Diagnosis and management of choledochal cysts. A review of 10 new cases


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RESUMEN

Objetivo: analizar las técnicas diagnósticas y las distintas posibilidades terapéuticas de esta rara patología.

Pacientes y métodos: presentamos un estudio retrospectivo de todos los casos de quistes de colédoco diagnosticados en nuestro hospital desde 1991. Incluye 10 nuevos casos y distintos tipos de quistes de colédoco (QC).

Resultados: de los 10 pacientes diagnosticados de QC siete tenían menos de 10 años. En cuanto a los diferentes tipos de QC: 7 eran de tipo I, 1 de tipo III, 1 de tipo IVa y otro de tipo V. Las manifestaciones clínicas habitualmente tenían un perfil biliar o pancreático. El diagnóstico se ha realizado mediante técnicas de imagen no invasivas (ECO, TAC) y CPRE. El tratamiento ha venido condicionado por el tipo de quiste: quistectomía con hepatico-eyunostomía en “Y de Roux” en los de tipo I, CPRE con esfinterotomía endoscópica en los de tipo III y trasplante hepático en la enfermedad de Caroli. En el único QC tipo IVa se realizó una papilotomía transductal y una colecistectomía. Todos han sido controlados periódicamente, sin que hayamos registrado complicaciones relevantes durante el seguimiento.

Conclusión: los QC son más frecuentes en niños pero no son excepcionales en adultos. Las técnicas de imagen y la CPRE son esenciales en el diagnóstico de esta patología. El tratamiento dependerá del tipo de quiste de colédoco. Es aconsejable el seguimiento periódico y prolongado de estos pacientes para detectar precozmente complicaciones como la transformación maligna.

Palabras clave: Quistes de colédoco. Diagnóstico. Tratamiento.

ABSTRACT

Aim: the aim of this study was to analyze the diagnostic and therapeutic options for the various types of this rare disease.

Patients and methods: 10 patients with choledochal cysts (CC) were diagnosed in our hospital since 1991. Type of cyst was established according to the Alonso-Lej classification.

Results: we report 7 type-I, 1 type-III, 1 type-IVa, and 1 type-V CC cases. Clinical manifestations were abdominal pain in all cases with biliary or pancreatic features. The diagnosis was established using abdominal ultrasonography, computed tomography, and endoscopic retrograde cholangiopancreatography (ERCP). All 7 patients with type-I CC underwent total cyst excision with Roux-en-Y hepatico-eyunostomy. For type-III CC an endoscopic sphincterotomy (ES) was performed, and in type-IVa CC a transuductal sphincterotomy and cholecistectomy was made. The patient with Caroli’s disease (type V) underwent liver transplantation. We have followed up all patients for several years without significant complications.

Conclusions: CC is more frequent in childhood, but is not exceptional in the adult. Imaging techniques and ERCP play an important role in the diagnosis, and also in the treatment of type-III cysts. Therapeutic options depend on cyst type, but due to the potential malignancy of this disease total cyst excision is recommended for types I, II and IV. In type-III CC endoscopic sphincterotomy is recommended, while liver transplantation is sometimes necessary for type V. Long-term follow-up is crucial to prevent malignant transformation except for type-III CC where this complication is very unusual.

Key words: Choledochal cyst. Diagnosis. Therapy.

INTRODUCTION

Choledochal cyst (CC) is a rare disease in our setting, with an incidence of 1 case per 100,000-150,000 live births (1). It is more common in Asia, particularly in Japan, and has a female predominance (2,3). It is usually
diagnosed in childhood, with 80% of cases being diagnosed before 10 years of age (3). Its cause is unknown and several hypotheses have been proposed, including abnormalities in the pancreatobiliary junction, which are frequently associated with biliary cysts (4-9).

