Adult's congenital bile duct cysts

Les dilatations kystiques congénitales des voies biliaires diagnostiquées chez l'adulte

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RÉSUMÉ

Introduction : La dilatation kystique congénitale des voies biliaires (CBDC) est une malformation congénitale rare. Elle résulte d'une anomalie de la jonction bilio-pancréatique (AJBP). Cette affection est diagnostiquée souvent à un jeune âge malgré qu'elle peut rester asymptomatique et de découverte qu'à un âge avancé. Le but de notre travail était d'étudier les aspects diagnostiques, thérapeutiques et évolutifs des CBDC à travers une série de 11 cas diagnostiqués chez des patients adultes.

Méthodes: Il s'agissait une étude rétrospective, monocentrique ayant colligé les patients opérés pour CBDC entre le 01/08/1999 et le 30/06/2009. Nous avons réalisé une étude descriptive analytique.

Résultats : L'âge moyen était de 45,3 ans. Deux hommes et neuf femmes. La douleur de l'hypochondre droit a été rapportée par tous les patients. A l'examen physique, un ictère a été noté dans cinq cas et le reste de l'examen était normal chez tous les patients. La biologie a objectivé une cholestase dans six cas et une cytolyse chez quatre patients. Un seul patient a présenté une hyper-amylasémie (cinq fois la normale).Le diagnostic préopératoire d'une CBDC a été évoqué dans huit cas. La cholangiographie peropératoire (CPO) a permis de poser le diagnostic positif et de classer les CBDC selon la classification de Todani. L'examen anatomopathologique a objectivé trois cas d'adénocarcinome vésiculaire associés. Les suites opératoires étaient simples chez dix malades. Un cas de reprise chirurgicale était nécessaire chez une malade pour pancréatite postopératoire nécrotique et infectée. Le recul moyen était de 40,6 mois avec des extrêmes allant d'un mois à 7 ans. Un seul cas de décès a été constaté, à un mois postopératoire. Aucun cas de dégénérescence à distance n'a été objectivé à nos jours avec un seul cas de cirrhose biliaire secondaire due à des poussées répétées d'angiocholite.

Conclusion: la CBDC est rare, Il faut y penser même à un âge avancé vue le risque accru de dégénérescence qui augmente avec l'âge et son pronostic péjoratif. Les examens radiologiques permettent de suspecter le diagnostic chez des patients jeunes et asymptomatiques pour assurer un traitement chirurgical bien conduit et à temps.

Mots-clés
Voies biliaires, dilatation, voie biliaire principale

SUMMARY

Introduction: Congenital bile duct cysts (CBDC) is a rare congenital malformation. It results from an anomaly of the biliopancreatic junction (AJBP). This condition is often diagnosed at a young age. Although, it can be asymptomatic and discovered only at an advanced age. The aim of our work was to describe the diagnosis, therapeutic and evolutionary aspects of BVCD through a series of 11 cases diagnosed in adult cases.

Methods: This is a descriptive, retrospective and monocentric study. It collects patients operated for CBDC between 01/08/1999 and 30/06/2009.

Results: The mean age was 45.3 years. Two men and nine women. The right hypochondria pain has been reported by all patients. On physical examination, jaundice was noted in five cases and the rest of the examination was normal. Biology showed cholestasis in six cases and cytolysis in four cases. Only one patient had hyperamylasaemia (five times normal). The preoperative diagnosis of a VBCD was reported in eight cases. Peroperative cholangiography (OCP) allowed to make the diagnosis and classify the CBDC according to the classification of Todani. Microscopic examination demonstrated three cases of associated gallbladder adenocarcinoma. The operative follow-up was simple for ten cases. A case of surgical recovery was necessary due to infection of necrotic pancreatitis. The mean follow-up was 40.6 months with extremes from one month to seven years. Only one case of death has been reported. No patient has presented a later biliary tract degeneration. Only one case of secondary biliary cirrhosis due to repeated attacks of angiocholitis has been reported.

Conclusion: CBDC is rare. It must be suspected at any age. It is characterized by the increased risk of degeneration. Radiological examinations can suspect the diagnosis in younger and asymptomatic patients to ensure a well-conducted and timely surgical treatment.

Key-words
Biliary, Dilatation, Common Bile Duct

Congenital bile duct cysts (CBVD) is a rare congenital malformation of the bile ducts. Its frequency is between 1/13000 and 1/200 000 births [1]. The CBDC is characterized by intercurrent cystic dilatations of the bile ducts [2-4]. BVDB is due to abnormal biliopancreatic junction (AJBP) [2, 5, 6]. It affects mainly the young patient [7]. There are five types according to Todani's classification (Figure 1) [8]. Several complications can occur, the most frequent is angiocholitis (11%) [9] and the most serious is tumor degeneration (0.7 to 6.8%) [10].The aim of our work was to study the diagnosis, therapeutic and the evolution of BVCD through a series of 11 cases diagnosed in adulthood.

