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SURGICAL MANAGEMENT OF BILIARY TREE CYSTS: A CASE SERIES

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ABSTRACT

Background:

Biliary tree cysts represent a genetic and pathological dilatation, focal or extended, of hepatobiliary system. Incidence in western populations is around 1/100.000-1/150.000 and is more frequent in Asiatic areas (1/1.000). Todani classification is based on site and morphology of cysts. Acute cholangitis is the main clinical presentation, cancer incidence is around 2,5-16%.

Material and methods:

From 2008 to 2018 we observed and surgically treated 6 cases.Clinical presentation was characterized by abdominal pain, jaundice, fever and sometimes weight loss. Five patients with involvement of intrahepatic tree and one patient with involvement of extrahepatic biliary duct.

Results:

All patients were surgically treated. We performed five hepatic resection and one resection of biliary tree reconstructed with bilio-digestive anastomoses sec. Roux. 30-days complications were 12,5%. 90-days mortality was 0%. Histology confirmed in a patient diagnosis of cholangiocarcinoma on Caroli disease. After two years of follow-up: 7 patients are alive and in good conditions, patient with cholangiocarcinoma died for disease progression after 12 months from surgery.

Conclusions:

In case of unilobar and symptomatic disease surgical resection is treatment of choice. Liver transplant is indicated in bilobar disease. Extrahepatic disease is treated with resection of biliary tree and reconstruction. It's important to treat these rare diseases in order to avoid risk of malignant developement.

INTRODUCTION

Biliary tree cysts represent a pathological dilatation, focal or extended, of hepatobiliary system. These rare congenital cystic malformation of bile ducts were first described in 1723 by Vater and Ezler[1].

Todani and collaborators created first in 1977 the classification that today is still universally accepted and divided morphology of malformations in five types [2]. Type I refers to a cystic dilatation of entire common bile duct, type 2 represents diverticulum of extrahepatic biliary tree, type 3 is choledococele and type 4 involves intrahepatic and extrahepatic biliary tree with multiple cysts.

Type 5 of Todani classification was first described by French gastroenterologist, Jaques Caroli, in 1958 and it is an isolated intrahepatic biliary cystic disease [3]. If it is associated with periportal fibrosis it is called Caroli syndrome [4].

Incidence in western populations is around 1/100.000-1/150.000 and is more frequent in Asiatic areas (1/1.000). These diseases less commonly affect men, with a female to male ratio of 4:1.

Clinical presentation includes abdominal pain in right upper quadrant hypochondrium, infections like cholangitis and acute pancreatitis, jaundice [5,6,7,8]. Hepatic abscess and sepsis are possible complications; portal hypertension occurs in Caroli syndrome.

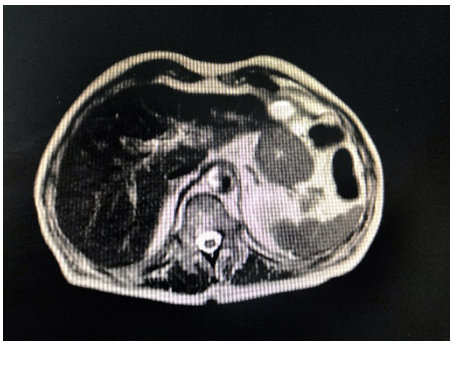
Ultrasound is the cheapest, less invasive and fast imaging we can use to visualize morphology of biliary tree. Tough more expensive imaging of choice is considered MRI and it gives precise patterns about disease extent and severity grade [9].

We describe five cases of Caroli disease and one case of Todani type 3-4 misdiagnosed several times for recurrent cholangitis episodes and abdominal pain. Our intent is to report these cases of rare diseases in order to highlight difficulties in diagnosis and to potentially prevent malignant evolution [10].

CASE SERIES

Case 1:

In August 2010 a female patient of 68 years old was accepted in our Unit for abdominal pain, increase of cholestatic indexes. It was performed a CT scan and images showed isolated dilatation of biliary tree of second and third segments (figure1). In her anamnesis she had recurrent episodes of cholangitis with hospital admission. In order to avoid the risk of a sever septic complication it was decided with gastroenterologist to switch to surgical approach. She underwent to a resection of S3 and consensual cholecystectomy with open technique. Post-operative course was uneventful.



