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Original articles

Choledochal cysts in adults and the importance of differential diagnosis

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Abstract
Background/Purpose. Choledochal cysts, congenital cystic dilatations of the hepatobiliary tree, present diagnostic dilemmas. In countries where hydatid disease is endemic, this rare pathology may be overlooked. The importance of this issue led us to reexamine our clinical material to evaluate our findings and management.
Methods. This study examined 12 patients (11 females, 1 male) who were treated at the University of Istanbul, Cerrahpasa Medical Faculty, with the diagnosis of choledochal cyst between 1981 and 2000. The cases were examined retrospectively, and the complaints, diagnostic methods, findings, and management were discussed.
Results. Ten of the cysts were type I (83.3%), one was type II (8.3%), and one was type IV (8.3%). Three of the patients underwent operation with the diagnosis of liver hydatid cyst, but during the operation the cysts were found to be choledochal cysts. In six of the patients, the cysts were correctly diagnosed, and they were excised.
Conclusions. Contemporary approaches to treating choledochal cysts focus on total removal of the cyst. In a country where hydatic diseases are endemic, hepatobiliary cystic lesions are often misdiagnosed clinically and radiologically as hydatid cysts. Considering that percutaneous and laparoscopic approaches are being increasingly adopted for treatment of hydatid cysts, the problematic area must be subjected to thorough examination for cystic lesions. The possibility of choledochal cysts must always be kept in mind; otherwise, an unfavorable process may begin for both patient and surgeon.

Key words Choledochal cyst · Differential diagnosis · Adults

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Introduction

Although considered a childhood disease, choledochal cysts are detectable in adults as well; they comprise 20% of all the cases.[1] This rare pathology, which is well defined as a congenital cystic dilatation of the biliary tree, may be overlooked, erroneously diagnosed, or detected unexpectedly during operation.[2–4] Its possibility must be considered during the diagnosis of a cystic mass in the hepatobiliary area. The importance of the issue led us to reexamine all our clinical material, the existing literature, and available up-to-date information.

Materials and methods

The study examined 12 patients treated at the University of Istanbul, Cerrahpasa Medical Faculty, with the diagnosis of choledochal cysts between 1981 and 2000. The cases were examined retrospectively; and complaints, diagnostic methods, and resulting operations were reviewed.

Results

Our series includes 12 patients: 11 females and 1 male. The youngest patient was 16 years old and the oldest 61 (mean 34 years). In six of the patients, the cases were correctly diagnosed before operation. Three of the patients were subjected to operation with a diagnosis of liver hydatid cyst, but the cysts were found to be choledochal during operation (Fig. 1). Between 1982 and 1984, another three patients were subjected to operation with a diagnosis of mechanical obstruction of the biliary tree but were rediagnosed during the operation as having choledochal cysts. As supportive methods of diagnosis, ultrasonography (US) and computed

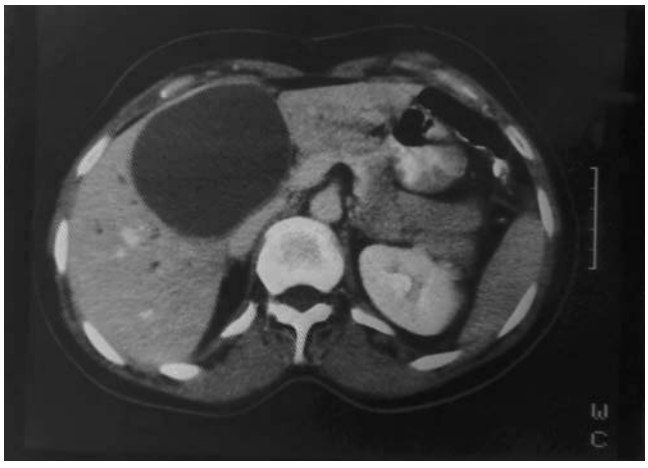


Fig. 1. Computed tomography (CT) image of a patient with the preoperative misdiagnosis of liver hydatid cyst and the peroperative diagnosis of choledochal cyst



Fig. 2. Endoscopic retrograde cholangiopancreatography (ERCP) image shows contrast material filling a type I choledochal cyst

