Cystic dilatation of common bile duct. Case report and review

Ann. Ital. Chir., LXXI, 1, 2000

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Abstract

The cystic dilatation of the common bile duct is a rare disease in Europe and in the USA; even rarer in adults. In fact less than 30% of cases are described in the over 20ies. There are some reports in the elderly. When observed in adults the diagnosis is usually due to the onset of symptoms of associated pathologies or to complications such as cholangitis or cancer. Ultrasound, CT and ERCP allow, in the majority of cases, an adequate pre-operative characterization of the lesion. Intra-operative cholangiography verifies the completeness of surgery – always resective – and excludes residual biliary pathologies. The treatment in patients who already underwent conservative surgery is more complex, in fact, in symptomatic patients a second observation and resection is mandatory while in asymptomatic patients a careful and complete follow-up is adequate without underestimating the cancer risk.

Septic and inflammatory complications, especially if chronic or recurring, can damage the liver; here resection or liver transplantion can be necessary.

The case here reported is a 24 year old woman with recurring cholangitis for at least 2 years in which US, CT and ERCP showed a Todani's type I cystic dilatation of the CBD. The total excision of the cyst with hepatic-jejunal anastomosis was carried out. A two-year follow up demonstrated no symptoms and normal laboratory findings; cholangioscintigraphy showed a good hepatic-biliary and anastomotic function.

Key words: Biliary duct cystic dilatation, choledochal cyst, hepatic-jejunal anastomosis.

Introduction

The biliary duct cystic dilatation (BDCD) can affect all the segments of the biliary tree, both intra and extra hepatic separately, or in association, up to a diffused involvement. The disease occurs in about one in every 2 million live births [1]; a greater incidence in oriental countries is reported, [2,3] more frequent in females than in males (sex ratio 3-4:1) [4]. Diagnosis is usually done in infancy or adolescence, in fact 25% of cases are diagnosed before 12 months old, and 60% before ten years [5, 6, 7]. The disease is very rarely seen in the elderly [8, 9]. This led to the hypothesis of the congenital nature of the disease. The etiopathogenesis can be related to a co-existent common pancreatico-biliary channel having a length of more than 1 cm. A high number of observations supported this theory [10]. In pediatric age, the typical symptomatology usually makes the diagnosis simpler. In adults the diagnosis is often delayed because of the atypical clinical picture [11]. The diagnosis in adults is therefore frequently made only due to the onset of complications or associated pathologies such as pancreatitis, biliary cirrhosis, portal hypertension, choleperitonitis (cystic rupture), malignant degeneration and, last but not least, lithiasis and recurring cholangitis [12, 13]. For these reasons an early diagnosis and the accurate pre-operative typing of the cystic pathology of the biliary ducts for a radical treatment are mandatory.

Case report

The patient *M. G.* female, 24 years old, unmarried, was admitted to the Department of Surgery, Unit II, University of Catania in November 1996 complaining of recurring abdominal cramps, associated with latent jaundice and fever for at least 2 years; nothing else was remarkable in her history. The hematochemical examinations showed increased transaminases, alkaline phosphatase, Gamma GT, and total and fractioned bilirubin. PT, PTT and AP were normal. US showed a large anechoic mass developing outside and under the liver, with high echogenic resonance formations inside; the

Pervenuto in Redazione il 22 Gennaio 1999

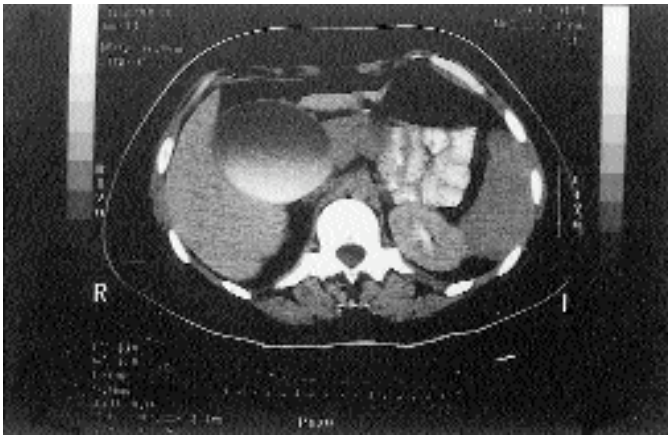

Fig. 1: Cholangio - CT.

liver showed an increased volume and an irregular echostructure. The gallbladder was tense and enlarged, with regular walls and with a lithiasic content. The size of the bile ducts at the hilum and the intrahepatic bile ducts were normal; the volume, morphology and echostructure of pancreas were normal too. ERCP showed an irregular duct of Wirsung with normal caliber and with a long common pancreatico-biliary channel (more than 1 cm). Echoendoscopy showed a mass in contact with the head of the pancreas and the CBD but was too large to establish its origin.