Choledochal cysts are divided into 6 types according to the classification established by Alonso-Lej in 1954 and modified by Todani in 1977 (10,11). Type I is most common (80-90% of all C.C) (12) and consists of a fusiform or segmental dilation of the common bile duct. Type II or diverticulum in the cystic duct, and type III or choledochocoele are the least common (2%) (13). Type IV is the second most common (10-15%) (12) and consists of multiple cystic dilations of the intra- and extrahepatic biliary tract (IVa), or only the extrahepatic biliary tract (IVb). Type V or Caroli’s disease is a diffuse cystic involvement of the intrahepatic biliary tract. The aim of this article is to present our case series of this rare disease, and to evaluate the diagnostic and therapeutic options used in each case.

PATIENTS AND METHODS

We reviewed 10 cases of C.C diagnosed in the Gastroenterology and Pediatric Surgery Departments of Miguel Servet University Hospital, Zaragoza (Spain), since 1991. The type of C.C was established according to Alonso-Lej’s classification as modified by Todani (10,11).

We captured clinical and biochemical data at the time of diagnosis, diagnostic method, and therapy used for all patients.

RESULTS

Ten patients with C.C have been diagnosed in our hospital since 1991. Seven patients had type-I C.C, 1 patient had type-III C.C, 1 had type-IVa C.C, and 1 type-V C.C. Seven patients were under 10 years of age at the time of diagnosis, 1 was 13 years old, and 2 were adults: The patient with choledochocoele (type-III C.C) was 44 years old, and the patient with Caroli’s disease (type-V C.C) was 45 years old. Age at the time of surgery ranged from 17 months in the youngest patient to 46 years in the oldest. Mean age in children was 5.1 years. Four of the patients were female and 6 were male.

The most common clinical presentation was abdominal pain, which was present in all cases and associated with nausea and vomiting in 6 patients. Six of them also had jaundice and/or cholangitis, and 2 of the children had recurrent pancreatitis. In the patient with type-III C.C the diagnosis was made after frequent, repeated episodes of abdominal pain, and clinically and biochemically documented pancreatitis. The adult with type-V C.C initially had nonspecific symptoms of abdominal pain, subsequently complicated by recurrent cholangitis (Table I).

Laboratory data revealed cholestasis with elevation of ALP, GGT, and bilirubin in 6 cases. Five patients also showed a slight elevation of liver enzymes, and hyperamylasemia in the context of pancreatitis episodes was noted in 3 patients.

After presenting these symptoms, an abdominal ultrasonogram (US) and/or computerized tomography scan (CT) was performed in all patients to guide diagnosis. The diagnosis was subsequently confirmed in 8 patients by ERCP. In patient no. 10 (CC type V) ERCP did not diagnose the illness, and the Caroli’s disease case was confirmed after left hepatectomy and histological study. In case no. 5 ERCP was not performed. In case no. 9 CT misdiagnosed a pancreatic pseudocyst, and this error was corrected with ERCP (Fig. 1). In only one patient, a common terminal duct for the common bile duct and the pancreatic duct were found on ERCP.

Treatment was surgical in a large majority of cases, and the technique used depended on the type of cyst. In all patients with type-I cysts, cystectomy with reconstruction of the biliary tract using a Roux-en-Y hepatojunostomy was performed. No complications occurred during the immediate postoperative period, and patients were discharged under long-term antibiotic therapy (amoxi-
cillin-clavulanic acid for 6 months). In the patient with choledochocele (type III) (Fig. 2), the only therapeutic intervention was endoscopic sphincterotomy performed during ERCP (Fig. 3) to ensure correct drainage of the biliary and pancreatic tracts. After 3 years of follow-up the patient has had no recurrences of abdominal pain or new episodes of pancreatitis. In the patient with type-IVa choledochal cysts, due to the difficulty of anastomosis if a complete resection of the cyst was performed, surgery was limited to transductal papillotomy and cholecystectomy in order to achieve, as in the previous case, adequate drainage of the biliary tract. In the patient with Caroli’s disease, as previously mentioned, a left hepatic lobectomy was initially performed due to recurrent cholangitis. After recurrence of symptoms and the detection of cystic lesions in the remaining portion of the liver, a liver transplantation was required (Table II).

