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Case Report

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Synchronous Distal Cholangiocarcinoma and Pre-Operative Unacknowledged Pancreatic Ductal Adenocarcinoma: An Extremely Rare Finding!

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Abstract

Background: the presence of a synchronous biliary tract and pancreatic cancers is extremely rare and has been described in few cases only. Herein, a case of a man who underwent pylorus-preserving pancreaticoduodenectomy for a distal cholangiocarcinoma is reported. However, on top of the cholangiocarcinoma a diagnosis of synchronous pancreatic ductal adenocarcinoma was made at pathological examination of the operative specimen.

Case: an 82-years-old man with history of multiple abdominal surgery, presented at the emergency room with jaundice. Abdominal ultrasound detected a bile duct dilatation, and contrast-enhanced abdominal-CT showed a suspicious 21mm enhanced lesion of the pancreatic head. No secondary lesions were detected. Endoscopic US described a hypoechoic lesion of 26,9x10,5mm of the distal bile duct suspicious for distal cholangiocarcinoma and an early chronic pancreatitis. No other lesions were found after a MRCP and restaging CT, and a pylorus-preserving pancreaticoduodenectomy was performed. The post-operative course was regular. However, on the pancreatic margin of the operative specimen a second neoplasia was found. This prompted the diagnosis of extrahepatic cholangiocarcinoma with synchronous pancreatic ductal adenocarcinoma, with 4/23 metastatic lymphnodes from adenocarcinoma-NOS. After referral to a specialized Center, considering comorbidities and personal medical history as well as the previous oncological treatment, no adjuvant chemotherapy was decided, and only a clinical-instrumental follow-up was established. After 18 months, the patient is still in good conditions with no signs of recurrence.

Conclusion: we report an extremely rare case of distal cholangiocarcinoma in an elderly patient with a synchronous pancreatic ductal adenocarcinoma with a surprisingly clinical surgical outcome.

Keywords: Cholangiocarcinoma; Pancreatic Ductal Adenocarcinoma; Synchronous; Pancreatic Surgery; Pylorus-Preserving Pancreaticoduodenectomy.

Introduction

Cholangiocarcinomas are rare tumours representing 3% of all gastrointestinal malignancies [1], arising in the 40% of the case in the distal portion of the biliary tract [2]. Exocrine pancreatic cancers are rare neoplasia and the worldwide incidence is about 4,9/100000 , representing overallthe 2,6% of all malignant neoplasias [3]. A primary malignant biliary tract neoplasia with synchronous primary pancreatic cancer is also an extremely rare condition described in few cases [4-7]. Herein, we report a case of an 82-years-old man with a diagnosis of distal cholangiocarcinoma who underwent pylorus-preserving pancreaticoduodenectomy followed by a diagnosis of synchronous pancreatic ductal adenocarcinoma at pathological examination.

Case Presentation

An 82-years-old man with Parkinson disease presented to Emergency Room of our center for weakness and jaundice. Previously, he had surgery for colorectal cancer with Hartmann's resection followed by adjuvant treatment, and also a partial cholecystectomy for chronic cholecystitis complicated by a cholecystoduodenal fistulas. Laboratory exams showed high circulating level of bilirubin (10.04mg/dL/9.25mg/dL) and Cancer antigen 19.9 (Ca19.9) (83.3 UI/mL) (Table 1).

Table 1. Laboratory data at ER admission

Laboratory data		
White blood cells	5.21	x 10^3/µL
Hemoglobin	12.3	g/dL
Platelets	214	x 10^3/µL
INR	1.13	
PCR	7.34	mg/L
Creatinine	0.66	mg/dL
Sodium	142	mMol/L
Potassium	3.60	mMol/L
Aspartate aminotransferase	110	UI/L
Alanine aminotransferase	26	UI/L
Gamma-glutamyl transpeptidase	406	UI/L
Total bilirubin	10.04	mg/dL
Direct bilirubin	9.25	mg/dL
Albumine	38	g/L
CEA	2.1	ng/mL
Ca 19.9	83.3	UI/mL