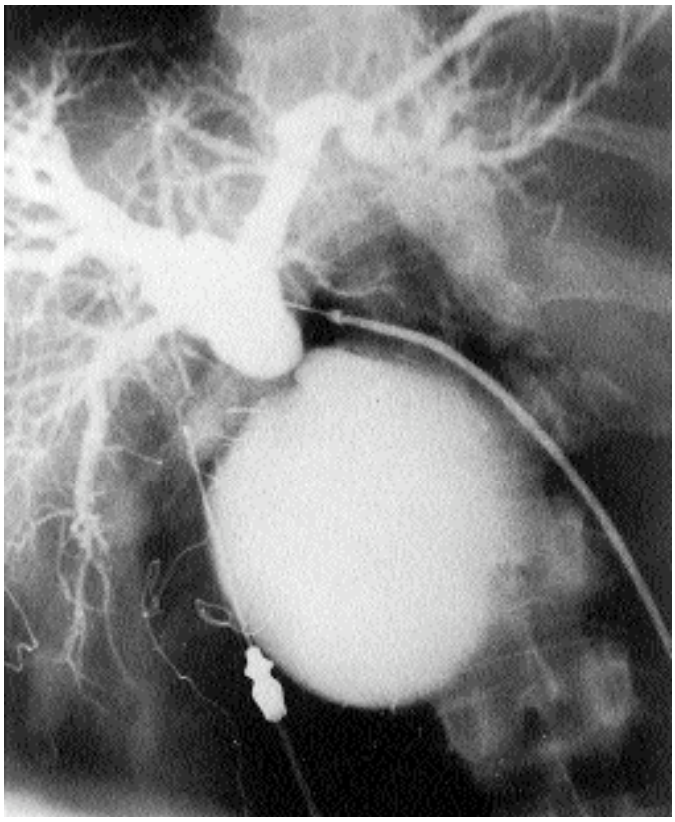

Fig. 2: Peroperative trans-cystic cholangiography.

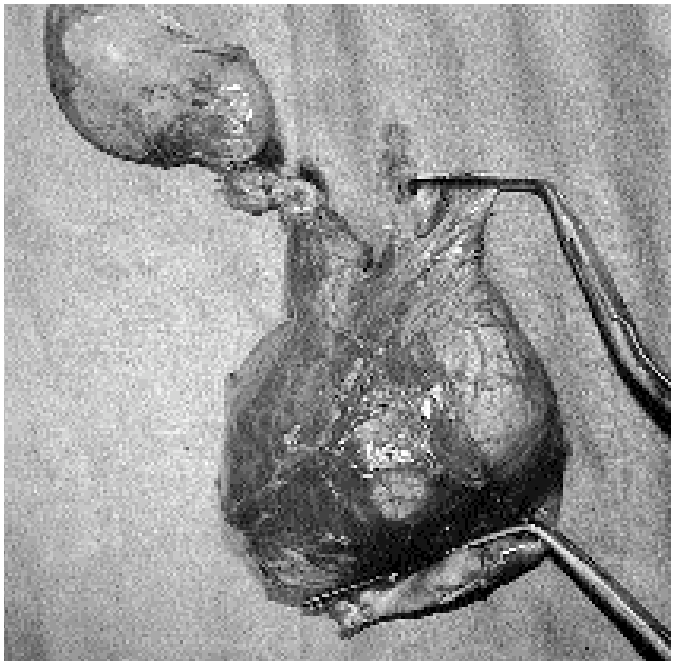

Fig. 3: Operative specimen.

MR with T1 and T2 image weighting by axial and sagittal scanning confirmed the presence of a round cystic structure under the liver – maximum width 70.7 mm – with a dense fluid content and small solid concretions. It was in contact with the head of the pancreas, the lower surface of the liver and, posteriorly, with the right kidney with which the cleavage plane was evident. The gallbladder was normal as were the body and the tail of the pancreas, where the head was not clearly seen.

The cholangio-CT confirmed the modest dilatation of the intrahepatic bile ducts and the progressive dilatation of the CBD, whose maximum equatorial diameter reached 7 cm, with small lithiasic concretions (Fig.1). The absence of dilatations of the intrahepatic bile ducts ruled out a percutaneous transhepatic cholangiography aimed at establishing the upper limits of the choledochal cystic dilatation.

