Clinics in Oncology

Case Report
Published: 18 Jun, 2020

Total Pancreatectomy with Spleen Preservation for Distal Cholangiocarcinoma Synchronous with Multifocal IPMNs and a Neuroendocrine Tumour of the Pancreas

Abdelhak Lamara[1], Badreddine Nini[1], Mohamed Boukhane[1], Sid Ahmed Medjahdi[1], Amel Soualmia[2], Abdelkrim Rehamnia[3], Nacereddine Lemaici[4] and Saliha Benyarbah[1]*

[1]Department of General Surgery, Regional Military University Hospital of Constantine, Algeria

[2]National Transplant Coordination, Algeria

[3]Department of Gastroenterology, Regional Military University Hospital of Constantine, Algeria

[4]Department of Pathology, Regional Military University Hospital of Constantine, Algeria

Abstract

Introduction: Total pancreatectomy is the treatment of choice for multiple tumor locations in the pancreas, especially when it comes to the multifocal association of different lesions.

Case Presentation: We report the case of a 62-year-old woman who underwent a complete pancreatectomy with spleen preservation for distal cholangiocarcinoma synchronous with multifocal IPMNs and a neuroendocrine tumor of the pancreas.

Discussion: In the presence of a diffuse disease of the pancreas and/or in the case of tumor association, total pancreatectomy remains the only valid oncological option.

Conclusion: Total pancreatectomy can be performed safely and without added risk in the presence of multiple or multifocal lesions of the pancreas.

Keywords: Total pancreatectomy; Multifocal lesions; TIPMN; Distal cholangiocarcinoma

OPEN ACCESS

Correspondence:
Abdelhak Lamara, Department of General Surgery, Regional Military Hospital University Hospital of Constantine, 5 RM, BP: 61 C, 25001, Constantine, Algeria, Tel: (213)661579759; E-mail: lamaraabdelhak2000@yahoo.fr

Received Date: *07 May 2020*
Accepted Date: *08 Jun 2020*
Published Date: *18 Jun 2020*

Citation:
Lamara A, Nini B, Boukhane M, Ahmed Medjahdi S, Soualmia A, Rehamnia A, et al. Total Pancreatectomy with Spleen Preservation for Distal Cholangiocarcinoma Synchronous with Multifocal IPMNs and a Neuroendocrine Tumour of the Pancreas. Clin Oncol. 2020; 5: 1712.

Introduction

The synchronous coexistence of three different types of tumors in a patient remains rare. It is sometimes not easy to differentiate between a Distal Cholangiocarcinoma (DCC) and an Adenocarcinoma of the Head of the Pancreas (AHP). The coexistence of intraductal tumor of the pancreas IPMNs and a DCC may be linked to the possibility of the pre-existing bile duct tumors (BT-IPMN). MRI and Echoendoscpic ultrasonogram (EUS) are very useful for diagnosis and monitoring. The presence of diffuse localizations on the pancreas always poses a problem concerning the type of surgical resection that must be proposed to ensure oncological control of the disease. Total Pancreatectomy (PT) is the only alternative for cancer surgery without added mortality or morbidity.

Case Presentation

A 62-year-old woman was seen in surgery consultation for fever and hypochondrium pain. The patient mentioned similar episodes associated with chills evoking flares of cholangitis; she had no pruritus. The patient is followed in dialectology for non-insulin-dependent diabetes and in cardiology for hypertension for 3 years. The patient has also had surgery for gallbladder stones 40 years ago. The clinical examination was normal. The biology showed: Alkaline phosphatases at 1.5 times the upper limit of normal (N), gamma-glutamyl-transferase at 4 N, total bilirubinemia at 4.8 mg/l. The serology was negative, CA 19-9 antigen was at 213 ng/ml. (5 N). Weight 73 kg high of 164 cm.

Imaging (ultrasound and CT scan) showed a centimeter round nodule in the distal bile duct (Figure 1). The EUS showed a tumor polypoid in distal bile duct without sign of aggression or loco regional extension (Figure 2).