We have controlled all patients every month with clinical and biochemical tests during the first six months, and every six months with abdominal US afterwards. Follow-up has ranged from 3 to 16 years and 8 patients are still being controlled. No morbidity or mortality was associated with surgery or ERCP. All patients were discharged without problems. No late complications have been recorded to date, and repeat surgery has not been required due to stenosis at the anastomosis. No case of degeneration to cholangiocarcinoma was seen during follow-up. One of the patients with a type-I biliary cyst underwent surgery for a mesenteric lipoma 6 years later, with subsequent reoperation for bands.

<table>
<thead>
<tr>
<th>Table II. Treatment</th>
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<tr>
<td>- Type I CC: Cystectomy + Roux-en-Y hepatojunostomy (7 cases)</td>
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<tr>
<td>- Type III CC: Endoscopic sphincterotomy (1 case)</td>
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<tr>
<td>- Type IVa CC: Cholecystectomy + transductal sphincterotomy (1 case)</td>
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<td>- Type V CC: 1. Left hepatectomy. 2. Liver transplantation (1 case)</td>
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DISCUSSION

In this study we report an interesting series of cases that summarizes the different types of CC and their therapeutic options.

The etiology of biliary cysts remains uncertain; it is still controversial whether they are a congenital or acquired condition (3). The most generally accepted theory is that they are caused by congenital abnormalities in the pancreatobiliary junction, which, according to several authors, are present in 39 to 96% of patients with CCs (4-7). The junction of the common bile duct with the pancreatic duct outside the duodenal wall, forming a long common channel, would favor reflux of pancreatic enzymes into the biliary tract with damage and dilation of the bile duct wall (6-8). For other authors dilation is caused by increased pressure in the biliary tract (9). However, there are CCs in which this particular feature is not found, and of all 9 ERCPs performed in our series it was reported only in one. Therefore, this association was rarely found in our series.

CCs are usually diagnosed during childhood, but 20% of cases are identified during adulthood (3). This is consistent with the proportion found in our case series.

The most common symptom of CC, abdominal pain, was not absent in any of our patients. It was associated in 6 cases with cholestasis or frank jaundice, and sometimes with cholangitis. The typical triad of pain, jaundice, and a palpable abdominal mass is rarely seen in full. In three of our patients pancreatitis was the form of clinical presentation. Pancreatitis may be caused by a difficult drainage of pancreatic enzymes in a sac-like papilla, or by direct compression by the cyst on the pancreatic duct (Fig. 1). In cases diagnosed during adulthood nonspecific symptoms are more commonly found, with intermittent abdominal pain and few alarm symptoms (14,15).

Imaging tests, such as abdominal US and CT, may suggest the diagnosis, but not always correctly (16,17). In our case no. 9, with clinical signs of recurrent pancreatitis, the diagnosis based on CT was of a pancreatic pseudocyst. An accurate diagnosis was reached by ERCP, which classified the cyst, measured its size and location, and showed its anatomic relationships prior to surgery (14,18). Magnetic resonance cholangiopancreatography (MRCP) may replace ERCP, particularly in children, since it is a noninvasive examination with fewer complications (18,19). We have this imaging technique available since 2004. In this period we have diagnosed no cases of CC, but ERCP should currently be reserved for therapy in this disease.

Treatment depends on type of CC. Authors agree in recommending resection surgery in type-I CCs. This resolves the clinical problem and prevents the occurrence of cholangiocarcinoma, whose incidence in patients over 20 with CC can be as high as 28% (2,3,6,14,20). The increased risk of cholangiocarcinoma in these patients may be due to sustained reflux of pancreatic enzymes and stagnation of bile salts, causing chronic inflammation of the cystic mucosa and subsequent malignant degeneration (2,21). If this occurs survival is very poor and most patients die within 2 years (2,14,20). The most widely used surgical technique is cystectomy with reconstruction of the anatomic continuity using a Roux-en-Y hepatojunostomy (3,6,14,15,20,22-25). The seven patients with type-I CC in our study followed this surgery and have been free of symptoms and complications. Some of them are young adults nowadays.

