



# Intraductal Papillary Neoplasm of the Bile Duct: A Diagnostic Challenge for Optimal Treatment<sup>☆</sup>

## Neoplasia papilar intraductal de vía biliar: un reto diagnóstico para un correcto tratamiento

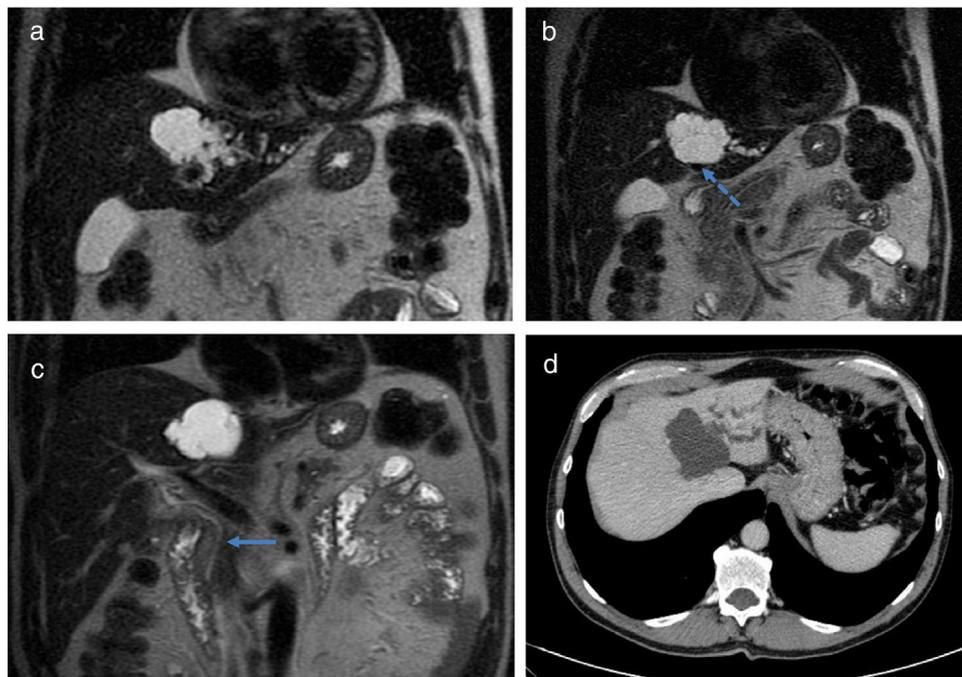
Intraductal papillary neoplasm of the bile duct (IPNB) is an uncommon condition characterized by the exophytic papillary growth of the biliary epithelium toward the ductal lumen. Previously included in the group of tumors with the same name located in the pancreas, these neoplasms present evident differences and, since 2010, they have been considered a separate entity with demonstrated potential for malignization toward cholangiocarcinoma.<sup>1,2</sup>

We present the case of a patient with IPNB who was operated on at our hospital.

The patient is a 59-year-old male, who was asymptomatic and had no pathological history. A routine ultrasound detected a 5 × 5 cm cystic lesion in the left hepatic lobe, with dilation of the intrahepatic left bile duct (BD), while the extrahepatic duct was normal. Computed tomography and magnetic resonance cholangiography showed evidence of a polylobulated cystic lesion in segment IV, with thin interior walls and no solid or enhanced areas. It caused compression

of the left portal branch and dilatation of the bile ducts of segments II-III, which came into contact with the lesion (Fig. 1). Endoscopic ultrasound confirmed the described findings, as well as the normal diameter of the extrahepatic BD (Fig. 2). The lab work-up showed normal liver function and tumor markers.

With the suspected diagnosis of mucinous cystic neoplasm of the liver, surgical treatment was indicated. A laparoscopic approach was used to perform intraoperative ultrasound, which located the lesion in segment IV, in close contact with the left portal vein and the middle hepatic vein, and BD dilatation in segments II-III was confirmed. Laparoscopic left hepatectomy was performed. The postoperative period was uneventful and the patient was discharged on the fourth day. The histopathological study revealed the presence of ducts with cystic dilatations containing mucin, covered by cylindrical epithelium, with foci in the shape of papillary projections and no invasion of the stroma, all compatible with



**Figure 1 – (a) Magnetic resonance cholangiography image showing the cystic lesion and dilatation of the left intrahepatic bile duct; (b) MRCP image showing the relationship of the lesion with the left portal branch, displacing it (dashed arrow); (c) MRCP image showing a normal diameter of the extrahepatic bile duct (solid arrow); (d) computed tomography image showing the lesion in segment IV and dilation of the left intrahepatic bile duct.**

<sup>☆</sup> Please cite this article as: de la Serna S, Pérez-Aguirre E, Díez-Valladares LI, García-Botella A, Torres García AJ. Neoplasia papilar intraductal de vía biliar: un reto diagnóstico para un correcto tratamiento. Cir Esp. 2018;96:457-459.