On the pancreas, EUS showed pancreatic steatosis with Intraductal Papillary and Mucinous tumor (IPMNs) of the pancreatic secondary canals with signs of degeneration at the isthmic and caudal level but without signs of loco regional infiltration. The pathology of the biopsies was in favor

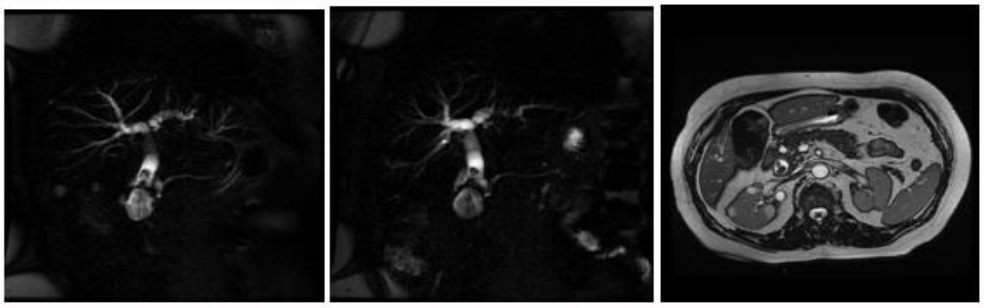

Figure 1: Magnetic resonance cholangiopancreatogram demonstrating distal Cholangiocarcinoma and multifocal secondary branch duct intraductal papillary mucinous neoplasms.

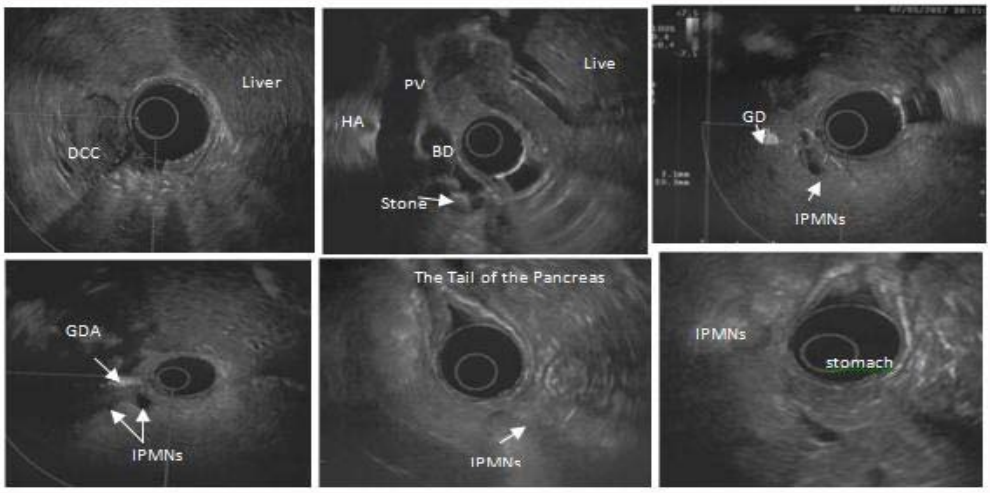

Figure 2: Endoscopic ultrasonogram showing a polypoide process in the retropancreatic distal bile duct purely intraanalar without signs of aggressiveness or locoregional extension.
Pancreatic steatosis with papillary and IPMNs of the pancreatic secondary duc with suspicious signs of degeneration at the isthmic and caudal level but without signs of locoregional infiltration. (EUS Performed by Doctor Nadjet SAKHRI).

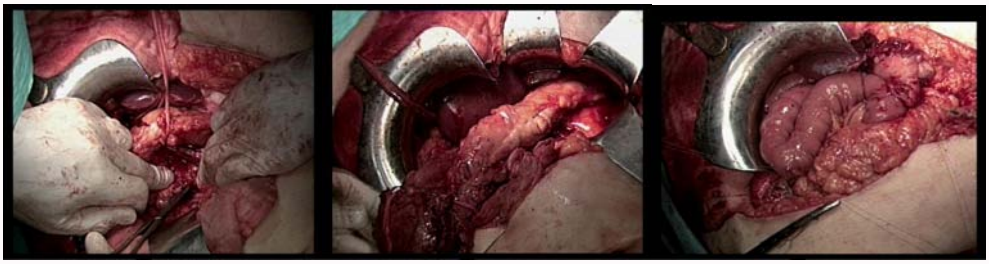

Figure 3: Total pancreatectomy procedure.

of a distal cholangiocarcinoma and IPMNs (Figure 2).

