

Case Report

Diffuse biliary papillomatosis (IPNB) associated with IPNM : a case report

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Introduction

Biliary papillomatosis is a rare event. It concerns the biliary epithelium. The definition of the World Health Organization is the presence of multifocal papillary lesions on intra hepatic and/or extrahepatic bile ducts 1. There is a risk of malignant transformation. The papillary lesions are producing mucus conducting to biliary obstruction. Right hypochondrium pain and jaundice are the more frequent symptoms. Some common points have been described between IPNB and IPNM 2,3. IPNB can be a diffuse disease of all the biliary tree, with a real risk of neoplastic transformation and it could be a therapeutic challenge. We report the case of a patient suffering from diffuse IPNB associated with mixt IPNM who underwent pancreaticoduodenectomy and liver transplantation.

Presentation of the case

A 60 years old man was followed and treated in our hospital since 3 years for a presumed IgG 4 related disease because he has an history of repeated acute pancreatitis and CT scan features of notches on the two kidneys. However, the rate of circulating IgG4 was normal. In april 2021, he was admitted with a jaundice. CT scan demonstrated an extra hepatic biliary stenosis with above a biliary dilatation (Fig.1). ERCP and spy glass examination found a stenosis and several bilateral defects in the biliary lumen in all the extra hepatic biliary common bile duct and in the right and left bile ducts (Fig. 2). Spy glass biopsies found low grade dysplasia on the left duct, high grade dysplasia on the right duct and the common bile duct. We decided to perform a pancreaticoduodenectomy (PD) with endoscopic peroperative examination of intra hepatic bile ducts and biopsies. During PD, we resected all the extra hepatic biliary tree and we noticed that the left pancreas had completely disappeared with thrombosis of the splenic vein and segmental portal hypertension due probably to the multiple episodes of acute pancreatitis. Biliary anastomosis was done on the upper biliary convergence which was clearly pathologic. Peroperative endoscopy confirm several papillary lesions on left and right biliary ducts. Specimen examination had described a mixed IPNM with high grade dysplasia on the pancreas and IPNB with high grade and low grade dysplasia particularly on the right duct and the upper biliary convergence. All the resected lymph nodes were normal. The post operative

period was marked by gastroparesis and glycemic imbalance. Finally, the evolution was favourable.

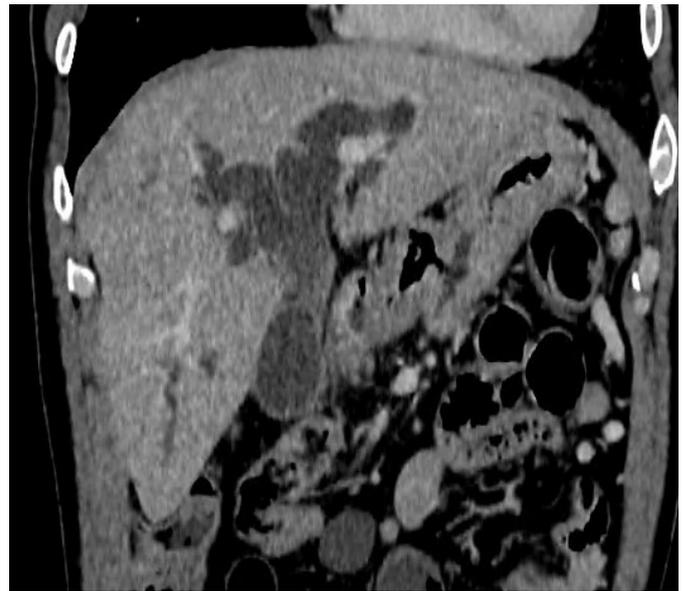


Figure 1 : Pre operative CT scan in coronal section, showing significant dilatation of the intra and extra hepatic bile ducts



Figure 2 : Initial ERCP showing images of substraction within the common bile duct corresponding to endobiliary papillary proliferation and mucus

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After a few months of relatively good general status, the patient developed fever, jaundice, malnutrition and intra hepatic abscess (Fig. 3). His condition needed multiple hospitalisations for antibiotics and enteral nutrition with jejunostomy. In six months, he has developed a secondary biliary cirrhosis.



Figure 3 : CT scan in axial section 1 year after total duodeno-pancreatectomy showing dilatation of the bile ducts and two hepatic abscesses on this section. Note the splenomegaly.

Decision was taken to register him on the french liver transplant waiting list with request of priority. Cadaveric donor was available in April 2022. The procedure was very difficult with multiple biliary abscess and important hemorrhage. Postoperative recovery was quite simple. Specimen analysis of the explant liver (Fig.4) described diffuse IPNB with numerous areas of high grade dysplasia and cholangiocarcinoma lesions on the upper biliary convergence, exactly on the site of the first bilio-enteric anastomosis.