**Figure 2 – Endoscopic ultrasound image showing normal main bile duct diameter (solid arrow) and dilatation of the left bile duct with intrahepatic cystic lesions (dashed arrow).**

pancreatobiliary IPNB, with low-grade dysplasia and free resection margins. After a 40-month follow-up, the patient did not present recurrence.

IPNB is a very infrequent pathology in our environment. It usually occurs in middle-aged Asian men, and risk factors include hepatolithiasis as well as clonorchiasis.<sup>3</sup> It tends to be multifocal and can occur at any point of the extra- or intrahepatic BD, the latter being more frequent in the left hepatic lobe,<sup>4,5</sup> as in our case. Although up to 30% of patients remain asymptomatic, presentation is usually in the form of pain, obstructive jaundice or cholangitis.<sup>4</sup>

Preoperative diagnosis is difficult and requires the combination of several imaging tests; the differential diagnosis should include cystic liver lesions. The radiological appearance observed on ultrasound, computed tomography and magnetic resonance is BD dilatation (with three possible patterns: diffuse duct ectasia, segmental dilatation or cystic dilatation), associated or not with intraductal masses.<sup>3,6</sup> In our case, the cystic, walled, compressive nature, with no apparent communication with the BD, led to the erroneous presumptive diagnosis of mucinous cystic neoplasm of the liver.<sup>6</sup> Endoscopic retrograde cholangiography may be useful for confirming BD dilatation and filling defects, but mucobilia is also evident in one-third of patients.<sup>5</sup> Intraductal ultrasound and cholangioscopy (transhepatic or oral) have also been used with biopsy, although these are only cost-efficient for the diagnosis of malignancy in 50% of cases, due to multicentricity.<sup>7</sup>

From the pathological standpoint, despite being more heterogeneous, these neoplasms are classified into four subtypes like pancreatic intraductal papillary neoplasms (pancreatobiliary, intestinal, gastric and oncocytic). The pancreatobiliary type is the most frequent in IPNB, which usually expresses mucin core proteins-1 (MUC-1) and has a poorer associated prognosis than the remaining subtypes.<sup>2,4,5,8</sup>

Considering the degree of dysplasia and depth of the invasion, IPNB are classified into 4 stages: (1) IPNB with mild-moderate dysplasia; (2) IPNB with high-grade dysplasia; (3) stage T1 cholangiocarcinoma of the American Joint Committee on Cancer; and (4) stage  $\geq$ T2 cholangiocarcinoma of the American Joint Committee on Cancer.<sup>3,9</sup> In 40–80% of the cases, there will be foci of invasive carcinoma or of mucinous or tubular adenocarcinoma, a figure that is significantly higher than in pancreatic intraductal papillary neoplasms.<sup>4,8</sup>

It is because of this potential for malignization that surgical treatment is indicated whenever distant disease is ruled out. Major hepatectomy is recommended, either with or without lymphadenectomy and associated or not with BD resection.<sup>7</sup> It is important to check the biliary resection margins since these tumors seem to have a superficial dissemination pattern, which, according to the series, is responsible for a recurrence rate (mainly locoregional) that reaches 47–60% within 5 years in cases of malignant degeneration.<sup>3,7</sup> Nevertheless, the prognosis of these lesions seems to be better than in conventional cholangiocarcinoma, with survival rates close to 69% 5 years after surgery.<sup>3,4,7,10</sup>

In conclusion, despite the infrequent presentation of IPNB in our setting and its difficult preoperative diagnosis, this entity with a potential for malignancy should be included in the differential diagnosis of cystic liver lesions. With radical surgical treatment and free resection margins, the prognosis of these patients with localized lesions is acceptable.

#### REFERENCES

1. Albores-Saavedra J, Kloppel G, Adsay NV, Sripa B, Crawford JM, Tsui WMS, et al. Carcinoma of the gallbladder and extrahepatic bile duct. In: Bosman FT, Carneiro F, Hruban

