Biliary papillomatosis
Case report

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Biliary papillomatosis. Case report

Biliary papillomatosis is a disease characterized by multiple papillary tumours of variable distribution and extent in the intrahepatic and/or extrahepatic biliary tree. Papillary carcinoma can develop within these lesions. Because biliary papillomatosis is a rare biliary pathological entity, its clinical features and outcome are not well known. The course of this disease is very prolonged. This diagnostic difficulty is due above all to the problematic distinction between biliary papillomatosis and cholelithiasis. Therapeutic strategy should be decided pre-operatively among resection, transplantation or stent. In many cases surgical strategy is decided during surgery, considering the mass extension, the local infiltration and the patient's age, and, when possible, with the aid of extemporaneous histological examination.
Recently we experienced a case of biliary papillomatosis of the common hepatic duct in a 82-year-old man, presented with obstructive jaundice and pain in the right upper quadrant and epigastrium radiated to the ipsilateral scapula. First we made cholecystectomy and we positioned the T-tube. The follow-ups performed with laboratory tests and T-tube cholangiogram showed no jaundice but the filling defect in the common bile duct was still present. So we performed a resection of the common bile duct and an hepatico-jejunostomy at the hepatic pedicle. The histological examination showed a villous adenoma of the common bile duct with high-grade dysplasia.

KEY WORDS: Biliary papillomatosis, Common hepatic duct, Surgery.

Introduction
Biliary papillomatosis is a disease characterized by multiple papillary tumours of variable distribution and extent in the intrahepatic and/or extrahepatic biliary tree [1-5]. Papillary carcinoma can develop within these lesions. Because biliary papillomatosis is a rare biliary pathological entity, its clinical features and outcome are not well known [5].
Recently we experienced a case of biliary papillomatosis of the common hepatic duct in a 82-year-old man.

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Case Report

In November 2009 an 82-year-old man was admitted to the hospital because of obstructive jaundice and pain in the right upper quadrant and epigastrium radiated to the ipsilateral scapula. Laboratory findings included total serum bilirubin 3.68mg/dl, direct serum bilirubin 2.79mg/dl, AST 379U/l, ALT 285U/l, alkaline phosphatase 189U/l, GGT 534mg/dl, leukocyte count 7,700/mm3. The patient reported having undergone an endoscopic retrograde cholangiopancreatography (ERCP) with drainage of the common bile duct five years before at another hospital. An abdominal ultrasonography showed dilatation of both the common bile duct and the intrahepatic ducts, dense material in the terminal common bile duct and distended gallbladder with biliary sludge. A first ERCP showed dilated common bile duct and intrahepatic ducts, with multiple gallstones. A sphincterotomy was performed and biliary sludge was extracted. Because of incomplete extraction, a second

ERCP was performed after one week with extraction of gallstones and sludge. The toilet was still incomplete, so a third ERCP was made the following month showing inhomogeneous and adherent material at the common hepatic duct bifurcation, which was impossible to remove. Surgical exploration was performed in January 2010: first we made cholecystectomy, then the intraoperative cholangiography showed the common bile duct dilated and full of mucus mixed with gallstones, and we positioned the T-tube. Histological examination showed subacute-chronic cholecystitis pseudofolliculitis.

The follow-ups in February 2010 and April 2010 performed with laboratory tests and T-tube cholangiogram showed no jaundice but the filling defect in the common bile duct was still present. In April 2010 we removed the Kehr tube.

In June 2010 the patient underwent a new ERCP which showed a dilatation and a filling defect in the common bile duct. After balloon catheter extraction, this resulted in an extrusion of friable polypoid masses. The specimen consisted of fragments of papillary neoplasm with moderate, focally severe, dysplasia .

In July 2010 the patient underwent a second surgery. Surgical exploration demonstrated a brownish mass extended within the common bile duct up to the hepatic pedicle. Therefore we performed a resection of the common bile duct and an hepatico-jejunostomy at the hepatic pedicle, and the resections margins were free. The patient was discharged on the 23rd postoperative day. The histological examination showed a villous adenoma of the common bile duct with high-grade dysplasia.