A total pancreatectomy was proposed given the coexistence of multiple lesions on the pancreas associated with a distal cholangiocarcinoma. The patient was well informed about her diabetes, how to deal with hypoglycemia, insulin injection techniques and self-monitoring, and pancreatic extracts at each meal. A total pancreatectomy with conservation of the spleen without conservation of the pylorus was performed, associated with a cleaning of the hepatic pedicle, the celiac trunk the splenic pedicle and total resection of the mesopancreas. The bile duct was resected just at level of the convergence. The patient received two globular pellets intraoperatively; the intervention time was 5 h 53 min (Figure 3).

In postoperative, we adopted the principle of fast track, postoperative analgesia with morphine, insulin therapy, and a diet comprising 55% carbohydrates, 25% lipids and 20% protein, enzyme substitution treatment with proton pump inhibitor during the 3 postoperative months and during follow-up. The patient was hospitalized at the diabetology unit for two episodes of hypoglycemia.

Definitive anatomopathology finds an ulcerobourgeonant tumor of 2 cm in the distal bile duct, and 19 peri-pancreatic nodes. The histology showed a well-differentiated cholangiocarcinoma of 2 cm from the distal bile duct infiltrating the muscularis. There is a presence of tumor vascular emboli, and an absence of perineural involvement. The nodes are not infiltrated (PT2NOMO). Several foci of infra-centimeter gastric type IPMNs, some of which are of low-

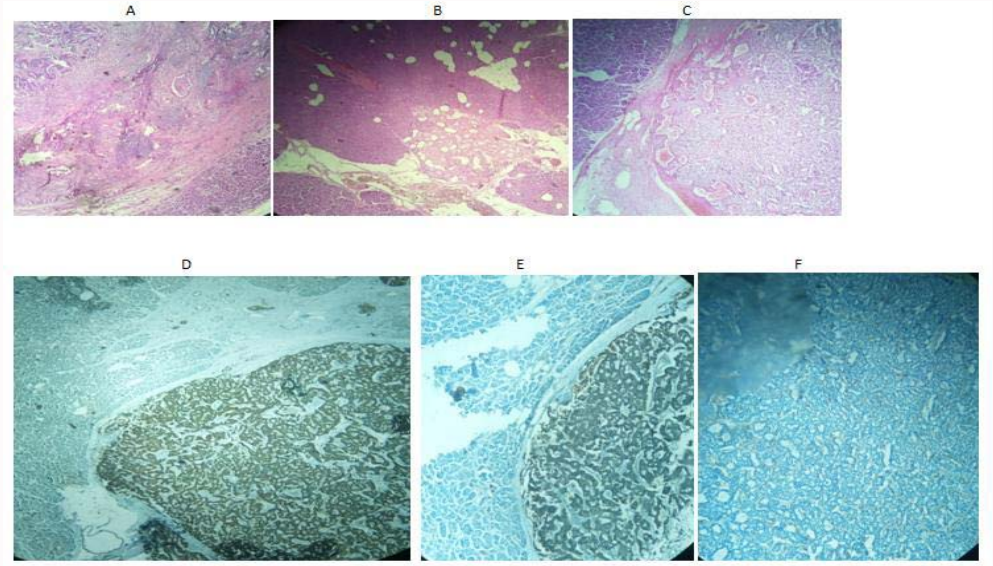

Figure 4: Definitive anatomopathology.
A) Distal cholangiocarcinoma infiltrating the pancreas. B) IPMN (HE: x40). C) Neurodocrinetumor (HE: x40). D) Immunohistochemistry chromogranine A positive. E) Immunohistochimistry NSE positive. F) Ki 67: 2%.

