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Duodenal Duplication Cysts - A Case Report of the Oldest Man in Literature

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Abstract

In the cluster of the congenital malformation of gastrointestinal system there is visceral duplication. This is a rare development abnormality that occurs mainly in infant and children and is detected for symptoms related to obstruction or to gastrointestinal bleeding. Only 30% of cases are diagnosed in adult. In Literature, is reported from 1 out of 25,000 to 1 out 100,000 deliveries of duplication of gastrointestinal system, but duodenum duplications are less than 5% of all. Duodenal duplication cysts are usually located in the second or the third part of duodenum, and can communicate with duodenal lumen or be extended into posterior mediastinum, communicate with gall bladder or communicate with the pancreaticobiliary duct.

We reported the case of a 70 years-old-man (the oldest one in literature) with a duodenal duplication cyst, his treatment and his outcome.

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Introduction

Among congenital gastrointestinal malformations, visceral duplication is a very rare development abnormality. In the majority of cases, the diagnosis is reached during infancy or childhood due to acute symptomatology related to obstruction or gastrointestinal bleeding. Only in about 30% of cases visceral malformations are supposed in adulthood [1,2]. The possible localization of duplication cysts is at any level of alimentary tract, from mouth to anus with a high variability of size, type and shape. Distal ileum, esophagus, colon and jejunum are the most frequent localization [2,3]. According to literature data, gastrointestinal system duplications is observed in 1 out of 25,000 or 1 out 100,000 deliveries, but duodenal duplications are less than 5% [2-4]. Duodenal Duplication Cysts (DDCs) are usually located in the second or the third portion of duodenum, communicating with duodenal lumen in about 25% of cases [2]. Moreover, they can be extended into posterior mediastinum [3], communicated with gall bladder in absence of other connections with biliary system [5]. In addition, DDC might cause recurrent pancreatitis especially in presence of communications with bilio-pancreatic ducts [6].

In consideration of the rarity and the unspecific symptomatology, the diagnosis is extremely hard [2]. In literature three cases of malignant transformation have been reported, even if in almost cases they are benign lesions [7]. According to several Authors, surgery -total excision, enucleation, digestive derivation- is the treatment of choice. Nevertheless, in consideration of the high postoperative morbidity, a minimal invasive endoscopic drainage is more and more proposed with very interesting results. Neoplasm position, size and extension are the main factors determining therapeutic strategy [2,3,7].

We report herein a rare case of DDC in a 70-year-old man, the oldest patient observed in Literature, undergone a derivative "open" surgical procedure.

Materials and Methods

In November 2015 a 70-year-old man presented to an emergency department with diarrhea and jaundice. Laboratory examination showed increased bilirubin levels and liver enzymes - total bilirubin =8.04 mg/dl (n.v. <1.2); AST-GOT =52 UI/L (n.v. <32); ALT-GPT =105 UI/L (n.v. =33). An abdominal Ultrasound (US) was performed and a hydropic gallbladder with thickened walls and a lumen occupied by biliary sludge associated with dilation of the intrahepatic bile ducts and

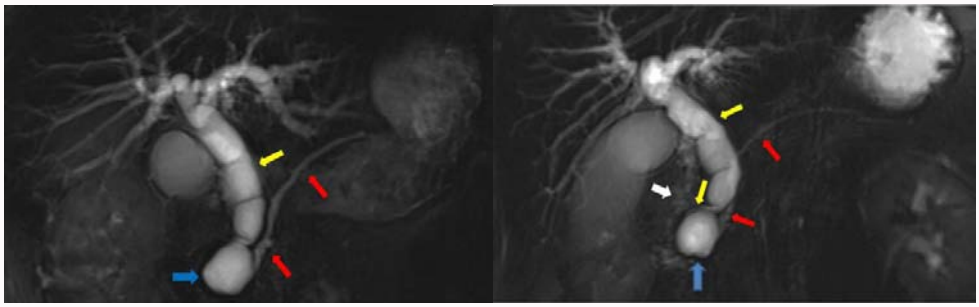


Figure 1a, 1b: Magnetic Resonance Cholangio-Pancreatography (MRCP) obtained with HASTE T2 radial sequences. The figures show the duplication duodenal cyst (duodenum: White arrow; cyst: Blue arrow) compressing the terminal choledochal (yellow arrows) and main pancreatic duct (red arrows) which appear severely dilated above the stricture.

of medium proximal common bile duct were observed. Subsequently a Magnetic Resonance (MR) described in correspondence of the periampullary region, a cystic formation of 27 mm × 24 mm × 25 mm with inhomogeneous content due to the presence of corpuscular slopes elements. This formation interrupted the Wirsung duct showing an apparent communication with it. Moreover, it displaced, compressed and imprinted the pre-papillary Choledochal segment, which appeared filiform and showed no signs of direct communication with the cyst itself. Wirsung duct, intra and extra hepatic *via* appeared markedly increased (Figure 1).