The likelihood of malignant degeneration is much lower in type-III CCs (2,6,21), and in these cases the treatment of choice is ES (6,17,20,24,26,27). This therapeutic approach allows an adequate drainage of the biliary and pancreatic tracts. We used ES in the sole patient with type-III CC, and achieved a long-lasting remission of symptoms. We have followed up this patient for 7 years, and finally discharged him 3 years ago.

The treatment of type-IVa CCs has been a subject of controversy. Most published studies recommend resection of extrahepatic cysts followed by a Roux-en-Y hepatojunostomy (6,20,24). Some authors are more aggressive and believe that hepatic lobectomy should be performed if intrahepatic cysts are confined to a single lobe (28). In the only patient we included with this type of cysts surgery was limited to ensuring external drainage for the biliary tract through transductal papillotomy and cholecystectomy. This was due to the technical difficulty in performing a hepatojugal anastomosis on a dilated intrahepatic bile duct.

Finally, in the patient with Caroli’s disease a precise diagnosis was only established after surgery (left hepatectomy). When clinical symptoms relapsed and diffuse hepatic damage occurred, which produced portal hypertension, the patient underwent liver transplantation with good results to date. This was the usual approach followed in the other studies reviewed. For unilobar disease the most effective therapeutic option is resection of the affected lobe (20,30). In the case of diffuse cystic involvement treatment should initially be medical with ursodeoxycholic acid and bile salt binders, but sometimes, if secondary biliary cirrhosis develops, liver transplantation is necessary (22,29-31).

Patients with CC require close regular follow-up, mainly aimed at an early detection of potential cholangiocarcinoma. Malignant degeneration occurs most often in type-I cysts, followed by type-IVa (2) cysts. In type-IVa cysts the intrahepatic portion of the cyst is most often left intact, and these patients should therefore be monitored very closely. We followed all patients every month for the first six months, and every six months afterwards using biochemical tests and abdominal US. Monitorization should last for life (32). Complete cyst resection substantially reduces this risk but does not prevent it completely, and there are several reports of cholangiocarcinoma years after cyst excision (2,33).
To conclude CCs are more frequent during childhood but are not uncommon in adults. Abdominal US, CT, and ERCP play an important role in the diagnosis of most patients, and in the case of type-III CC also in their treatment. CRNM is presently replacing ERCP as a diagnostic method in this disease. Therapeutic options have usually been determined by type of cyst: cystectomy with Roux-en-Y hepaticojunostomy in type I, ERCP with ES in type III, and initial medical treatment in Caroli’s disease, although hepatic lobectomy and an eventual liver transplant were required. In type IVa at least extrahepatic biliary tract cysts should be resected when possible. Long-term follow-up is required in these patients for an early detection of possible complications such as malignant degeneration, except in type-III CC, where transformation to colangiocarcinoma is very rare.

REFERENCES

6. Lópezes RR, Pinson CW, Campbell JR, Harrison M, Katton R. Varia-
17. García-Cano J, Nieto I, Godoy MA, Gómez Ruiz CJ, Sánchez-Man-
19. Lam WW, Lam TPW, Staing H, Chan FL, Chan KL. MR cholangi-
ography and TC cholangiography of pediatric patients with choledochal cysts. AJR 1999; 173: 401-5.
22. Shi LB, Peng SY, Meng XK, Peng CH, Liu YB, Chen XP, et al. Dia-
gnosis and treatment of congenital choledochal cyst: 20 years experi-
24. Scudamore CH, Hemming AW, Teare JP, Fache JS, Erb SR, Watkin-
28. Todani T, Narusue M, Watanabe Y, Tabuchi K, Okajima K. Manage-
29. Medrano-Caviedes R, Artigas V, Sancho FJ, Martín-Hargraves G, Ro-
31. Madariaga JR, Iwatsuki S, Starzl TE, Todo S, Selby R, Zetti G. He-