Table 1. Overview of the patients

Pt. no.	Age/ gender	Preop. diagnosis	Diagnostic method	Date of the operation	Type of cyst	Type of operation
1	16/F	MOBD	Physical exam + lab	04/09/1982	I	Cholecystectomy + Roux-Y cystojejunostomy
2	31/F	MOBD	Physical exam + lab	09/12/1982	I	Cholecystectomy + cystoduodenostomy
3	25/F	MOBD	Physical exam + lab	18/10/1984	I	Cholecystectomy + cystoduodenostomy
4	39/M	Choledochal cyst	Physical exam + lab; US + CT + ERCP	03/05/1987	I	Cholecystectomy + cystoduodenostomy, T-tube drainage
5	22/F	Choledochal cyst	Physical exam + lab; US + CT + ERCP	21/09/1989	I	Cholecystectomy + cyst excision + Roux-Y hepaticojejunostomy
6	20/F	Choledochal cyst	Physical exam + lab; US + CT + ERCP	08/08/1990	I	Cholecystectomy + cyst excision + Roux-Y hepaticojejunostomy
7	61/F	Hydatid cyst + cholelithiasis	Physical exam + lab; US + CT	27/05/1992	I	Cholecystectomy + cystoduodenostomy
8	47/F	Hydatid cyst	Physical exam + lab; US + CT	19/09/1994	I	Cholecystectomy + cyst excision + Roux-Y Hepaticojejunostomy
9	19/F	Choledochal cyst	Physical exam + lab; US + CT + ERCP	04/10/1994	I	Cholecystectomy + cystoduodenostomy
10	23/F	Choledochal cyst	Physical exam + lab; US + CT + ERCP	15/06/1999	I	Cholecystectomy + cyst excision + Roux-Y hepaticojejunostomy
11	46/F	Hydatid cyst + cholelithiasis	Physical exam + lab; US + CT	30/12/1999	IVa	Cholecystectomy + Parsiyel cystectomy + T-tube drainage
12	60/F	Choledochal cyst	Physical exam + lab; US + CT + MRCP	12/06/2000	II	Cholecystectomy + cyst excision + T-tube drainage

MOBD, mechanical obstructive biliary disease; US, ultrasonography; CT, computed tomography; ERCP, endoscopic retrograde cholangiopancreatography; MRCP, magnetic resonance cholangiopancreatography

tomography (CT) were applied to nine of the patients, endoscopic retrograde cholangiopancreatography (ERCP) was used in five (Fig. 2), and magnetic resonance cholangiopancreatography (MRCP) was used in one. Demographic specifications, cyst types, and operations are depicted in Table 1. There was no mortality due to the operations. In a 46-year-old woman (patient 11 in Table 1), the biliary fistula that appeared after removal of the T-tube, closed spontaneously after endoscopic sphincterotomy.

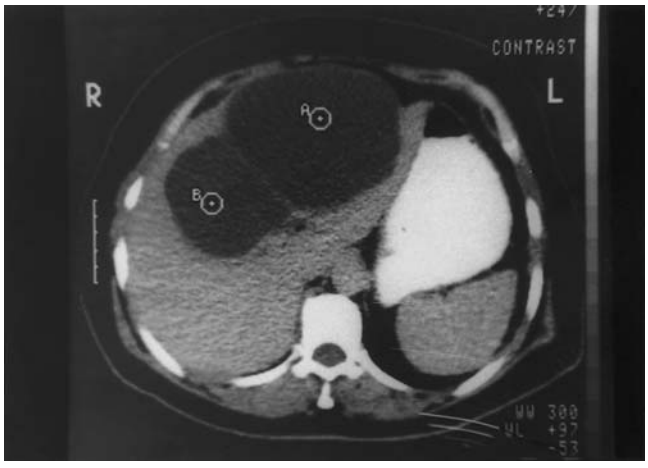


Fig. 3. CT image of a patient who had a giant choledochal cyst (type IVa) operated on with a diagnosis of liver hydatid cyst

