



Carcinosarcoma of the Choledochal Cyst

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Abstract

We report the case of a 19-year-old young man with choledoch carcinosarcoma secondary to anastomosis of choledochal cyst and jejunum. The patient suffered from the choledochal cyst and received anastomosis of choledochal cyst and jejunum at 4-year-old. The patient was hospitalized because of abdominal pain, fever and obstructive jaundice. The imaging examination showed a huge heterogeneous tumor located in the hilar area with dilatation of intra hepatic bile duct. Resections of choledoch cyst and jejunum loop associated with pancreaticoduodenectomy were performed during the operation. Carcinosarcoma was pathologically diagnosed after the operation. Pelvic metastases were diagnosed 8 months postoperatively and sigmoid colostomy was therefore performed. The patient died 4 months later.

Keywords: Carcinosarcoma; Choledoch cyst; Bile duct

Introduction

Carcinosarcomas are rare malignant and biphasic tumors composed of both carcinomatous and sarcomatous components. They have been reported in multiple organs, such as uterus, lung, bladder and liver. However, only few cases have been reported in the extra hepatic biliary system, especially in the common bile duct [1]. Only seven cases of common bile duct carcinosarcomas have been described in English literature [1-7]. There is no case report of carcinosarcoma secondary to congenital choledoch cyst. Here, we report a case of choledochal cyst carcinosarcoma which could be the first case worldwide.

Case Presentation

A 19-year-old young man was referred after suffering abdominal pain, fever and obstructive jaundice for one month. He had a history of congenital choledochal cyst and received the anastomosis of choledochal cyst and jejunum when he was 4 years old. A solid mass in upper abdomen was palpated reaching 3 cm below the costal margin and 4 cm below the ensisternum. Biochemical tests showed elevation in bilirubin, transaminase and alkaline phosphatase levels (ALT 133 U/L, AST 99 U/L, T-Bil 15.7 mg/dL, D-Bil 11.9 mg/dL, ALP 266 U/L, GGT 505 U/L). An enhanced CT scan showed a huge tumor (12cm*10cm) located in the hilar area with heterogeneous enhancement during the arterial phase (Figure 1a and b). The dilatation of intrahepatic bile ducts was detected by CT scan. Analysis of serum tumor markers revealed an elevated carbohydrate antigen 199 (CA199) level of 1199 U/ml (normal range: < 35 U/ml). However, both alpha fetoprotein and carcinoembryonic antigen levels were within normal ranges. Under a preoperative diagnosis of cholangiocarcinoma of choledochal cyst, the patient underwent an exploratory operation. A huge cystosolid tumor was detected in the hilar area, which reached from the bifurcation of hepatic bile duct to the intrapancreatic bile duct (Figure 1c). Resection of choledochal cyst and jejunum loop associated with pancreaticoduodenectomy was performed during the operation. Macroscopically, a lot of grayish yellow soft tissues were detected in choledochal cyst with the total size of 20×16×10 cm and areas of hemorrhage and necrosis were found in the cut surface. Histologically, the solid part of the tumor was comprised of both moderately-poor differentiated adenocarcinomic and sarcomatous component. The adenocarcinoma cells were arranged in trabecular and tubular patterns intermingling with spindle-shaped cells (Figure 2a). Immunohistochemical staining revealed that the adenocarcinoma cells were positive for cytokeratin while the sarcomatous cells were positive for vimentin (Figure 2b and c). The patient suffered an intra peritoneal abscess after the operation and percutaneous catheter drainage was performed. He was discharged on post-operative day 42 and the serum CA199 level had decreased to normal level. Neither chemotherapy nor radiotherapy was conducted after the operation. Pelvic metastases were diagnosed 8 months postoperatively and sigmoid colostomy was therefore performed. The patient died 4 months later.

