CONGENITAL CYSTIC DILATATIONS OF THE BILE DUCTS: ATTEMPT FOR MODIFICATION IN CLASSIFICATION

Letter to the Editor

INTRODUCTION

Congenital cystic dilatations of the bile duct are infrequent affecting the intrahepatic bile duct as much as the extrahepatic or both at the same time. Vater was who first described a congenital dilatation of the extrahepatic bile duct, but was Douglas' description in 1852 that gave full knowledge about this rare affection. On the other hand, Jacques Caroli described in 1958 the congenital cystic dilatation of the intrahepatic bile duct as an infrequent cause of intrahepatic cholestasis. These dilatations are characterized by their polymorphism as the dilatations can affect any part of the bile duct. The rareness of this pathology has caused the majority of the studies to be clinical case reports and the importance of its knowledge lies in the fact that the treatment can be wrong.

The aim of the present communication is to present eight cases and revise the literature with the intention to evaluate if there have been changes in the therapeutic suggestions in recent years and suggest the aggregation of two subtypes in the classic classification.

METHODS

In a descriptive study of a series of cases data was collected from the patients with congenital cystic dilatations of the bile duct between 1997 up to the present (23 years). For its classification we used the classification proposed by Alonso Lej with Todanis' modification. To classify them were used cholangiographic in pre, intra and postoperative associated to the results of the pathologic anatomy in the case of resection. Also associated with diagnostic confirmation were the indirect signs that may accompany them, such as thinning of the dilatation wall, hypoplastic gallbladder and increased duodenal-pancreatic vascular aspect, among others. The presence of the anomaly proposed by Babbit and later re-classified by the French Association of Surgery based on that of Komi et al. was investigated (Figure 1). In patients with Caroli disease, it was specified if it was diffuse (bilobar) or unilobar; if the dilation was saccular or tubular; and if there was intrahepatic lithiasis. Caroli syndrome was considered when there was an association of congenital liver fibrosis and/or polycystic kidney disease.

CASES REPORT

In Table 1 the principal data is presented of eight patients, three with Caroli disease and the other five with choledochal cysts, four were of type I and one was of type VI A. In two patients with choledochal cysts type I in which the bile duct was resected, there were anatomic changes associated to this pathology, in both there was hypoplastic gallbladder and increased duodenal-pancreatic vascular aspect. In a patient with a type 1 cyst and in the type one VI A the existence of an anomalous pancreaticobiliary junction could be objectified. As the recollection of patients was carried out in 23 years there exist different study modalities between them. In two surgical treatment was performed without preoperative
knowledge of the correct diagnosis and these patients were initially approached to resolve lithiasic pathology of the bile duct. The imaging methods were of very little help in the preoperative of the patient with the choledochal cyst type VI A that operated initially approached to resolve lithiasic pathology of the bile duct.


discussion

Congenital cystic dilations of the bile duct in adults are rare, for Valayer cystic dilation of the extrahepatic bile duct (CHD) would occur in 1 out of 2 million births\(^4\); other western authors report incidence of 1 in 100,000 to 190,000 live births\(^4\); the incidence in the East would be much higher, being reported as 1 in 1,000 births in some Asian populations\(^14\). The female gender is more affected in the cystic dilation of the extrahepatic bile duct with a 4:1 relation, while in CD the distribution by gender is equal\(^5\). The majority of patients with cystic dilation of the extrahepatic bile duct are presented in childhood\(^5\). Yamaguchi\(^15\) on 1433 reports that approximately the 51% of patients were in the first decade of life and this number jumped to almost 70% in the first two decades of life, being much less frequent in adult. In comparison CD is usually diagnosed in adult.

Alonso Lej et al.\(^1\) made the most utilized classification and later on suffered various modifications being Flanagan’s in 1975\(^6\) and Todani’s in 1971\(^11\) the most representatives. The last one is more used and has the particularity of having incorporated, as type V, the Caroli disease. In the year 2001, we proposed a subdivision of type V in two subtypes in relation to whether it was CD or CS\(^16\). Today, we propose a subdivision in three types I choledochal cyst type I - are the more frequent followed by type IV, that coincides with our patients\(^17\). The last one is more affected in the cystic dilation of the extrahepatic bile duct with a 4:1 relation, while in CD the distribution by gender is equal\(^5\). The majority of patients with cystic dilation of the extrahepatic bile duct are presented in childhood\(^5\). Yamaguchi\(^15\) on 1433 reports that approximately the 51% of patients were in the first decade of life and this number jumped to almost 70% in the first two decades of life, being much less frequent in adult. In comparison CD is usually diagnosed in adult.