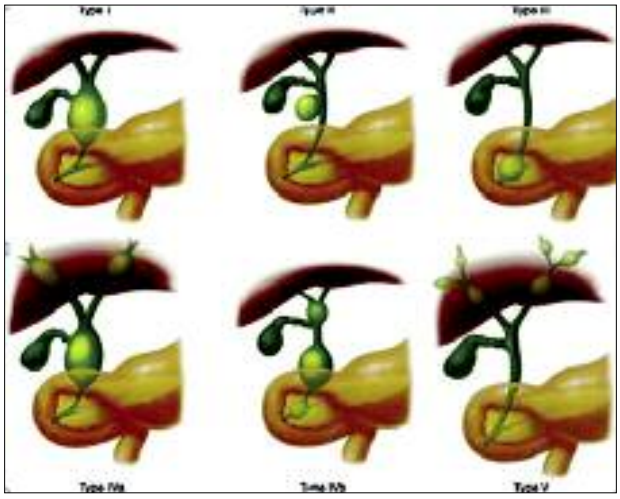

Figure 1: Todani's classification of cystic dilatation of the biliary tract

METHODS

This is a monocentric, retrospective and descriptive study. It collects all cases of CBDC in the general surgery department of Fattouma Bourguiba Hospital in Monastir between 01/08/1999 and 30/06/2009. Observations were summarized by a detailed study of the records concerning the characteristics of the ground, the clinical and exploration data, the operative finding, microscopic examinations and operative suites. We specified the therapeutic modalities as well as short and long term results for each type of CBVD.

RESULTS

We collected nine women and two men. The mean age was 45.3 years (Extreme from 17 to 77 years). Right hypochondriapain was found in all patients. Physical examination found jaundice in five cases. Biology showed cholestasis in six cases and cytolysis in four cases. Only one case had hyperamylasemia (five times normal). All our cases underwent radiological investigations. The abdominal ultrasound was performed in seven cases. It allowed us to evoke the diagnosis of BVCD in a single case by showing a cystic formation in continuity with the biliary tract. However, the problem of differential diagnosis with liver hydatid cyst was mentioned in two other cases. Abdominal CT-scan was performed in eight cases. It strongly referred the diagnosis of BVCD in all of these cases. Magnetic resonance imaging (MRI) was performed in three cases. It allowed the confirmation of the CBDC in a patient and to specify its type. No cases of AJBP has been found. Diagnosis has been confirmed in eight cases. Surgical treatment was chosen in front of degeneration risk. All cases were operated using a right subcostal incision. There were no liver metastases or peritoneal carcinomas. Peroperative cholangiography was performed in all operated cases. It confirmed the diagnosis of CBDC in all the cases and classified them according to Todani's classification. In our series, Todani type I was present in nine cases and Todani type IV in two cases. The surgical procedure consisted in resection of the primary biliary tract with hepaticojejunal anastomosis in eight cases with Todani Type I CBDC. A large bile duct and hepatic resection with hepaticojejunal anastomosis were performed in a patient who presented a gallbladder carcinoma confirmed by a microscopic examination. A biliodigestive and gastrojejunal double palliative anastomosis were performed in two cases who had extensive gallbladder tumor to the duodenum and lymphatic extension. Microscopic examination was performed in cases who underwent CBDC resection. Three cases of associated gallbladder carcinoma were retained. Immediate surgical follow-up was simple in ten cases. A case of surgical recovery was needed due to infection of necrotic pancreatitis whose operative follow-up was simple. The mean follow-up was 40.6 months with extremes from one month to seven years. Only one case of death has been reported. It was the case who underwent double biliodigestive derivation after one month of the operation. The death was related to uncontrolled infection due to acute angiocholitis.

DISCUSSION

In our series of adult CBDC, the average age of discovery was relatively advanced. It was 45.3 years. The discovery at this age makes the interest of our cases. The majority of reported cases are among childhood [1, 2]. According to Mutricy, it is diagnosed in 44% of cases before the age of 10 years, some authors even reach 60%.It can remain asymptomatic and be fortuitously discovered in adulthood. The discovery at the stage of complications is rare [3]. The CBDC represents 1% of all benign biliary affections [4-6]. It is more common in Asia and Japan than North Africa [7]. It represents 0.26% of the cases operated on for benign biliary pathology. In Tunisia, there are no

large series that report this malformation. To our knowledge, this is the second study of the CBDC after those of Manai et al [8]. It is essentially a female affection with a sex ratio of 3/1 [5]. This is consistent with our results. Pathophysiology at the origin of this malformation is subject to doubt. The most commonly accepted is advanced by Babbit in 1969 [7]. This theory suggests that CBDC is the consequence of an anomaly of choledochal and Wirsung duct junction (AJBP), which causes a reflux of the pancreatic secretion to the bile duct, which leads to the appearance of the most feared complication that is degeneration of the bile ducts [9]. This complication is more frequent and early after digestive anastomosis or incomplete cyst resection [7]. It is a cholangiocarcinoma in 90% of cases [9]. This AJBP is defined by three anatomical criteria [7, 10]: The existence of a common channel longer than 15 mm, an extra duodenal junction of the biliopancreatic channel at a distance from the sphincter and a connection angle greater than 30°. It was not possible to specify the rate the AJBP rate in our cases because MRI has not been done in all our cases. Generally, clinical triad "pain, jaundice, mass" evokes a CBDC. But this condition may be asymptomatic. In most cases, radiological examinations allow diagnosis and classify the CBDC (Figure 1) [11]. Abdominal ultrasound and abdominal CT scan allow diagnosis, especially in types I, IV and V [8]. Endoscopic retrograde cholangiopancreatography (ERCP) is currently supplanted by cholangio-wirsungo-MRI [8]. Both of these examinations show biliary malformation and AJBP in a non-invasive technic and have a biliary tract mapping allowing surgeons to plan the treatment (Figure 2).