These findings were compatible with choledochal cysts (type II or III) or Intraductal Papillary Mucinous Neoplasms (IPMN). The patient underwent a hepatobiliary scintigraphy (185 Mbq of 99m-Tc-HIDA i.v.) that revealed strong hepatic fibrosis due to the absence of biliary washout.

After two weeks, he presented nausea and intense jaundice. Abdominal examination revealed mild tenderness in the epigastrium without rigidity or palpable masses. Laboratory examinations show a total bilirubin of 12.28 mg/dl; direct bilirubin of 10.57 mg/dl (n.v. = <0.3), γGT of 203 UI/L(n.v. =5-36); AST of 53 UI/L; ALT 72 UI/L.

Finally, according to endoscopic ultrasonography three possible diagnoses were supposed: DDC, for its structural characteristics, choledochocele and cystic dystrophy of the duodenal wall, even if groove pancreatitis was not observed.

Considering diagnostic doubts and simultaneous presence of gallbladder lithiasis, we opted for a subcostal laparotomy. A hydropic gallbladder, a severe dilatation of the main bile duct, and a duodenal cistic structure in correspondence of the papilla were observed. A cholecystectomy was performed and a cholangiography excluded choledochocele. The duodenotomy demonstrated a voluminous cyst protruding inside the lumen of the second portion. The cyst was then marsupialized through the common wall, and a cytological examination of the cystic content and a wide cystic wall biopsy was performed intraoperatively. The results were negative for malignancy, therefore a cystoduodenostomy was performed and a 12 French T biliary drainage was positioned (Figure 2).

Histopathological examination showed fibromuscular tissue fragments, partially covered by columnar epithelium, presumably expression of benign mucous cysts. A steady decrease of total bilirubin level with complete regression of jaundice was slowly observed. Six months after operation an ultrasound examination showed a reduction of dilatation of the biliary tract in absence of morphological abnormality of duodenal- biliopancreatic structures.

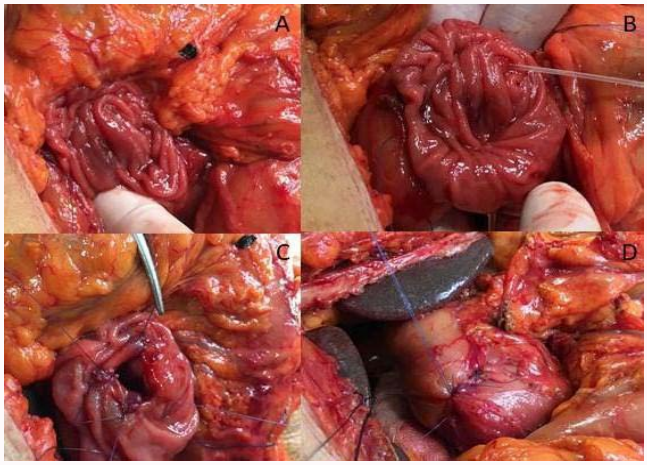


Figure 2: Intraoperative time. A: duodenectomy with eversion of mucosa. B: identification of cystic wall. C: cystic marsupialization. D: closure of duodenal wall.

Nowadays, 5 years after surgery, patient is in good health and without any symptoms correlated with this abnormality.

Results and Discussion

Duplication cysts of GI tract are rare congenital anomaly of unclear etiology [6]. The main criteria for a correct diagnosis are: 1) an intimate relation to the GI tract; 2) the presence of a smooth muscular coat; 3) the presence of an alimentary mucosal lining.

A small percentage (less than 5%) of all cases is represented by duodenal duplication [6]. In 1967 was reported the first case and, since then, a limited number of interesting clinical pictures have been reported. Performing a PubMed, Google Scholar literature review, a total of 91 cases have been found between 1999 and 2015: 48% were diagnosed before 10 years of age, whereas 22% were found in the second decade of life and the remaining 30% were in patients older than 20 years, with a mean age of 22 (1 to 42 years-old). The case presented is extremely rare and interesting because is the older patient reported in literature with an asymptomatic history for pancreatitis or abdominal pain (Figure 3).

This congenital anomaly can be cystic or tubular, communicant or not with duodenal lumen, pancreatic duct or biliary system and can be stuffed of mucosal secretion, purulent content or bile. The most frequent type is cystic and not communicant [4]. These are generally located at the medial border of the first and second parts of duodenum and extend to the anterior or posterior side. In our case,

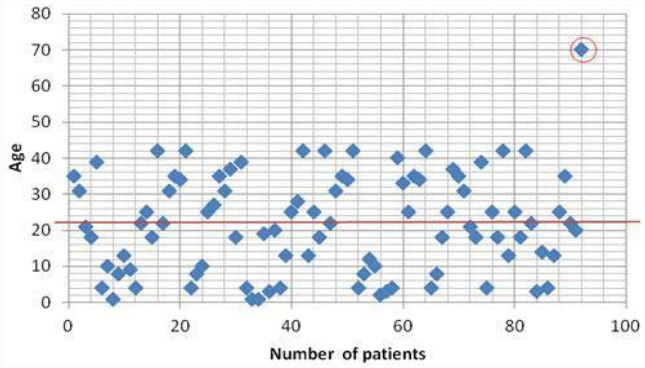


Figure 3: Graphical distribution of patient's age in literature. Red circle evidence our patients.

the duodenum duplication was cystic, not communicant, located in the second part of duodenum on the lateral border and extending to the posterior side.