A bilateral subcostal laparotomy consented an accurate exploration of the entire contents of the abdominal cavity and showed a voluminous dilatation (ø about 10 cm) of the CBD from the hepatic ducts to the supra pancreatic segment, dislocating the pancreas and the II and III duodenal portions anteriorly. Also the cystic duct appeared dilated while the gallbladder was partially intrahepatic and folded. After cholecystectomy the CBD was carefully isolated from the hepatic artery and the portal vein. A peroperative trans-cystic cholangiography showed a modest dilatation of the hepatic ducts, with regular

intrahepatic ducts and the presence of a common pancreatico-biliary channel of 1-cm length (Fig. 2). The resection was carried out from the confluence to the sovrapancreatic CBD after the placing of De Bakey's clamps above and below the bile duct dilatation. A retrocolic end-to-side hepatic-jejunostomy with 4/0 absorbable monofilament was performed. The distal stump of the CBD was isolated from the pancreatic tissue and closed with interrupted sutures. The enteric continuity was re-established by a side-to-side jejuno-jejunal anastomosis using GIA 60 and TA 55. A sub-hepatic Penrose's drainage was applied. The cystic dilatation (Fig. 2) was of 9 cm in diameter with a smooth surface, greenish in color and thickened (Fig. 3). Histologically the almost total absence of biliary mucousa and the hyperplastic phlogosis of the gallbladder were demonstrated.

Discussion

The first description of BDCD dates to the anatomical studies of Vater and Ezler [14] in 1723, while the first resection was in 1924, carried out by G.L. McWorter [15]. The first classification of Alonso-Ley were made in 1959 [16], whereas today the modifications proposed by Todani, 1977, are preferred [17]. He divides the cystic dilatation of the biliary ducts into five types: type I (dilatation of the common biliary duct), type II (diverticulum of the common biliary duct), type III (Choledochocele), type IV A (multiple intra and extra hepatic cysts), type IV B (multiple extra hepatic cysts), and type V or Caroli's disease (multiple or isolated intra-hepatic cysts). Type I is statistically the most frequent in various case studies in literature; our case is of this type. The etiological aspect of the disease is still not perfectly clear even if the congenital nature is certain, supported by the young age of patients, like the one we describe here, even if the onset of the symptomatology appears quite recent.

Numerous theories based on pancreatico-biliary alterations have been proposed. Babbit and Coll. in 1969 [18] believed the disease to be related to a high insertion of biliary duct in the pancreatic duct with a long common pancreatico-biliary duct. This observation has, in fact, been reported in 36-90% of cases [19]. This theory is, moreover, confirmed by the absence of a sphincterial anti-reflux activity of the choledoch [20] and by the finding of high concentrations of amylase in the cystic fluid [21]. Kusunochi [22] which strengthen the above mentioned hypothesis postulating a primitive neuronal dysfunction, with a reduced number of ganglia cells in the cystic wall. Today less credit is given to the theory of congenital disorders associated with distal obstruction [16] and that of anomalous choledochal rechannelisation during embryonic development [23].

Genetic anomalies have not been demonstrated, although some observations have been recorded in members of the same family [24].The clinical picture shows the classic triad of jaundice, palpable abdominal mass and pain in not more than a third of the cases studied [25]. Jaundice appears more frequently in newborns and adolescents, about 80% of observed cases, while in adults there is prevalently a symptomatology of abdominal pain [25]. Congenital cysts should be considered in the differential diagnosis of recurrent abdominal pain, especially in young female patients [26]. Stenosis, rupture and portal hypertension globally occur in 25% of cases with cystic pathology of choledoch [27]. Cancer is seen with a frequency closely linked to age at diagnosis, with a maximum incidence of 28.3%, 20 times greater than in the general population [28]. The forms more at risk seem to be type I and IV according to Todani. The site of origin, in 60% of cases, is the cyst wall. However, the carcinoma (of which adenocarcinoma is the most common histological finding [29]), can also arise in the intra hepatic ducts and in the gallbladder [30-33]. The most accredited etiopathogenetic theory holds that the common channel as responsible for the degeneration through the continuing chronic inflammation due to biliary stasis and pancreatico-biliary reflux [29, 30]. The aspecific symptomatology leads, in almost all cases, to malignant degeneration when eradication is no longer suitable and the average survival does not exceed 8-9 months [34].