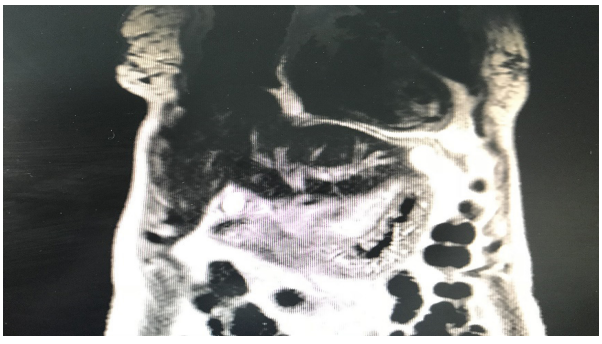


Figure 1

Case 2:

In June 2016 a 63 years old man was admitted to our unit to underwent to elective left hepatectomy for hepatic congenital cystic disease. In his clinical history it was reported recurrent episodes of acute biliary pancreatitis starting thirty years ago followed by cholecystectomy. Later, he was admitted several times in hospital for cholangitis episodes caused by lithiasis of biliary ducts treated with ERCP. Last CT scan showed atrophic left liver associated to dilatation of consensual biliary tree (Figure 2). Before moving to surgical treatment, a biopsy was made but it was not diagnostic, and it was positioned a stent for choledocholithiasis. In a multidisciplinary context it was decided to move to surgical approach and the patient underwent to a left hepatectomy. During surgery a biopsy of fibrotic tissue close to biliary tree was made and pathology report excluded malignant degeneration. Post-operative course was uneventful.

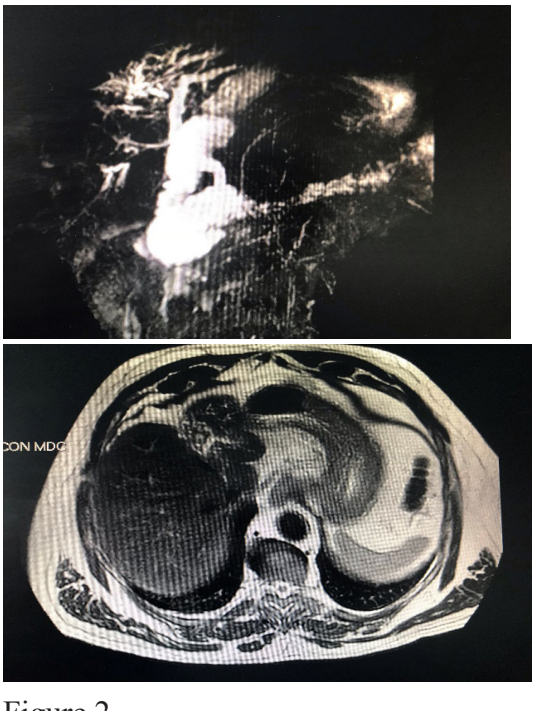


Figure 2

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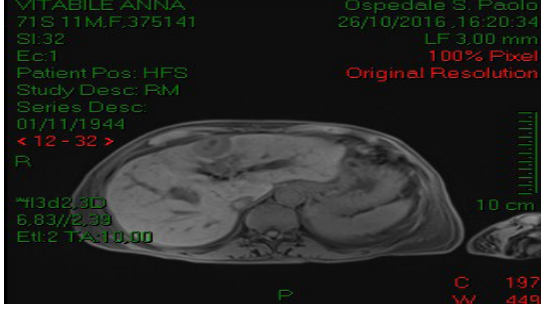
Case 3:

In October 2016 a woman of 69 years old was admitted for weight loss, asthenia, abdominal pain episodes, fever and anaemia. We reported in her clinical history a previous cholecystectomy and bilio-digestive anastomosis performed for cholangitis 30 years before. She hadn't important comorbidities. Us images showed in epigastric region a lesion of 9x6 cm L-L. Blood tests were characterized by leucocytosis and low haemoglobin. A CT scan was made. Images showed irregular mass with colliquative areas 10x5 cm A-P involving predominantly left liver and cranially extended to the stomach, that was probably involved (Figure 3). Left biliary tree was dilatated.

Cholangiocarcinoma markers were negative. Case was multidisciplinary discussed, and a US guided biopsy was performed. Histological findings revealed that lesion was a probably a rare case of liver fibrosarcoma. In multidisciplinary context we decided to move to explorative laparoscopy as diagnostic tool to exclude peritoneal carcinosis or visceral infiltration and to establish the feasibility of a surgical procedure with radical intent. Abdomen were free from distant lesions and stomach were cleavable from hepatic mass, we converted to open surgery and performed a left hepatectomy. Postoperative course was characterized by an intraabdominal collection treated in conservative way. Patient was discharged after 20 days from surgery.



