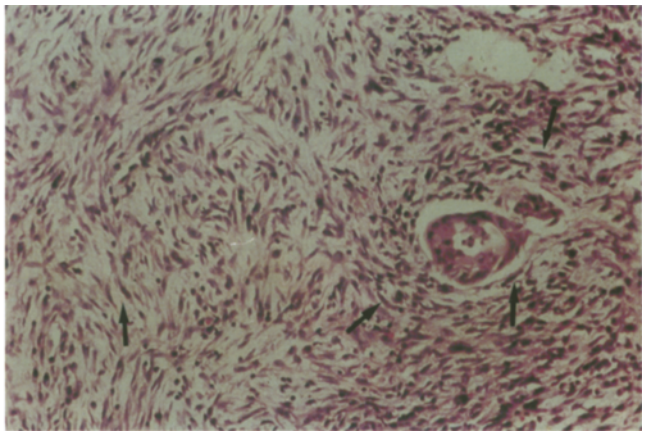

**Fig. 4.** Histopathology shows tumor cells arranged compactly, with a glandular formation, surrounded by elongated spindle cells. H&E, × 134

Unfortunately, the patient experienced a stormy postoperative course, with hepatic insufficiency, which resulted in death 10 days after operation. Autopsy was not permitted.

## Discussion

Obstructive jaundice induced by a malignant tumor at the hepatic hilum is usually due to cholangiocarcinoma. Histologically, the majority of these tumors are adenocarcinoma. Fewer than 10% are squamous cell