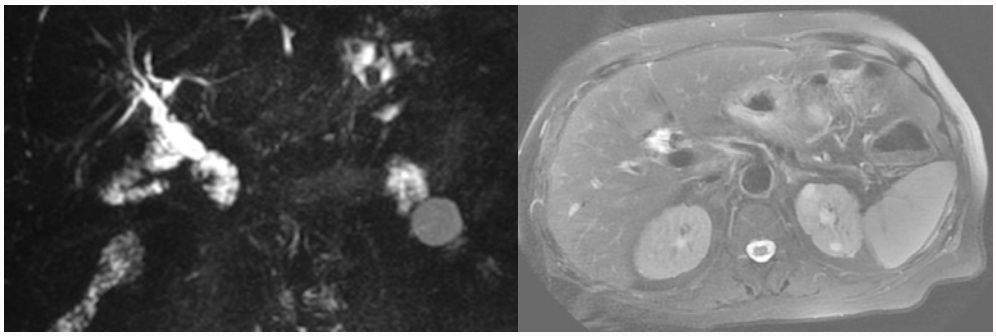

Figure 5: MRI 18 months after operation: No recurrences.

grade dysplasia in the head, and a well differentiated neuroendocrine tumor of 05 mm (G1) from the tail of the pancreas were also found (Figure 4). CT scan and MRI at 6 months, 12 months and 18 months did not show signs of recurrence (Figure 5).

Discussion

The association of IPMNs and DCC seems very rare and requires more observations. In the case of our patient the question that arises is: Is it an association with degenerate BT-IPMN and IPMNs?

Papillary tumors of the bile ducts (BT-IPMN) are rare, often multifocal and obstructive; transformation into invasive carcinoma is possible. Invasive forms have a very poor prognosis [1,2]. CT scan, C-MRI and UE are the key first-line exam for diagnosis and follow-up of IPMNs.

The presence of jaundice in a patient with cystic lesions of the head of the pancreas, the presence of a solid component within the cystic mass or of a wall nodule taking contrast and reaching the main pancreatic duct >9 mm are in favor pejorative forms [3]. Although these neoplasms share similar clinical and histological characteristics. Based on their significant risk of malignancy, BT-IPMN requires radical surgical resection and careful monitoring [4].

It is not uncommon to confuse DCC with AH of the pancreas. UE can be useful in differentiating between the two pathologies, especially in cases where CT scan cannot detect an intrapancreatic mass or dilation of the pancreatic duct [5]. CCD has a favorable prognosis compared to adenocarcinoma of the pancreas [6,7]. The objective of surgical resections is to allow the removal of all progressive lesions. However, the damage can be multifocal and therefore requires a total pancreatectomy, as is the case with our patient. A total pancreatectomy performed for degenerate IPMNs associated with CCD and the histology revealed an associated pancreatic neuroendocrine tumor. Total pancreatectomy may be performed for diffuse pancreatic disease. Mortality from complications of diabetes is rare. The quality of life and the difficulties of managing diabetes after total pancreatectomy should be taken into account and reserve this technique for carefully selected patients [8].

Operative mortality is improved compared to previous reports. Greater survival benefits have been seen in younger patients with smaller resected tumors with the negative margin and in the absence of lymph node infiltration [9]. Long-term survival was equivalent to a partial pancreatectomy. Therefore, total pancreatectomy can be used in multifocal or diffuse pancreatic locations. Patients should receive adequate doses of synthetic insulin and pancreatic enzyme

supplementation to ensure long-term survival [10]. It is important to define the survival characteristics of patients with invasive pancreatic tumors who have undergone TP and above all to provide a basis for comparative analyze on the benefits of removing the entire gland with the intention to erase all the microscopic disease [9]. Oncological resection RO is a predictor of DCC survival, 5-year survival goes from 0% to 27% depending on the radicality of the surgical excision [11,12]. The perineural invasion and the extra-capsular lymphatic extension also represent poor prognostic factors [11,13]. The presence of positive Lymph node is associated with poor prognosis [11,13,14]. Neoadjuvant treatment based on Gemcitabine or 5FU seems to have a benefit on the possibility of radical surgery R0, 33% complete response [15], survival is better after neoadjuvant chemotherapy *vs.* adjuvant chemotherapy (53% *vs.* 23%), postoperative radiotherapy can be useful for the control of microscopic residues [15,16].

Conclusion

The coexistence of multifocal pancreatic tumor lesions and distal cholangiocarcinoma leaves no choice for partial resection of the pancreas or even preservation of the central pancreas. Total pancreatectomy remains the only guarantee of an oncologic resection of such lesions. The reduction in mortality rate and the possibility of managing postoperative complications prompted surgeons to propose total pancreatectomy as the reference treatment for multiple and multifocal lesions of the pancreas.