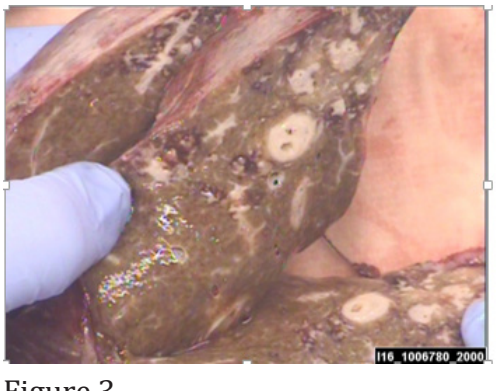


Figure 3

Case 4:

In January 2018 a male patient of 53 years old was admitted in ER for epigastric pain. Anamnesis was characterized by previous abdominal-thoracic surgery for trauma, numerous hospital admissions for biliary cholic and he has patent foramen ovale and meningioma. At physical examination abdomen was tender and painful. Blood test were negative. Abdominal US showed dilatation of left biliary tree and suspect of endoluminal lithiasis, gall bladder was inflamed and containing stones. He was admitted in our unit for diagnostic investigations. We initially treated him with medical therapy and planned a colangioMRI. MRI images showed a focal ectasia of S3 bile duct characterized by multiple endoluminal filling defects. A spherical neoformation was localized at distal portion of S3 biliary duct (Figure4). Gall bladder was inflamed, distended and lithiasic. Radiologist hypnotize first inflammatory process complicated by abscess. He underwent to laparoscopic exploration and it was consensually performed a resection of S3 and cholecystectomy. Initially post-operatory course was uneventful. However, the patient was readmitted for fever due to a hepatic abscess close to surgical margin, treated by percutaneous drainage. He was discharged after 15 days from second admission.

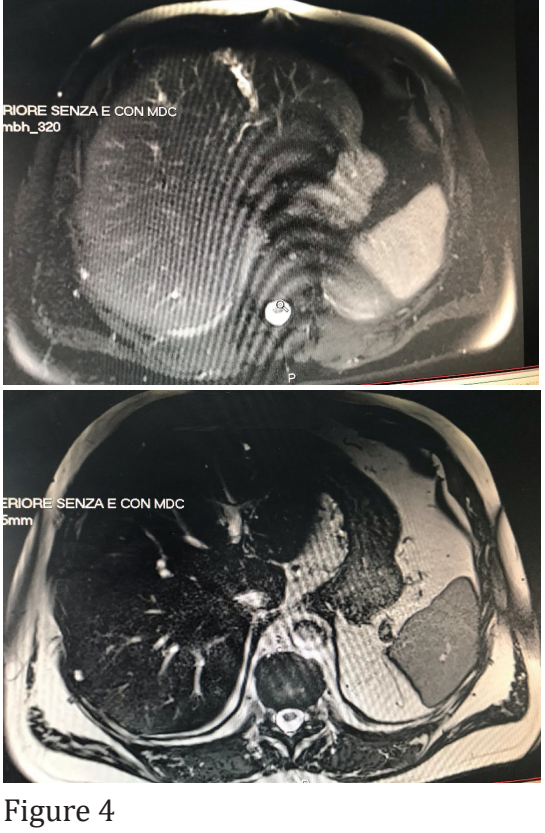


Figure 4

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Case 5:

In March 2013 a Chinese woman come to ER unit for right hypochondrium pain and weight loss. Blood tests showed high levels of CA 19,9, d-dimer, ALP and GGT. Her clinical history was characterized by a cholecystectomy fifteen years ago and several episodes of abdominal pain in right quadrants. At US left liver was disomogenous with biliary ducts dilatated. CT scan was performed: left biliary tree was ectasic and associated to an irregular lesion suspected for cholangiocarcinoma. MRI showed left portal branch infiltrated (Figure 6). We discussed the case in multidisciplinary reunion and move to explorative laparotomy. Patient underwent radical left hepatectomy, all intraoperative biopsies on nodes were negative for malignant infiltration. Post-operative course was uneventful. Histological findings revealed cholangiocarcinoma (CCA) on Caroli disease (Figure 7). Patient died after 12 months from surgery for disease progression.