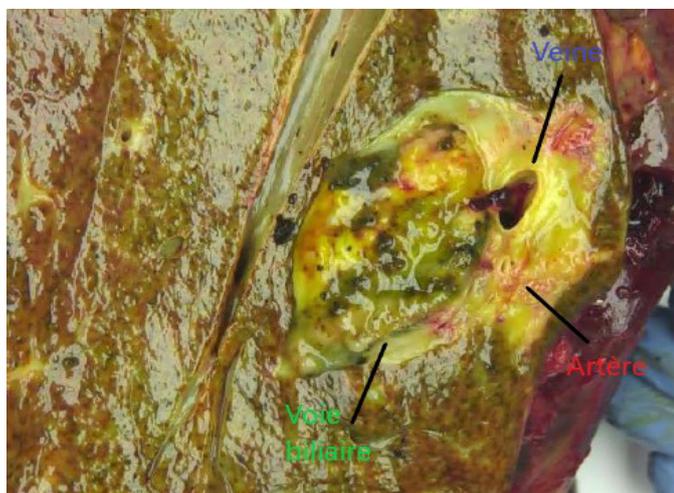


Figure 4 : Section of anatomopathology of the explant liver. We see the endobiliary proliferation, the vein with a thrombus

Discussion

IPNB can be localized or diffuse. The diffuse form represents a real therapeutic challenge. Endoscopic biopsies can not evaluate correctly the importance of the disease because different evolutionary stages exist in different part of the biliary tree. Currently, the curative treatment of the diffuse form must be resection of all the biliary tree that's to say pancreaticoduodenectomy and liver transplantation. Previous cases of liver transplant have been reported in this disease 4,5. Some authors have proposed a first surgical procedure (Liver resection or pan-

atocoduodenectomy) to stage at the best the disease and eliminate advanced cases with lymph nodes encasement, 5,6. It was our strategy in the case report and we have been confronted to a quick evolution to cholangiocarcinoma. The pancreatic disease of the patient, it was a mixed form, spanning the entire pancreas, with high grade dysplasia and some similar cases, combining IPNB and IPNM, have been reported 7–13. There are some common points between IPNB and IPNM. Four subtypes of epithelium have been described: biliary pancreatic, intestinal, gastric and oncocytic. In IPNB and IPNM, the subtype has an influence on prognosis 6. Thus, pancreatobiliary type is associated to more invasive lesions, lymph nodes encasement and postoperative recurrence. IPNB and IPNM have mucin secretion and risk of neoplastic transformation 2,3. Some what, IPNB is the biliary mirror of IPNM. However, there are differences: the type more frequent in IPNB is pancreatobiliary while it is the gastric type in IPNM 14. IPNB can have non secreting form. Finally, IPNB had a worse prognosis.

In our case, the patient was followed since two years for IgG 4 related disease. A posteriori, this diagnosis was wrong, images of notch on the kidneys would be infectious sequelae.

Conclusion

IPNB are very rare biliary tumors which can be localized or diffuse with a high risk of neoplastic transformation. The treatment is the complete resection which can conduct to liver transplantation. IPNB has been described as the « biliary equivalent » of IPNM but IPNB seems to be a more aggressive disease.

Reference

1. Histological Typing of Tumours of the Gallbladder and Extrahepatic Bile Ducts | SpringerLink. Accessed May 31, 2022. <https://link.springer.com/book/10.1007/978-3-642-84241-2>
2. Zaccari P, Cardinale V, Severi C, et al. Common features between neoplastic and preneoplastic lesions of the biliary tract and the pancreas. *World J Gastroenterol.* 2019;25(31):4343-4359. doi:10.3748/wjg.v25.i31.4343
3. Biliary papillary tumors share pathological features with intraductal papillary mucinous neoplasm of the pancreas - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/17058219/>
4. Papillomatose multifocale des voies biliaires: une indication de transplantation hépatique - EM consulte. Accessed May 31, 2022. <https://www.em-consulte.com/article/16276/papillomatose-multifocale-des-voies-biliaires-une->
5. Vibert E, Dokmak S, Belghiti J. Surgical strategy of biliary papillomatosis in Western countries. *J Hepato-Biliary-Pancreat Sci.* 2010;17(3):241-245. doi:10.1007/s00534-009-0151-1
6. Intraductal papillary neoplasm of the bile duct - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/24379576/>
7. Ren X, Zhu CL, Qin XF, Jiang H, Xia T, Qu YP. Co-occurrence of IPMN and malignant IPNB complicated by a pancreatobiliary fistula: A case report and review of the literature. *World J Clin Cases.* 2019;7(1):102-108. doi:10.12998/wjcc.v7.i1.102
8. Intraductal mucinous tumors occurring simultaneously in the

- liver and pancreas - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/12522542/>
9. A case of mucin hypersecreting intraductal papillary carcinomas occurring simultaneously in liver and pancreas - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/15729259/>
 10. Intraductal Papillary Mucinous Tumor Simultaneously Involving the Liver and Pancreas: A Case Report. Accessed May 31, 2022. <https://www.jpatholm.org/journal/view.php?number=2802>
 11. Simultaneous intraductal papillary neoplasms of the bile duct and pancreas treated with chemoradiotherapy - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/22403738/>
 12. Xu XW, Li RH, Zhou W, et al. Laparoscopic resection of synchronous intraductal papillary mucinous neoplasms: A case report. *World J Gastroenterol WJG*. 2012;18(44):6510-6514. doi:10.3748/wjg.v18.i44.6510
 13. [Synchronous malignant intraductal papillary mucinous neoplasms of the bile duct and pancreas requiring left hepatectomy and total pancreatectomy] - PubMed. Accessed May 31, 2022. <https://pubmed.ncbi.nlm.nih.gov/24561701/>
 14. Laurent L, Lévy P. La néoplasie papillaire intraductale des voies biliaires (IPN-B), équivalent « biliaire » de la TIPMP? *Hépatogastro Oncol Dig*. 2014;21(6):460-466. doi:10.1684/hpg.2014.1030