

CASE REPORT

Carcinoid Tumor of the Common Bile Duct

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(Received 24 October 1993)

A case of a primary carcinoid tumor of the common bile duct is presented. Diagnostic and therapeutic uncertainties of this extremely rare cause of jaundice are discussed.

KEY WORDS: Carcinoid tumor biliary tract obstructive jaundice
pancreaticoduodenectomy

INTRODUCTION

Argentaffin, carcinoid tumor producing cells, are present in all parts of the gastrointestinal tract including the biliary system¹. The most frequent gastrointestinal primary sites for the development of carcinoid tumors are the small bowel, rectum, appendix and stomach². Carcinoid tumors of the biliary tract, on the other hand, are sporadically reported and arise mostly in the gallbladder^{3,4}, whereas such tumors of the main bile ducts are extremely rare and have been reported only on four occasions⁴⁻⁷.

We present herein another case of a carcinoid tumor of the common bile duct presenting with obstructive jaundice. Diagnostic and therapeutic uncertainties are emphasized.

Case report

A 44 year-old male patient was admitted with resolving obstructive jaundice. Previous ambulatory ultrasonography has demonstrated a mild distention of the intra- and extra- hepatic bile ducts and a dilated gallbladder. This has been followed by an endoscopic

retrograde cholangiography (ERCP), which disclosed a fusiform stricture of the distal common bile duct; it has been dilated and a 10 F stent was inserted. There were no positive findings on physical examination except for residual jaundice. After the jaundice subsided the patient underwent an explorative laparotomy. A 0.5 cm' lesion was found on the anterior surface of the left hepatic lobe and was fully excised; frozen section reported an anaplastic carcinoma. The extra- hepatic bile ducts, gallbladder and pancreas appeared macroscopically normal. The condition was assessed as a 'metastatic carcinoma of presumably biliary or pancreatic origin' and a palliative choledochoduodenostomy was undertaken. Frozen section of the choledochotomy's normally appearing edges revealed an anaplastic carcinoma as well.

The postoperative course was uneventful. Pathological examination of the paraffin sections showed a small (0.5 cm') metastatic carcinoid tumor in the liver. The tumor cells were arranged in groups of small uniform cells without conspicuous atypical changes (Fig. 1). The diagnosis of carcinoid tumor was supported by a positive immunohistochemical stain for chromogranin. The tumor infiltrating the wall of the common bile duct showed the same small uniform groups of cells (Figure 2) and perineural infiltration was seen. This tumor was interpreted as a carcinoid of the bile duct.

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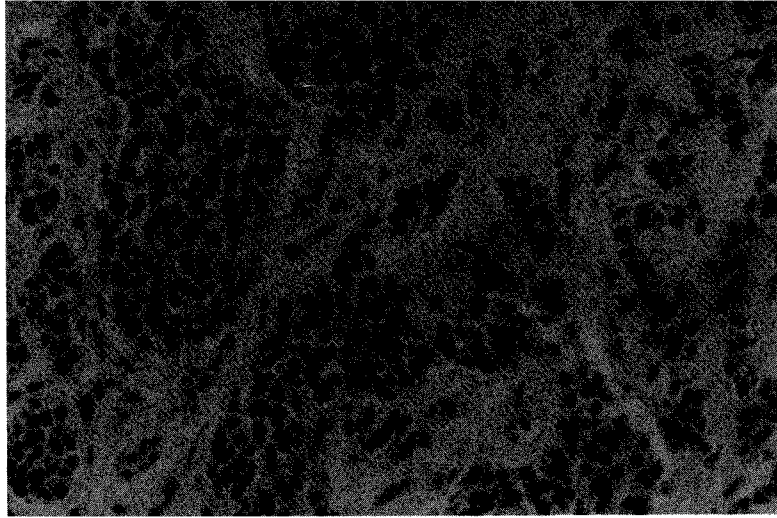


Figure 1 The metastatic carcinoid tumor in the liver (H & E, $\times 300$).