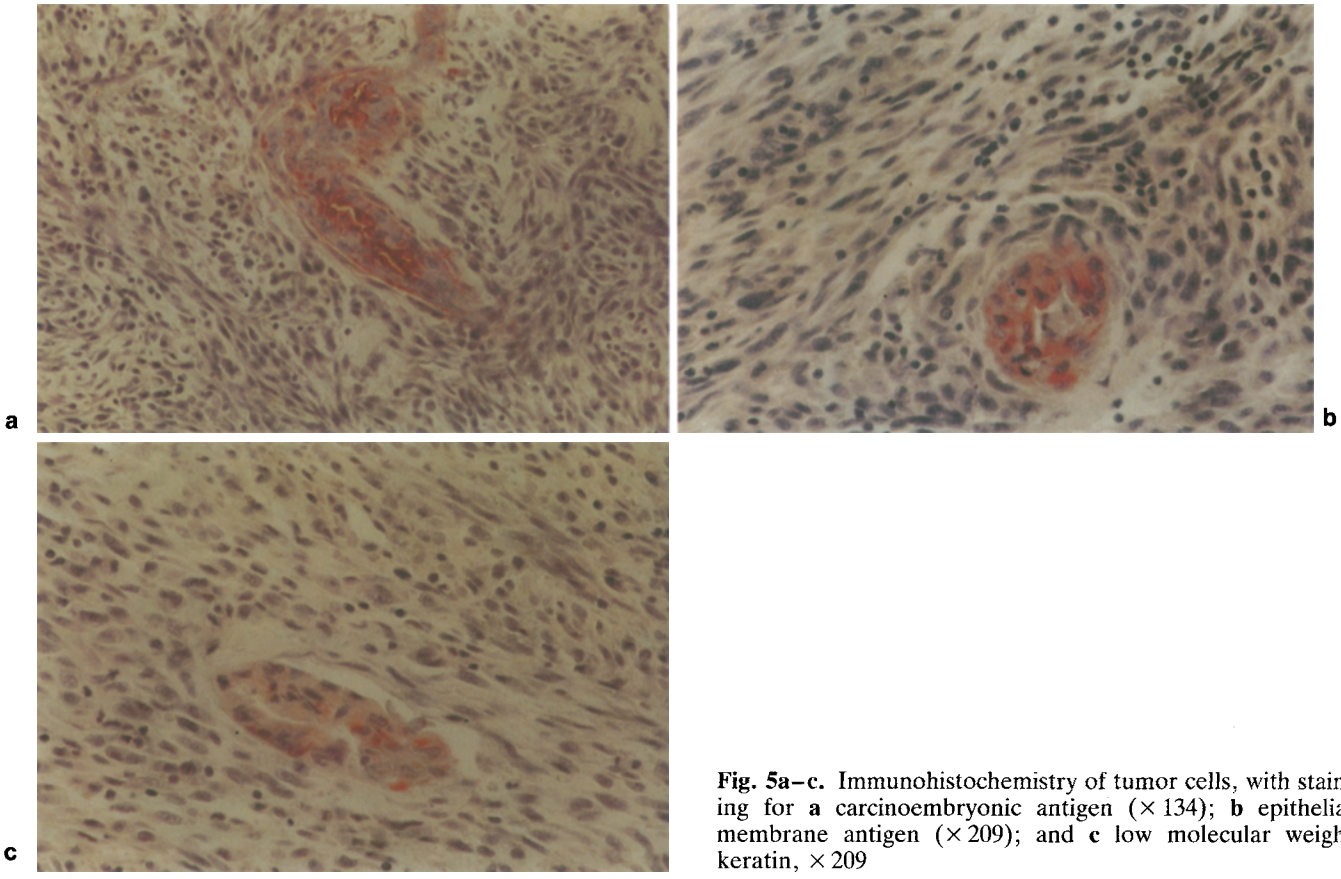


**Fig. 5a–c.** Immunohistochemistry of tumor cells, with staining for **a** carcinoembryonic antigen (× 134); **b** epithelial membrane antigen (× 209); and **c** low molecular weight keratin, × 209

carcinoma.[13] Sarcomatous (or spindle cell) differentiation of squamous cell carcinoma has been recognized in various sites, such as the upper aerodigestive tract,[1–4] skin,[5] lung,[8–10] and urogenital tract.[14] However, to the best of our knowledge, there has been no report concerning spindle cell sarcomatous change of the hepatic ducts.

Clinically, in our patient, the spindle cell sarcomatous change manifested as obstructive jaundice, due to its location in the hepatic ducts. The results of laboratory examination suggesteds only extrahepatic cholestasis. Elevation of serum levels of CEA and CA19-9, in contrast to a normal AFP level, precludes the diagnosis of hepatocellular carcinoma. Diagnostic imaging revealed a suspicious infiltrative lesion at the porta hepatis that could have been hepatocellular carcinoma, invasive carcinoma of the gallbladder, metastatic carcinoma, or stenotic lesions induced by another benign process. After excluding these possibilities by diagnostic imaging, cholangiocarcinoma remained the tentative clinical diagnosis, despite the abundance of spindle cells in the resected tumor histologically.

Considerable controversy exists concerning the histogenesis of spindle cell carcinoma. There are three theories concerning its histogenesis: it represents reactive connective tissue,[15] it is a true sarcoma,[16] and it represents carcinoma with pseudosarcomatous features.[15] The majority of recent reports favor a neoplastic and epithelial origin for the sarcomatous component. Positive immunoreaction with keratin, epithelial membrane antigen, and CEA suggests an epithelial origin, while positive reaction with desmin, actin, myosin, and vimentin implies mesenchymal origins.

However, it is known that renal cell carcinoma[17] and anaplastic carcinoma of the thyroid[18] may show positive vimentin staining. Upton et al.[19] have also reported that vimentin was sometimes expressed in adenocarcinoma, and it has been shown that tumors containing cells positive for vimentin are not always sarcoma.[8] Zarbo et al.[3] reported that even individual cells in sarcomatous areas sometimes co-expressed keratin and vimentin. Therefore, positive vimentin staining in our case was not needed to identify the tumor as sarcoma. We have also experienced poorly differentiated cholangiocarcinoma revealing weakly positive vimentin staining. From the standpoint of morphologic appearance and histochemical reactivity, the neoplasm was consistent with a cholangiolar origin.

Spindle cell sarcomatous change is a rare disease that may occur in hepatic ducts, resulting in obstructive jaundice. This location of the tumor leads to a dismal outcome.

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