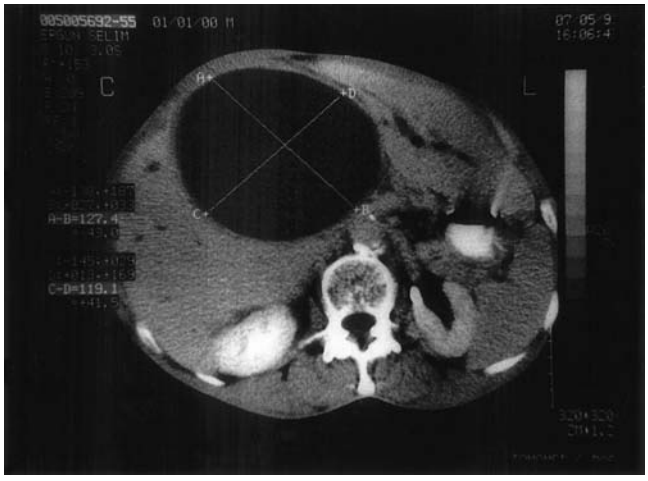


Fig. 4. CT image of a liver hydatid cyst

Postoperative follow-up was carried out regularly in six of the patients for 1–12 years. During this period one of the patients previously treated by cholecystectomy and choledochoduodenostomy had recurrent cholangitis 2 years after operation. Roux-Y hepaticojejunostomy was performed because of narrowing of the anastomosis. The remaining five patients are currently living disease-free.

Discussion

It is emphasized in the literature that type I cysts are mostly detected according to the Alonso-Lej classification. Todani refined the classification system of bile duct cysts by combining the Alonso-Lej classification and the variants of Caroli's disease.[5–8] In our series, 10 patients had type I cysts (83.3%), 1 had a type II cyst (8.3%), and one had a type IV cyst (8.3%).

The classic triad of jaundice, pain, and palpable mass was not seen in any of our cases. Five patients suffered from cholangitis symptoms, such as jaundice, fever, and pain. One patient with a giant cyst (Fig. 3) presented with the complaint of a painful mass in the upper right quadrant. In other patients who had ambiguous dyspeptic complaints, cysts were detected by US examination.

Prompt operation after the diagnosis was advised because of the possibility of hepatic failure, pancreatitis, biliary sludge, and stone formation.[9] During an era when US and CT had not been widely used diagnostically, these patients were diagnosed with only clinical and biochemichal findings and were operated on with a preoperative diagnosis of mechanical obstructive biliary disease. Their diagnoses were based on the presence of clinical and biochemical findings.

At present, US is the tool of choice for diagnosing biliary disorders. The communication with the biliary ductal system of the cyst must be demonstrated to diagnose a choledochal cyst. Depending on the type of choledochal cyst, the intrahepatic ducts may or may not be dilated, and other intrahepatic cysts may or may not be demonstrated. Therefore, when a choledochal cyst is demonstrated sonographically, cholangiography should be the next step. A choledochal cyst may be confused with several other cystic lesions, including hepatic cyst, enteric duplication cyst, pancreatic pseudocyst, hepatic artery aneurysm, and spontaneous perforation of the common bile duct. These entities can all be differentiated using a careful scanning approach and with the use of duplex and color Doppler imaging.[10]

In countries where hydatic disease is endemic, hepatobiliary cystic lesions are often misdiagnosed clinically and radiologically as hydatid cysts. The hydatid cyst in Fig. 4 has characteristics similar to those of the choledochal cyst in Fig. 1. Figure 5 shows the US findings of the same case as in Fig. 1. Because of economic problems, further radiological studies cannot always be done for each new patient with a cystic liver mass. Hence, three of our patients were operated on with a diagnosis of hydatid cyst.

There is only one article on this issue, published by Hira et al.[11] They emphasized that, apart from choledochal cysts, lesions such as simple liver cysts and pancreatic pseudocysts are often misdiagnosed as hydatid cysts in endemic areas. This confusion in diagnosis appears to be even more likely regarding giant cysts. Cholangiography is an indispensable diagnostic procedure, although endoscopic retrograde (ERCP) and magnetic imaging (MRCP) cholangio-pancreatography may be preferred.[12–14] Considering that percutaneous and laparoscopic approaches are increasingly being



Fig. 5. Ultrasonography shows a large cystic mass

used to treat hydatid cysts, problematic areas concerning cystic lesions must be subjected to thorough examination, and the possibility of choledochal cysts must always be kept in mind. Otherwise, an unfavorable process may begin for both the patient and surgeon.

Contemporary approaches to treating choledochal cysts focus on total removal of the cyst. It is advisable to remove the cyst, as it may develop into a cholangiocarcinoma.[7,8]

Our series might be criticized because serological tests were not done in patients who were misdiagnosed as having a hydatid cyst of the liver. The answer to such criticism is that negative results from serological tests do not exclude the possibility of a hydatid cyst.[15] Initial internal drainage operations for the cysts have been replaced by their total removal in our clinic. No neoplastic developments have so far been encountered during follow-up of our patients.

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