Intrahepatic and extrahepatic bile duct dilatation was detected at abdominal ultrasound (US) whereas computed tomography (CT) showed a suspicious slightly enhanced lesion of 21 mm of the pancreatic head with bile duct dilatation (26mm) without secondary lesions (Figure 1A-B). Progressive, total and direct bilirubin levels increased (up to 14.65/12.79 mg/dL) and an endoscopic retrograde cholangiopancreatography (ERCP) detected double bile duct stenosis: the first of less of 1 cm in the supra-papillary tract and the second of 12-13 mm localized more proximal in the common bile duct. During the procedure, brush cytology and an endoscopic drainage with plastic stent were performed, obtaining a reduction of bilirubin levels. The cytology examination did not detect any neoplastic cells. An endoscopic US showed a hypoechoic lesion of 26.9 x 10.5 mm of the distal (supra and intrapapillary) bile duct which potentially suggested a primary malignant distal bile tract cancer. The pancreatic parenchyma was slightly lobed mimicking an initial chronic pancreatitis while no other lesions were found. The magnetic resonance cholangiopancreatography (MRCP) confirmed the suspicion of a single distal cholangiocarcinoma (dCCA) (Figure 2A-B). No other abdominal or thoracic lesions and/or metastasis were found.

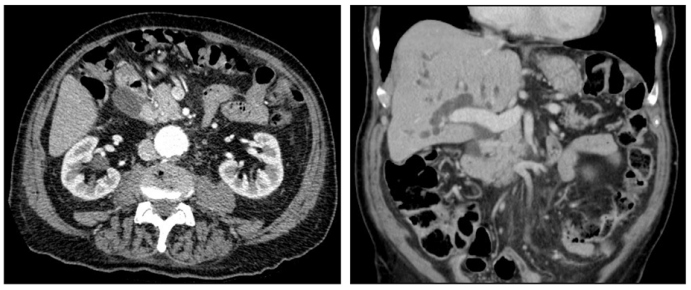

Figure 1. *abdominal computed tomography (CT) showing a suspicious slightly enhanced lesion of 21 mm of the pancreatic head A: axial section; B: coronal section.*

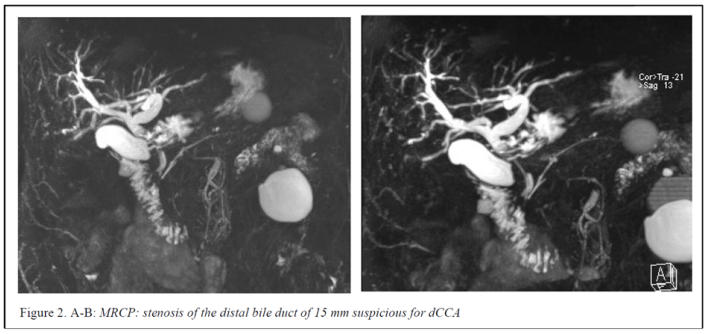

Figure 2. A-B: *MRCP: stenosis of the distal bile duct of 15 mm suspicious for dCCA*

At first, the patient and his relatives rejected a surgical treatment and chose a non-operating management with a clinical and instrumental revaluation after few months. Six months later, the patient was stable and the restaging CT imaging confirmed the single neoplasm of the distal bile duct without other lesions and metastasis. At this time, the laboratory exams showed again high

circulating level of bilirubin and a higher Ca19.9 (Table 2).

This time the patient accepted the surgical procedure and a pylorus-preserving pancreaticoduodenectomy was performed. Surgery lasted 420 minutes and the blood loss was about 500 mL; the pathologic examination performed during surgery on frozen section of the bile duct did not detect any neoplastic cells. Two drainages were placed near the anastomosis and the post-operative course was uneventful: the first drainage was removed on 5th post-operative day and the second one on 8th POD, with a length-of-stay of 11 days. At a further pathological examination, the dCCA of the extra-pancreatic bile duct (pT1 with R0 resection) was confirmed but a second neoplasm of 18 mm on the pancreatic margin was detected as pancreatic ductal adenocarcinoma (PDA) (pT1c with R2 resection). Out of 23 resected lymph nodes, 4 had metastases from adenocarcinoma NOS (pN2).