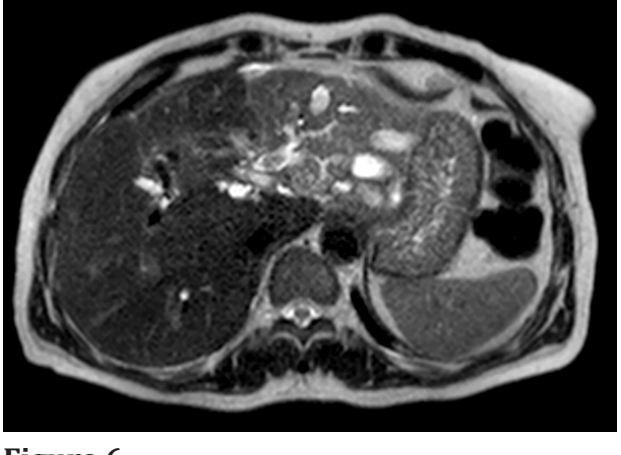

Figure 6

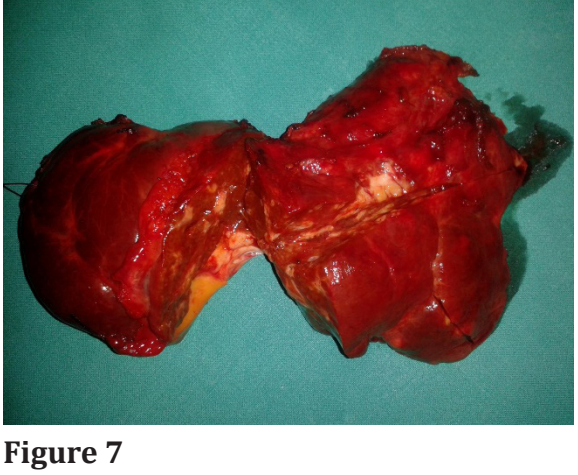

Figure 7

Case 6:

In 2016 we admitted 30 years old woman for abdominal pain localized in right hypochondrium. US images showed an anomalous anatomy of biliary tree. CT scan and cholangioMRI were performed to complete diagnosis. We find out that it was present a diverticulum of extrahepatic biliary tree (Todani disease type II) associated with choledococele (Todani disease type III) (Figure 8). Because of the young age, good performance status and symptomatic disease impacting her quality of life it was decided in multidisciplinary context to move to surgical approach. She underwent to a laparoscopic resection of extrahepatic biliary tree with bilio-digestive anastomosis sec. Roux. Post-operative course was uneventful.

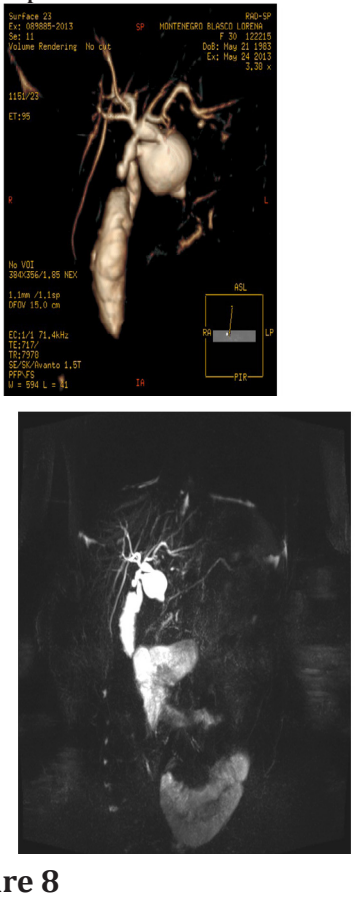

Figure 8

RESULTS

Ninety days mortality rate was 0%. Complication rates were 12,5% (Clavien-Dindo grade III). Histological findings of all patients revealed Caroli disease. Case 6 patient was characterized by a diagnosis of cholangiocarcinoma (CCA) on Caroli disease and she died after 12 months from surgery for disease progression. The rest of patients are in good conditions, alive and free from hepatobiliary septic complication or cancer.

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DISCUSSION

Etiopathogenesis of biliary tree cysts is still an open discussion: different causes may play a synergic role. There are different theories that involving embryology and multy-symptomatic disorders.

Babbitt's theory was one of the first accepted in literature: cystic malformations were connected to an anomaly of the biliopancreatic duct union (APBDU) [11, 12]. This alteration is seen in 30% to 70% of all CC and represents a congenital malformation, in which the pancreatic and bile ducts join anatomically outside the duodenal wall, forming a markedly long common channel.

The sphincter of Oddi located in the duodenal wall, allowing reflux of pancreatic secretions into the biliary tract and exposing epithelium to digestive and irritant action of juices. This pathologic mechanism leads to citostructure changes (progression to CCA) and higher probability of malignant degeneration. Komi et al describe nine types of APBDU.