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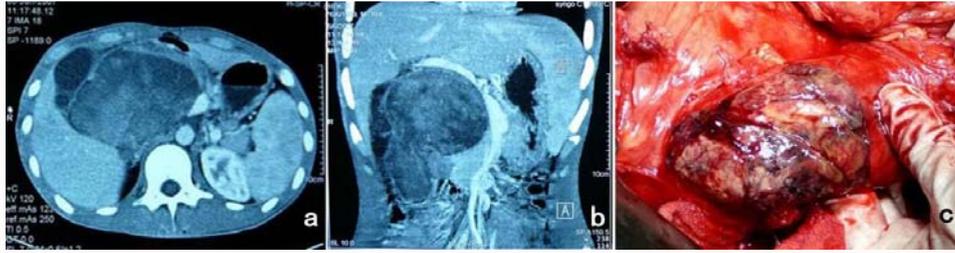


Figure 1: a, b) CT scan showed a huge tumor (12 cm×10 cm) located in the choledoch and loop of jejunum with heterogeneous enhancement during the arterial phase. c) A huge cystosolid tumor was detected in the hilar area, which reached from the bifurcation of hepatic bile duct to the intrapancreatic bile duct.

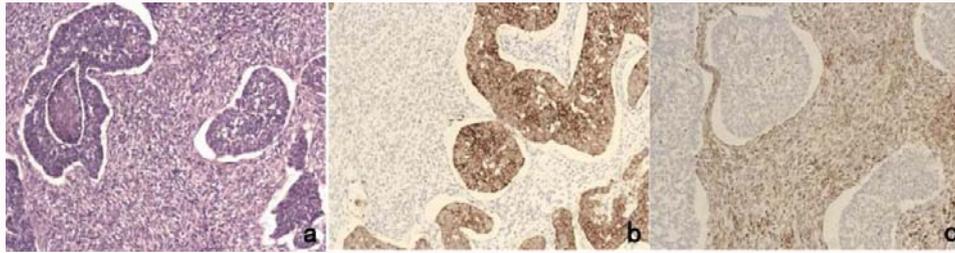


Figure 2: a) Histologically, the solid part of the tumor was comprised of both moderately-poor differentiated adenocarcinoma and sarcomatous component. The adenocarcinoma cells were arranged in trabecular and tubular patterns intermingling with spindle-shaped cells. b) Immunohistochemical staining revealed that the adenocarcinoma cells were positive for cytokeratin c) while the sarcomatous cells were positive for vimentin.

Discussion

Carcinosarcoma is a very rare malignant disease which consists of both carcinomatous and sarcomatous components. Its exact histogenesis has not been clearly elucidated. Among various hypotheses, two major theories have been proposed for the predisposing factors of the origin of carcinosarcoma: the “Multiclonal Theory” which regards a carcinosarcoma as being a collision tumor composed of separate epithelial and mesenchymal origin, and the “Monoclonal Theory” which regards the carcinomatous and sarcomatous components are derived from a single pluripotent stem cell [8,9]. In immunohistochemical staining, most of the carcinomatous components are positive for epithelial markers such as cytokeratins and epithelial membrane antigen, while the sarcomatous components are positive for vimentin and smooth muscle actin. In this case, the adenocarcinoma cells were positive for cytokeratin but negative for vimentin, while the spindle cells were on the contrary.

Carcinosarcoma has been reported in the female genital tract, lung, kidney, bladder and the gastrointestinal tract [10]. But carcinosarcoma itself is a very rare lesion of the hepatobiliary pancreatic system, especially for the common bile duct [11]. To our knowledge, only seven cases carcinosarcoma of common bile duct have been reported [1-7], while no case of choledochal cyst carcinosarcoma has ever been reported. This may be the first case report of common bile duct carcinomasarcoma secondary to congenital choledochal cyst. In the previous reports, most of the cases were elderly female patients. However, our case was a young male patient and congenital choledochal cyst may be the reason for the pathogenesis. According to the previous reports, this is the youngest patient with carcinosarcoma of common bile duct.

Due to the rarity and lack of specific features of imaging, accurate preoperative diagnosis is difficult for carcinosarcoma. Most of the previously reported cases were diagnosed as cholangiocarcinoma preoperatively. Postoperatively pathological examination would confirm the diagnosis of carcinosarcoma. However, even if accurate

diagnosis was made preoperatively, the patients may receive the same surgical procedure. The only possibly curative treatment for carcinosarcoma is surgery. The prognosis of patients with carcinosarcoma of common bile duct remains uncertain. But according to the case reports, many patients died of metastasis within one year after the operation. At present, there are no reports of effective chemotherapy or radiotherapy for carcinosarcoma, irrespective of the affected organ. Further investigations using new agents may be helpful to improve the surgical outcome for patients with carcinosarcoma.

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