Regarding the etiology, some differences are recommended not to talk about "cystic dilation of the extrahepatic bile duct", including them in what is called “the fibropolycystic family of liver disease”, that is, a constellation that includes other
Babbit\footnote{9} proposed an anomaly in the biliopancreatic junction with two variations and with a common duct between the choledochus and the Wirsung duct longer than 10 mm round the outside of the duodenal wall. Later, other authors\cite{7,11} add a third more complex variety in “anse de seau” (Figure 1) and it is the most common etiological theory with a frequency of up to 90% and that justifies basically the cystic dilation of the extrahepatic bile duct type I and IV\cite{4,15}. This theory justifies the reflux of pancreatic liquid inside the biliary tract with the consequent digestion and inflammation with later dilatation that would justify the values of amylase superior to 10000 U/L that is usually found in the bile of the dilatations\cite{8}. Both CD and the cystic dilation of the extrahepatic bile duct type II and III have their etiology in recessive autosomal disorders that would also justify the presence of polycystic kidney disease and congenital fibrosis in CS\cite{5,19,22}.

Regarding the clinical presentation, the classic triad of abdominal pain, jaundice and palpable tumor in cystic dilation of the extrahepatic bile duct is not present in all patients. We can say that these symptoms can occur associated with others such as fever, choluria or the presence of lithiasis.

Our series is 23 years old and some patients were insufficiently studied. The MRCP is undoubtedly an element of great value to reach a diagnosis, while during surgery, the procedure devised by Mirizzi will also be of great help in making therapeutic decisions. The percutaneous or endoscopic approaches will not only be able to collaborate with the diagnosis, but also have the addition of being able to drain the bile duct in patients with cholangitis or hepato-cellular disorders due to cholestasis.

One of the most important points is the malignization of the dilatation, mainly in those of type I and IV, between 2.5 % and 17.5% are reported\cite{3,4,8,10,17,18,19}. Bruguera et al\footnote{5} refer, that the possibility of malignancy in CD would range between 7-14%. Benjamin\footnote{4} reports that biliary stasis favors the formation of secondary bile acids that would have mutagenic power. In our series we only had one patient with cancer, and it was from the gallbladder, the patient had an abnormality of the biliopancreatic junction and this anomaly has also been associated with the genesis of gallbladder cancer. The increased risk of developing a cholangiocarcinoma makes surgical treatment one of the first
treatment options.

In what refers to the cystic dilatation of the extrahepatic bile duct, mainly in the two most frequent types, resection of the cystic bag with the performance of a Roux-en-Y hepaticojejunostomy is the treatment of choice, although some authors suggest that performing a hepaticoduodenostomy would have the same results. We believe that the latter favors reflux episodes of postoperative cholangitis. We have resected the dilation in three patients with good subsequent evolution, and in two we performed insufficient treatments, one of them with a multiple pathology of the bile duct that included cancer of the gallbladder and the other in which we arrived at the diagnosis postoperatively and were refused the resection. In regard to types II and III dilatations, choledochocole may be a recurrent cause of pancreatitis and this is the main indication for resection, whereas in the choledochocole diverticulum pain or its complications would be the reasons for the resection.

Regarding type V dilatations, we propose subdivision as: Va, segmental or lobar; Vb, diffuse CD; and Vc, CS with fibrosis. This classification, in our view, is because there are different therapeutic approaches in relation to each subtype. Va type requires resectional treatment of the affected part with good results in terms of symptom control. Vb type can be managed with medical treatment (ursodeoxycholic acid, antibiotics, etc.) and endoscopic papillotomy and in cases where control of symptoms is difficult, liver transplantation will be a weighted option. Vc type, which is associated with liver fibrosis, has indication to liver transplant, having life expectancy up to five years in 72.4%.

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