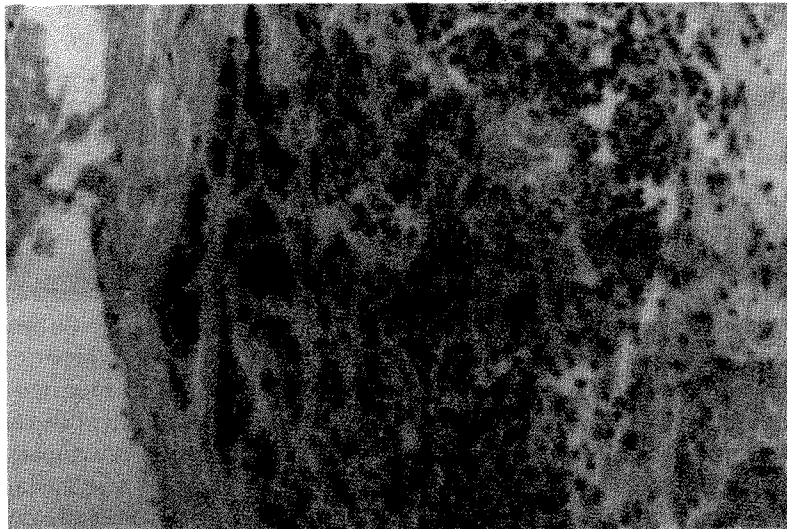


Figure 2 The carcinoid tumor in the wall of the common bile duct (H & E, $\times 300$).

The literature was searched for therapeutic guidelines to no avail. Taking into consideration the minimal hepatic involvement and the patient's young age it was decided to proceed with a pancreaticoduodenectomy in order to eliminate the primary tumor which was suspected still to exist at the site of the choledocoduodenostomy. The latter was excised at a pylorus-preserving Whipple's procedure during which the liver and regional lymph-nodes appeared normal. Careful pathological assessment of the resected specimen failed to find any evidence of residual carcinoid tumor. All excised lymph nodes were free of

tumor. Presently, 18 months after the operation the patient is well.

DISCUSSION

The extreme rarity of carcinoid tumor causing obstructive jaundice makes its pre-operative diagnosis very unlikely. This case of a carcinoid tumor of the common bile duct is the fifth reported in the English literature. Also the four previously described patients presented with obstructive jaundice; one was

diagnosed at autopsy⁴, another at operation⁶ and the details of the remaining two were not mentioned^{5,7}.

In this respect the situation is similar to that occurring with other uncommon benign tumors and pseudotumors or sarcoma of the bile ducts, all conditions clinically mimicking the commonly occurring cholangiocarcinoma⁸. Obviously, routine pre-operative investigations of the jaundiced patient such as ultrasound, CT, and ERCP are not able to pinpoint the correct diagnosis. As reflected in this case and emphasized by others²⁻⁴, the primary carcinoids of the gastrointestinal tract may be of a small diameter and yet metastases may be found. Perhaps, needle aspiration of a mass, if visualized on CT or brushing cytology at ERCP, could achieve pre-operative diagnosis. Also accurate pre- and intra-operative histological diagnosis may be problematic because the differentiation between carcinoid tumors and undifferentiated carcinoma is very difficult (Figures 1 & 2), particularly on frozen section specimen.

This case exemplifies the dangers of "blind" stenting of the obstructed bile duct, without histological diagnosis, in the jaundice patient labeled (following CT and ERCP examinations) as suffering from 'malignant obstruction with metastatic disease'. Such approach may deny definitive treatment in the occasionally curable patient while exposing him to the prolong morbidity of non-operative palliation.

The paucity of published experience with carcinoid tumor of the bile duct makes therapeutic recommendations not more than a learned guess. However, experience with carcinoids elsewhere in the gastrointestinal tract, which suggest the advisability of aggressive resection or debulking may be applied^{9,10}.

Primary tumors should be removed and if deemed unresectable debulking should be attempted. Likewise, solitary liver metastases should be resected. The role of pancreaticoduodenectomy as performed in this patient, remains unproven. Interestingly, no evidence of tumor was found in the specimen of our Whipple's procedure attesting to the minute size of the primary carcinoid causing jaundice in this patient, and the possibility that it was completely destroyed during biopsy and choledoduodenostomy.

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