At pathological examination (Figure 3A-D), ectasia of the proximal two-thirds of the extra pancreatic common bile duct was observed. The distal third up to the entrance to the pancreatic parenchyma showed two distinct lumens with walls of increased consistency. Histological examination revealed two lesions with evidence of interposed spared tissue: a first lesion was in the extra pancreatic portion of the bile duct with the aspect of a well differentiated biliary-type adenocarcinoma, dCCA, infiltrating the wall and extending to the peribiliary adipose tissue and with foci of high-grade biliary intraepithelial neoplasia (BilIN-HG). This neoplastic lesion was positive for Cytokeratin 7, Cytokeratin 19 and CDX2; Cytokeratin 20 and SATB2 were negative and nuclear positivity for SMAD/DPC4 was lost. The second neoplastic focus was a well differentiated PDA, extended to the retro pancreatic adipose tissue and to the resection margin of the pancreatic parenchyma. It was positive for Cytokeratin 7, Cytokeratin 19 and CDX2; Cytokeratin 20 and SATB2 were negative and nuclear positivity for SMAD/DPC4 was lost. As described, from a morphological point of view, although similar, the two lesions showed some cytoarchitectural differences, while from an immunohistochemical point of view, as expected, they showed an overlapping pattern. Furthermore. The presence of aspects of BilIn-HG suggested for primitivity of the dCCA. The analysis of the metastatic lymph nodes did not show any morphological and immunohistochemical features that could not be unequivocally referred to one of the two neoplasms.

Table 2. Pre-operative laboratory data

Laboratory data		
White blood cells	10.53	x 10^3/µL
Hemoglobin	13.0	g/dL
Platelets	198	x 10^3/µL
INR	1.08	
Creatinine	0.69	mg/dL
Sodium	135	mMol/L
Potassium	4.15	mMol/L
Aspartate aminotransferase	63	UI/L
Alanine aminotransferase	61	UI/L
Gamma-glutamyl transpeptidase	407	UI/L
Total bilirubin	6.71	mg/dL
Direct bilirubin	5.89	mg/dL
Albumine	36	g/L
CEA	1.2	ng/mL
Ca 19.9	80.5	UI/mL