References

1. Adimoolam V, Sanchez MJ, Siddiqui UD, Yu S, Dzuira JD, Padda MS, et al. Endoscopic ultrasound identifies synchronous pancreas cystic lesions not seen on initial cross-sectional imaging. Pancreas. 2011;40(2):1070-2.
2. Kubo H, Nakamura K, Itaba S, Yoshinaga S, Kinukawa N, Sadamoto Y, et al. Differential diagnosis of cystic tumors of the pancreas by endoscopic ultrasonography. Endoscopy. 2009;41(8):684-9.
3. Crippa S, Bassi C, Salvia R, Malleo G, Marchegiani G, Rebours V, et al. Low progression of intraductal papillary mucinous neoplasms with worrisome features and high-risk stigmata undergoing non-operative management: A mid-term follow-up analysis. Gut. 2017;66(3):495-506.
4. Minaguawa N, Sato N, Mori Y, Tamura T, Higure A, Yamaguchi K. A comparison between intraductal papillary neoplasms of the Biliary Tract (BT-IPMNs) and Intraductal Papillary Mucinous Neoplasms of the Pancreas (P-IPMNs) reveals distinct clinical manifestations and outcomes. Eur J Surg Oncol. 2013;39(6):554-8.
5. Yokoyam Y, Ebata T, Igami T, Sugawara G, Mzuno T, Yamaguchi J, et al. Different clinical characteristics between distal cholongiocarcinoma and pancreatic head carcinoma with bilary obstruction. Pancreas. 2017;46(10):1322-6.
6. Ethun CG, Lopez-Aguiar AG, Pawlik TM, Poultsides G, Idrees K, Fields RC, et al. Distal cholongiocarcinome and pancreas adenocarcinoma: Are they really the same disease? A 13-institutio study from the US extrahepatic biliary malignancy consortium and the central pancreas consortium. J Am Coll Surg. 2017;224(4):406-13.
7. Nakanuma Y, Kakuda Y. Pathologic classification of cholangiocarcinoma: New concepts. Best Pract Res Clin Gastroenterol. 2015;29(2):277-93.
8. Wu W, Dodson R, Makary MA, Weiss MJ, Hirose K, Cameron JL, et al. A contemporary evaluation of the cause of death and long-term quality of life after total pancreatectomy. World J Surg. 2016;40(10):2513-8.
9. Johnston WC, Hoen HM, Cassera MA, Newell PH, Hammill CW, Hansen PD, et al. Total pancreatectomy for pancreatic ductal adenocarcinoma: Review of the National Cancer Data Base. HPB (Oxford). 2016;18(1):21-8.
10. Suzuki S, Kajiyama H, Takemura A, Shimazaki J, Nishida K, Shimoda M. The clinical outcomes after total pancreatectomy. Dig Surg. 2017;34(2):142-50.
11. Murakami Y, Uemura K, Hayashidani Y, Sudo T, Hashimoto Y, Ohge H, et al. Prognostic significance of lymph node metastasis and surgical margin status for distal cholangiocarcinoma. J Surg Oncol. 2007;95(3):207-12.
12. Fernandez-Ruiz M, Guerra-Vales JM, Colina-Ruizdelgado F. Comorbidity negatively influences prognosis in patients with extrahepatic cholangiocarcinoma. World J Gastroenterol. 2009;15(42):5279-86.
13. Yoshida T, Matsumoto T, Sasaki A, Morii Y, Aramaki M, Kitano S. Prognostic factors after pancreatoduodenectomy with extended lymphadenectomy for distal bile duct cancer. Arch Surg. 2002;137(1):69-73.
14. Katayose Y, Nakagawa K, Yamamoto K, Yoshida H, Hayashi H, Mizuma M, et al. Lymph nodes metastasis is a risk factor for bone metastasis from extrahepatic cholangiocarcinoma. Hepatogastroenterology. 2012;59(118):1758-60.
15. McMasters KM, Tuttle TM, Leach SD, Rich T, Cleary KR, Evans DB, et al. Neoadjuvant chemoradiation for extrahepatic cholangiocarcinoma. Am J Surg. 1997;174(6):605-8.
16. Nelson JW, Ghafoori AP, Willett CG. Prognostic factors after pancreatoduodenectomy for distal bile duct cancer. J Radiat Oncol Biol Phys. 2009;73:148-53.