However, it's evident that this theory doesn't justify the Caroli disease because APBDU doesn't involve intrahepatic biliary tree.

A new etiologic theory came out to explain Caroli disease: the ductal plate malformation [9]. The development of the biliary tree is a multistep process that ends in the formation of a system of intrahepatic and extrahepatic bile ducts. The ductal plate is double layers tubular structure of epitelium of biliary ducts surrounding portal vein radicals which undergoes remodelling into the intrahepatic biliary tree. Remodelling process of these ductal plates occurs during the 12th embryonic week to form bile canaliculi of various sizes beginning from the hilum to the periphery of the liver. When remodelling errors of ductal plate occur early, they lead to dilatation of lobar or segmental ducts (Caroli disease), later they lead to dilatation of interlobular ducts associated with periportal fibrosis (Caroli syndrome) up to dilatation of the smallest bile ductulus (biliary hamartoma or Von Mayemburg complexes).

Exploring pathogenesis of biliary tree cystic alterations, we have to discuss about the polycystic diseases. This group of multy-symptomatic diseases involves polycystin proteins, that intermediate cell proliferations and adhesion in ciliated cells within the liver and the kidney. According to the genetic mutation we have different diseases. Mutation of protein kinase Sec63 causes autosomal dominant polycystic liver disease (ADPLD) that it's rare, asymptomatic and typical of adult age. Renal and hepatic synchronous involvement is typical of autosomal dominant polycystic kidney disease (ADPKD) and autosomal recessive polycystic kidney disease (ARPKD) and it's caused respectively by policystin 1, policystin 2 and fibrocystin [13, 14].

In these last ten years more importance has been attributed to genetical alterations amenable to ciliopathies. Satir et al. [15] introduced the concept of ciliopathies: a group of disorders associated with genetic mutations which result in abnormal formation or function of motile/no motile cilia. This concept represents now an expanding disease spectrum that could explain majority of multisymptomatic disease because cilia are a component of almost all vertebrate cells.

Cholangiocytes express primary cilia extending from their membranes, which detect stimuli from bile. The information received from these cilia is traduced into several cellular signalling pathways involved in secretion, proliferation and apoptosis. Detention of morphological and/or functional defects of these organelles leads to pathological changes like cholangiocyte hyperproliferation, altered fluid secretion and absorption. Cilia dysfunction has been observed in epithelial tumours, including cholangiocarcinoma (CCA), in addition, the loss of cilia is associated with dysregulation of cellular communication process resulting in CCA development and progression [16, 17].

The main risk of biliary ducts cystic diseases is malignant evolution. The incidence of CCA and gallbladder cancer in these patients is respectevely 2,5-16% and 10-25%, CCA occurs more in type I and type IV Todani disease [18]. The presence of an abnormal cystic dilatation of biliary duct is thought to play a role in carcinogenesis and hepatocellular damage due to reflux of pancreatic contents into the bile duct. The risk of cholangiocarcinoma development has been reported to be as high as one hundred-fold greater in patients with biliary tree cysts than in the general population [19, 20].

Surgery decision making factors should be summed firstly in evaluation of localisation and extension of the disease, in literature surgical treatment is indicated only for symptomatic cases. Surgery should be tailored according to disease extension: in bilobar cases is indicated hepatic transplant and in unilobar cases is indicated surgical resection [21]. Biliary tree cysts have to be treated as neoplasia and resected with free parenchymal margin in order to avoid malignant degeneration. The indication to operate is due as well to the higher risk of symptomatic patients to undergo a septic complication, in addition we don't' have to forget that chronic liver disease could cause portal hypertension.

CONCLUSIONS

Biliary tree cysts are a group of rare diseases which pathogenesis is still an open discussion. However, surgeons don't have to underestimate cystic alteration of biliary ducts because these diseases could evolve to malignancy. Multidisciplinary discussion with radiologist, medical oncologist and gastroenterologist is mandatory for these patients, especially when they are symptomatic. Surgery is indicated in patients with symptoms and should be planified on disease extension; transplant should be considered in patient with bilobar disease. In the future with new discovering in molecular biology more explanation could come out to light origins of this diseases and probably a consulting for genes mutation should be suggested to patients with these alterations. The real incidence of Todani and Caroli disease is missing, we are promoting an Italian Survey on behalf of Italian Society of Surgical Oncology (ESSO affiliated) to avoid this lack.

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