The diagnosis of cystic dilatation of the main bile duct is confirmed by US, which shows the localization, shape and dimensions of the dilatation. The method also allows the prenatal diagnosis from the second three months of pregnancy [35]. Scintigraphy, with Tc99m HIDA, is useful in functional investigations and is important in follow-up [36]. The role of CT in the diagnostic protocol has not found agreement in literature. Both Katyal and Lees, and Rha and Coll [25, 37] do not cite CT among the methods of imaging to be used preoperatively, while in the work by Mai and Coll [38] it seems to have a unquestionable role. In our case cholangio-CT has had a primary role in the definition of the relationships and of the nature of the cysts, which were not clearly identifiable using ultrasound and MR.

ERCP showed the existence of a long common pancreatico-biliary channel with irregular duct of Wirsung and a marked dilatation of the main bile duct, confirming the reliability of the technique whose sensitivity is almost 100% [39]. There were no indications for a percutaneous transhepatic cholangiography in the absence of dilatation of the intrahepatic biliary tree. Instead we carried out peroperative cholangiography to show an eventual associated biliary pathology. We thus carried out an intraoperatory hepatic biopsy that showed a liver that was still healthy. Recently [25, 34] liver biopsy has been proposed to evaluate a initial concomitant hepatic cirrhosis or fibrosis, which is often present in patients with cystic pathologies of the common bile duct [38].

Management of choledochal cyst actually requires the

complete surgical excision while drainage procedures are no longer used, as these have been associated with a high incidence of complications. In most series Roux-en-Y hepaticojejunostomy is the treatment of choice [37]. The valved jejunal conduit interposed between the common hepatic duct and duodenum is described as another effective procedure [37, 40-42].
The therapeutic strategy does not appear completely definite as regards the patient who has already undergone internal drainage procedures and lives with the risk of developing carcinomas that cannot be radically removed. For these patients it is believed advisable to re-operate for exeresis , above all when there is recurring cholangitis. Exeresis greatly reduces the risk of malignant degeneration but does not eliminate it; in fact 40% of cancers develop elsewhere, other than in the cystic dilatation. The treatment of asymptomatic adults with entero-cystic anastomosis is still doubtful though they must have an accurate clinical-laboratory-instrumental surveillance [25]. In the absence of standardized protocols of follow-up, we examined the patient after 6, 12 and 24 months from surgery, using clinical and echographic methods. Cholangioscintigraphy was carried out at 60 days, one year and two years from surgery. We found no episodes of cholangitis or biliary pain. Hepatic functionality was within the norm and the scintigraphic and echographic methods showed a normal biliary flux without anastomotic stenosis.

Conclusions

In conclusion, after a careful revision of literature, we believe that greater attention should be given to the observation of this uncommon adult pathology, as a diagnostic delay, incorrect diagnosis or treatment can lead to problems that are not easily treatable. In fact, some severe complications, such as multiple lithiasis and the cystic dilatation of the intrahepatic biliary tree with marked segmental parenchymal atrophy can necessitate a hepatic resection, while, for patients with severe secondary hepatobiliary insufficiency, organ transplant may be the only solution [43].

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Commento Commentary

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Il caso clinico qui descritto ci offre l'opportunità di rivedere un raro ma insidioso aspetto della chirurgia biliare.
Gli aspetti clinici e le metodiche diagnostiche illustrate dagli AA sono elementi di grande importanza così come la radicalità dell'approccio chirurgico. Suggestiva ed interessante è l'iconografia presentata.
Allo stato attuale il semplice drenaggio interno della cisti senza asportazione della stessa non trova più indicazione a causa della elevata incidenza di colangiti e del rischio di cancerizzazione, per queste ragioni la diagnosi deve essere raggiunta preoperatoriamente.
In particolare va data grande importanza alla associazione di dolore addominale ed ittero, specialmente in giovani donne.
I1 caso presentato da Cappellani e Coll. rappresenta un buon esempio di corretto approccio diagnostico-terapeutico.

The cases here reported represents a good occasion for a review of a rare but deceitful aspect of the biliary surgery.
The clinical aspect and the diagnostic procedures illustrated by the authors are elements of high importance as well as radical surgical treatment.
Suggestive and interesting are figures presented by Cappellani e Coll.

The simple internal drainage of the cyst without excision is not recommended because of high incidence of recurring cholangitis and cancer. For this reason the diagnosis must be done preoperatively, giving high importance to the addominal pain and jaundice specially in young female patients.
The case reported by the authors is a good example of correct management of this disease.

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