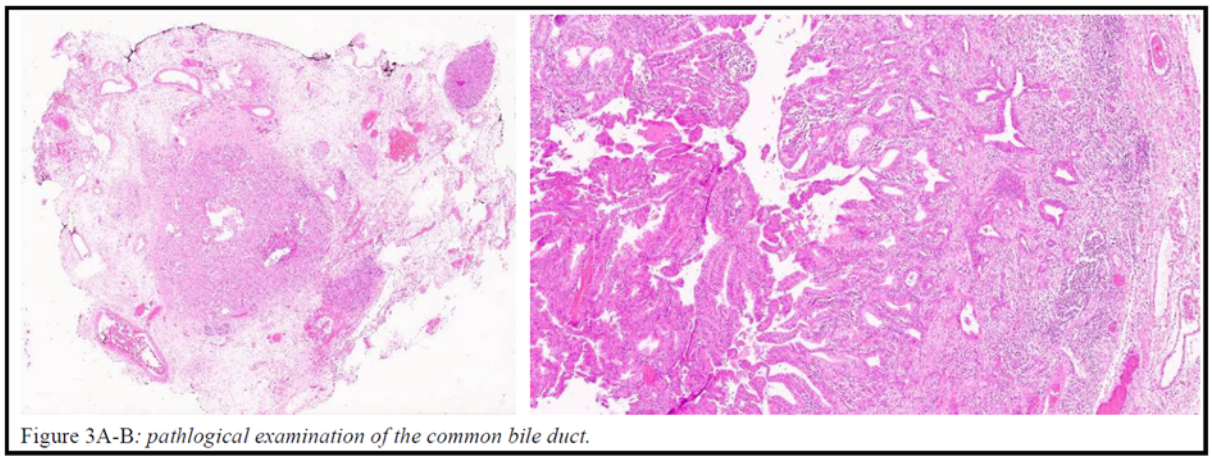

Figure 3A-B*: pathlogical examination of the common bile duct.*

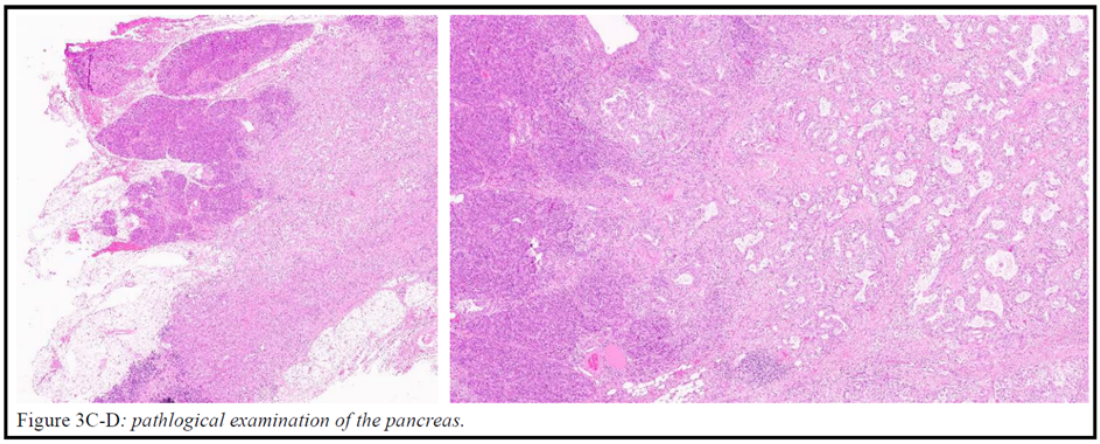

Figure 3C-D*: pathlogical examination of the pancreas.*

After the dismission, the patient was referred to a specialized Center, where no adjuvant chemotherapy was decided due to his comorbidities and age, and only a clinical and instrumental follow-up was established. One year after-surgery, no signs of recurrence have been observed on contrast-enhanced CT. After 18 months the patient is still stable, oral feeding is regular and no gastrointestinal disorders were referred. The last laboratory data were normal range except for a slight increase of the Ca19.9 marker (Table 3).

Table 3. Laboratory data 18 months after surgery

Laboratory data		
White blood cells	6.53	x $10^3/\mu L$
Hemoglobin	12.0	g/dL
Platelets	190	x $10^3/\mu L$
Creatinine	0.81	mg/dL
Aspartate aminotransferase	15	UI/L
Alanine aminotransferase	<5	UI/L
Gamma-glutamyl transpeptidase	10	UI/L
Total bilirubin	0.37	mg/dL
CEA	1.6	ng/mL
Ca 19.9	73.5	UI/mL

Discussion

We report an unusually rare case of distal cholangiocarcinoma synchronous with a pancreatic ductal adenocarcinoma. The detection of the second neoplasm was possible only at pathologic examination, because the neoplasia, despite its dimensions, was not seen during the preoperative imaging (abdominal CT, MRCP, and the endoscopic US). Although, a R0 resection of the dCCA was performed, the resection for the unacknowledged pancreatic adenocarcinoma must be considered as a R2 resection. The patient's age, his comorbidities, and the potentially low-aggressive nature of both neoplasias allowed the survival of the patient during the 18 months, without any treatment before or after the surgery. Actually, radical resection is the only curative therapy either for dCCA and PDA. Therefore, despite the poor 5-year overall survival [8,9], in this case the surgical treatment has been the best therapeutic option for the patient allowing him to live without major complications up to now.

Conclusion

Our approach on elderly patient affected by a rare overlap of dCCA and PDA, was surprisingly successful, prolonging survival and a satisfactory quality of life.

Conflict of interest: The authors have not conflict of interest to declare for this work.

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