Discussion

Biliary papillomatosis is a rare disease. The pathogenesis is still unknown, but various biliary conditions (such as recurrent pyogenic cholangitis and congenital choledochal cyst) were reported to be associated with this pathology [6-9]. Biliary papillomatosis is more common in men than in women, in their sixth or seventh decade of life and it is not so different from usual cholangiocarcinoma [5].

The most common clinical manifestations are recurrent abdominal pain, repeated episodes of acute cholangitis and obstructive jaundice due to cholestasis. Acute cholangitis is the second most common manifestations of biliary papillomatosis [5].

Some considerations can be made as a result of our experience. The course of this disease is very prolonged: in fact, in our case, about seven years passed before the diagnosis was made. This diagnostic difficulty is due above all to the problematic distinction between biliary papillomatosis and cholelithiasis. In literature we found other cases where the diagnosis was difficult.

Yeung et al. reported seven cases of biliary papillomatosis. One patient, who presented only epigastric pain, was diagnosed to have hepatoma and only surgical exploration revealed biliary papillomatosis in the left hepatic lobe and a small hepatoma in the right lobe. In another patient, who presented cholangitis and jaundice, the correct diagnosis was delayed for 2 years. He was treated initially as in choledocholithiasis with cholecistectomy and exploration of the common bile duct. The postoperative T-tube cholangiogram and an ERCP performed 2 years later revealed the same filling defects and the diagnosis of biliary papillomatosis was hypothesized [6].

Also, Imvrios et al. published a case of a 43-year-old man admitted to the hospital with obstructive jaundice and treated initially with stent insertion in the common bile duct by ERCP. Two years later, because of repeated episodes of cholangitis, he underwent exploratory laparotomy with choledochotomy and cholangioscopy which revealed extensive masses and stones in the left and right intra- and extrahepatic bile ducts [10].

These objective difficulties depend on the difficulty of making a correct diagnosis by means of the instrumental examinations that we have. Ultrasound can demonstrate non-specific bile duct dilatation and intraductal solid masses with no distal acoustic shadowing. Magnetic resonance cholangiopancreatography (MRCP) shows multiple irregular filling defects or the presence of intraductal masses connecting with a pedicle to the bile duct. In ERCP typical endoscopic features are multiple small filling defects and direct visualization of excessive mucus discharge from the papilla of Vater [11-13]. It is necessary to be careful because also intraductal papillary neoplasm of the pancreas is characterized by mucus ipersecretion and mucobilia [14-15]. On cholangioscopic evaluation, sludge material intermingled with pus frequently covering the papillary masses and multiple papillary masses were observed within the bile duct lumen [5].

Cholangioscopy would be the best diagnostic instrument because it allows direct visualization of the lesion, but it is not available to all hospitals.

On the base of our experience, we can say that the first characteristic which can be helpful to make the diagnosis is the lesion morphology, particularly if the filling defect remains the same over the time and can't be removed by endoscopy. In our experience the possibility to make an histological examination of material from the papilla of Vater was decisive. The histological findings allowed us to make the diagnosis.

After the diagnosis, the choice of the best treatment is very difficult. Resection is the treatment of choice when biliary papillomatosis is localized, according to preoperative imaging workup and with the support of intraoperative ultrasound or cholangioscopy [6,11,16,17]. The extent of resection should be decided in most cases during surgery. If the patient cannot undergo major surgery, local ablation, stenting or drainage palliative procedures are considered [6,11]. In the case of diffuse biliary papillomatosis liver transplantation is the treatment of choice [18,19]. Bilobar or recurrent disease, as well as the high risk

of malignant transformation should favour total hepatectomy and liver transplantation to be considered as the ultimate curative approach [6].