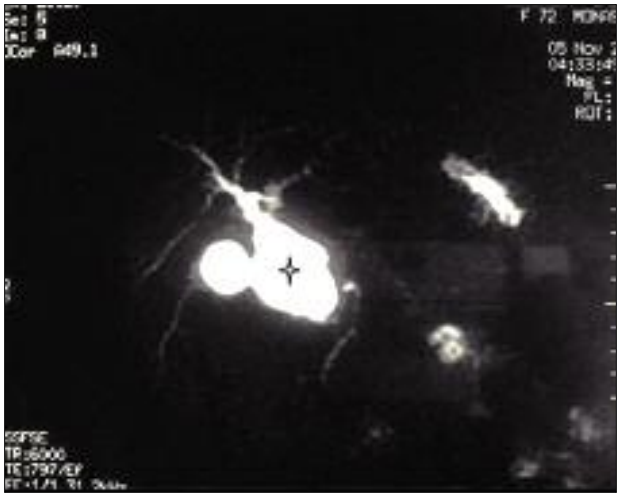

Figure 2: Bili-MRI showing a fusiform dilatation of the choledochus suggestive of a type I CBDC of Todani.

While no imaging examinations can be used to confirm the degeneration of a CBDC [8]. The three degenerated cases of our series were discovered all in the surgical treatment and confirmed by microscopic examination. The risk of angiocholitis and the rate of degeneration, which can reach 50% beyond the age of 50 years and the survival is 5% at 2 years of age, then the diagnosis must be established as early as possible [12, 13]. The proposed gesture is based on the type of Todani's classification. In type I, a complete excision of the cystic dilatation and the gallbladder is indicated with a hepaticojejunal Roux-en-Y anastomosis using classic or laparoscopic method [6]. This treatment took a long time to become apparent due to the technical difficulties and initial complications (hemorrhage, pancreatitis, fistula or papillary disintegration) which can be seen especially during a severe or traumatic dissection of the pancreatic bile duct [8], whereas its current risk is less than 1% [7].For CBDC type II, it is sufficient to perform an excision by closing the orifice of communication with the biliary tree [1]. Type III surgical treatment involves transduodenal excision of the choledochocele associated to surgical sphincteroplasty or sphincterotomy that may be sufficient. For type IVa, if hepatic involvement is localized, the treatment is similar to that of type I associated with a hepatectomy carrying the affected biliary tract [1]. If the hepatic involvement is diffuse (Figure 3), the treatment is similar to type V. Treatment of the CBDCtype IVb is the same as type I. For CBDC type V, it is not necessary to respect the common bile duct. In Caroli disease, type V localized, that is no therapeutic problem. Hepatic resection can allow a definitive cure of this disease. The problem arises in the case of diffuse bile duct involvement, in these cases transplantation is the intervention of choice [1], and it is discussed according to the age, evolution of the disease and existence of associated portal hypertension.

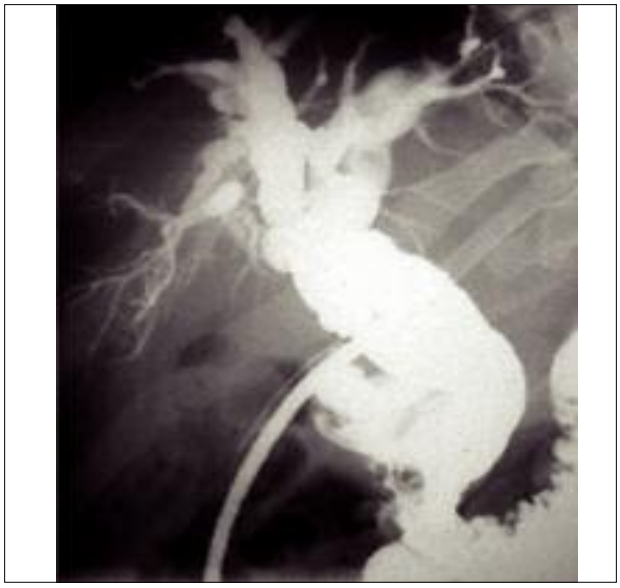

Figure 3: Intraoperative cholangiography show dilatation of bile duct tract.

CONCLUSION

CBDC is rare. It is necessary to think of it even at an advanced age. Advanced age is correlated with an increased risk of degeneration with pejorative prognosis. Radiological exams suspect the diagnosis at an early age and in asymptomatic patients. The treatment is always surgical. It should involve a removal of the dilated bile ducts followed by a biliodigestive anastomosis. Postoperative complications should be prevented by a good surgical technique and an early treatment.

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