Like other intestinal duplications, most duodenal duplications are diagnosed in infancy and childhood [4]. Even if in about 70% of cases DDCs are discovered during the first and second decades of life, however they can remain asymptomatic until adulthood. In the presented case the patient, 70 years old, indeed, is the eldest of English Literature and our paper confirms the diagnostic difficulty related to this asymptomatic rare congenital anomaly. Etio-pathological causes inhering DDC onset in elderly patient should be object of very interesting studies but nevertheless the debate is still open. Even if several theories have been proposed etiology remains unknown. In matter of fact, clinical features of DDC vary from asymptomatic cases to nonspecific symptoms such as abdominal pain, abdominal distension and vomiting [7].

In most of cases the dimensions are less than 5 cm but rare larger cysts palpable mass, gastric outlet or small bowel obstruction are other possible clinical pictures. Moreover, DDCs, for the direct pressure applied against the pancreatic duct, may cause recurrent pancreatitis. The presence of an ectopic mucosa is responsible of duodenal bleeding or peritonitis and stone formation inside the cyst has been reported. Jaundice is commonly described while hemorrhagic ascites is very rare complication [8].

In the presented case jaundice and nausea were the main symptoms. In addition, intestinal or biliary atresia, malrotation, gastric diverticulum, complete large bowel duplication, situs inversus, and intraspinal neurenteric cyst should be investigated as associated pathologies.

Preoperative diagnosis might be very difficult and rarely does it result accurate. The differential diagnosis comprises all cystic lesions in the bilio-pancreatic region like choledochoceles, pancreatic pseudo-cysts, pancreatic cystic tumors, mesenteric cysts and duodenal diverticulum [9].

In matter of fact, considering from an embryologic point of view that the hepatobiliary tree originates from the alimentary tract during early development, it is very difficult distinguish between DDC and type III of choledochal cysts. Unlike typical enteric duplication cysts, choledochal cysts by definition communicate with the biliary tree and can thus lead to multiple pancreaticobiliary pathologies. Large choledochoceles of the terminal common bile duct might cause biliary obstruction and pancreatitis and can be easily misdiagnosed as DDC, which would not be expected to communicate with the biliopancreatic tree [2]. In effect also in our case, preoperative MR defines the finding compatible with choledochal cysts (type II or III) or Intraductal Papillary Mucinous Neoplasms (IPMN) and only endoscopy ultrasonography-guided allowed to hypothesizes the diagnosis of "duplication cyst of duodenum". So this last diagnostic tool should be considered of paramount importance in the preoperative phase [2,4].

DDC treatment is object of intensive research and is still controversial. In case of asymptomatic lesions of small dimensions a "wait and see" policy is undoubtedly preferable. In the other cases surgeons are divided between favorable and detractors of radical resection, considering negligible incidence of malignant transformation and significant perioperative morbidity, sometimes reported following biliopancreatic operations. Type and localization of cyst, patients' characteristics, and clinical status should be considered as the main factors determining the choice of therapeutic strategy between surgical and mini invasive endoscopic procedures [7]. Complete surgical resection is recommended in case of suspect malignant transformation, that however was reported only in three cases in English Literature [4,7]. DDC radical resection, allowing a precise pathological examination, might require the association with dangerous procedures on biliary tree or pancreatic gland and so it should be indicated in selected cases or in patients with serious suspect of malignant neoplasms. Derivative procedures – cystojejunostomy or marsupialization with duodenum - should be suggested following endoscopic approach failure or if a "wide biopsy" is indicated. In the reported case cystoduodenostomy and a large biopsy of cystic wall was the preferred surgical procedure, preserving biliopancreatic structures and allowing a rapid functional recovery.

In the remaining symptomatic cases - old patients, ASA III - mini invasive procedures offer a rapid and efficacious resolution with short hospital stay and reduced costs. Recently, more and more papers, reporting favorable outcomes of DDC endoscopic treatment, have been focused this interesting issue and the debate is still open [10-14].

Conclusion

DDC is a congenital pathology of childhood and the onset in adults is very rare in clinical series. The Authors reported a case of 70 years old patient- the oldest patient reported in English Literature- in which definitive pathology confirmed diagnosis, following "open" cystoduodenostomy and full biopsy. Preoperative diagnosis might be very difficult and endoscopic ultrasound is of paramount importance excluding duodenal dystrophy or pancreatic neoplasms.

Follow-up is indicated in asymptomatic DDCs of small dimensions. Endoscopic treatment might be the treatment of choice in symptomatic patients reserving surgery in selected cases or in rare conditions of suspected malignant disease.

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