We choose in our case to perform a resection of the common bile duct and an hepatico-jejunostomy at the hepatic pedicle, because in our patient the lesion was in the extrahepatic bile duct below the hepatic pedicle. Then we reviewed the literature to compare our surgical strategy with others, and we found different choices according to the extension of the papillomatosis .

Hanafy and McDonald published a case of villous adenoma of the common bile duct including the cystic duct in a 76-year-old man, treated by resection of the mass and the gallbladder in one piece [20].

Delaunay et al. published a case of biliary papillomatosis of the common bile duct in a 74-year-old man, treated by means of a pancreaticoduodenectomy. A MRCP performed three months after surgery revealed residual biliary papillomatosis at the hepatic pedicle [21].

Chae et al. published a case of biliary papillomatosis of the common bile duct and the left intrahepatic duct in a 77-year-old man. In consideration of the patient's age and general condition, resection of the common bile duct and cholecystectomy were performed, while extensive surgeries as left lobectomy and pancreaticoduodenectomy were abandoned [22].

Yeung et al. published seven cases of biliary papillomatosis and among these only one case of biliary papillomatosis localized in the common bile duct, a 70-year-old man treated by pylorus-preserving pancreaticoduodenectomy [6].

Sotona et al. published a case of papillary adenoma of the common bile duct, treated by bile duct resection with Roux-en-Y hepaticojejunostomy [23].

Doberaurer et al. published a case of villous adenoma of the common bile duct in a 75-year-old woman, treated by partial resection of the common bile duct [24].

Guglielmi et al. published a case of papillomatosis of the biliary tract in a 75-year-old woman, treated by cholecystectomy, choledochotomy and positioning of a permanent T-Tube [25].

Sagar et al. published a case of a 57-year-old woman who underwent cholecystectomy and exploration of the common bile duct for cholelithiasis and choledocholithiasis. Two years later she presented again obstructive jaundice and the ERCP showed several filling defects in the lower end of the common bile duct. At laparotomy, a transduodenal approach to the common bile duct was preferred to avoid reopening it. A sphincteroplasty was made and a balloon catheter was inserted, with extraction of a solitary stone and abnormal tissue [26].

Therapeutic strategy should be decided pre-operatively among resection, transplantation or stent. In many cases surgical strategy is decided during surgery, considering the mass extension, the local infiltration and the patient's age, and, when possible, with the aid of extemporaneous histological examination.

Riassunto

La papillomatosi della via biliare è una patologia caratterizzata da multiple formazioni papillari delle vie biliari sia intra- che extra-epatiche, su cui può svilupparsi un carcinoma papillare. Le caratteristiche cliniche di questa patologia della via biliare non sono ben conosciute, in quanto è molto rara. Il suo decorso è generalmente molto prolungato. Questa difficoltà diagnostica deriva dalla difficile distinzione tra papillomatosi e calcolosi della via biliare. Una volta fatta la diagnosi, la strategia terapeutica dovrebbe essere decisa preoperatoriamente tra resezione, trapianto o stent. In molti casi viene decisa durante l'intervento chirurgico stesso, considerando l'estensione della massa, l'infiltrazione locale e l'età del paziente e, dove possibile, grazie anche all'aiuto dell'esame istologico estemporaneo.

Recentemente abbiamo avuto un caso di papillomatosi del dotto epatico comune in un uomo di 82 anni, che si era presentato con ittero ostruttivo e dolore in epigastrio e ipocondrio destro, irradiato posteriormente. Inizialmente abbiamo eseguito la colecistectomia con la coledocolitotomia, posizionando un tubo di Kehr. I follow-up hanno dimostrato con gli esami di laboratorio e con la colangiografia trans-Kehr che l'ittero era scomparso, mentre persisteva il difetto di riempinmento della via biliare. Quindi abbiamo eseguito la resezione della via biliare principale con epaticodigiunostomia all'ilo. L'esame istologico ha mostrato un adenoma villoso della via biliare principale con displasia di alto grado.

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