- RH, Theise ND, editors. WHO classification of tumors of the digestive system Lyon: IARC Press; 2010; p. 266-73.
- Nakanuma Y, Kakuda Y, Uesaka K, Miyata T, Yamamoto Y, Fukumura Y, et al. Characterization of intraductal papillary neoplasm of bile duct with respect to histopathologic similarities to pancreatic intraductal papillary mucinous neoplasm. *Hum Pathol.* 2016;51:103-13.
  - Wan XS, Xu YY, Qian JY, Yang XB, Wang AQ, He L, et al. Intraductal papillary neoplasm of the bile duct. *World J Gastroenterol.* 2013;19:8595-604.
  - Rocha FG, Lee H, Katabi N, DeMatteo RP, Fong Y, D'Angelica MI, et al. Intraductal papillary neoplasm of the bile duct: a biliary equivalent to intraductal papillary mucinous neoplasm of the pancreas? *Hepatology.* 2012;56:1352-60.
  - Ohtsuka M, Shimizu H, Kato A, Yoshitomi H, Furukawa K, Tsuyuguchi T, et al. Intraductal papillary neoplasms of the bile duct. *Int J Hepatol.* 2014;2014:459091.
  - Egri C, Yap WW, Scudamore CH, Webber D, Harris A. Intraductal papillary neoplasm of the bile duct: multimodality imaging appearances and pathological correlation. *Can Assoc Radiol J.* 2017;68:77-83.
  - Kim WJ, Hwang S, Lee YJ, Kim KH, Park KM, Ahn CS, et al. Clinicopathological features and long-term outcomes of intraductal papillary neoplasms of the intrahepatic bile duct. *J Gastrointest Surg.* 2016;20:1368-75.
  - Kloek JJ, van der Gaag NA, Erdogan D, Rauws EA, Busch OR, Gouma DJ, et al. A comparative study of intraductal papillary neoplasia of the biliary tract and pancreas. *Hum Pathol.* 2011;42:824-32.
  - Hwang S, Lee YJ, Song GW, Park KM, Kim KH, Ahn CS, et al. Prognostic impact of tumor growth type on 7 th AJCC staging system for intrahepatic cholangiocarcinoma: a single-center experience of 659 cases. *J Gastrointest Surg.* 2015;19:1291-304.
  - Luvira V, Pughem A, Bhudhisawasdi V, Pairojkul C, Sathitkarnmanee E, Luvira V, et al. Long-term outcome of surgical resection for intraductal papillary neoplasm of the bile duct. *J Gastroenterol Hepatol.* 2017;32:527-33.

Sofía de la Serna\*, Elia Pérez-Aguirre,  
Luis Ignacio Díez-Valladares, Alejandra García-Botella,  
Antonio José Torres García

Servicio de Cirugía General y del Aparato Digestivo, Hospital Clínico San Carlos, Instituto de Investigación Sanitaria del Hospital Clínico San Carlos (IdISSC), Madrid, Spain

\*Corresponding author.

E-mail address: [sdlserna@me.com](mailto:sdlserna@me.com) (S. de la Serna).

2173-5077/

© 2017 AEC. Published by Elsevier España, S.L.U. All rights reserved.

## Ruptured Anastomotic Pseudoaneurysm of the Common Femoral Artery After Wrongful Diagnosis of Inguinal Hernia<sup>☆</sup>

### Rotura de pseudoaneurisma anastomótico de arteria femoral común tras diagnóstico erróneo de hernia inguinal

In recent years, femoral artery pseudoaneurysms have been an increasingly frequent complication. They can be caused by peripheral catheterization or appear at the site of a bypass anastomosis (vein or stent).<sup>1,2</sup> According to the latest series, anastomotic pseudoaneurysms (APA) complicate arterial anastomoses from 1.4% to 4%. Their mean interval for presentation is 6 years, and even less if associated with superinfection (the presence of bacterial colonization in APA is 80%).<sup>3,4</sup> APA present as a dilation of the arterial wall adjacent to the anastomosis, and its most common manifestation is a painful and hyperpulsatile inguinal mass,<sup>2</sup> which is why it should be included in the differential diagnosis of all inguinal masses. Thrombosed APA that are overinfected may present as a non-pulsatile mass with surrounding erythema, imitating a strangulated hernia.<sup>5</sup> There is a life-threatening risk for APA rupture and hemorrhagic shock, entailing a surgical emergency that makes its early diagnosis of utmost importance.<sup>6</sup>

We present the case of a 76-year-old hypertensive, insulin-dependent diabetic, obese male patient with a body mass index of 37.5. His medical history included revascularized ischemic cardiomyopathy with a triple coronary bypass and atrial fibrillation treated with Sintrom<sup>®</sup>. His surgical history of interest included left carotid endarterectomy and repair of an uncomplicated abdominal aortic aneurysm with aortobiiliac bypass and right iliofemoral bypass with Dacron<sup>®</sup> prosthesis together with right femoral endarterectomy 12 years earlier. The patient came to the general surgery outpatient clinic for a symptomatic right inguinal mass that increased with Valsalva maneuvers and had grown progressively over the past year; he was diagnosed with right inguinal hernia and was pending weight reduction to schedule surgical repair.

<sup>☆</sup> Please cite this article as: García Blanco VE, Ruales Romero AM, Galera Martínez MC, Dóiz Artázcoz E, Rodríguez Piñero M. Rotura de pseudoaneurisma anastomótico de arteria femoral común tras diagnóstico erróneo de hernia inguinal. *Cir Esp.* 2018;96:459-461.

