ABSTRACT

Duodenal duplication cysts (DDC) are rare congenital malformations that are usually found in infant and children, and DDC rarely presents in adulthood. The diagnosis of DDC is usually suspected by abdominal CT and endoscopic retrograde cholangiopancreatography (ERCP). Treatment is mainly surgical and total excision, if possible, is the procedure of choice. However, in some cases, alternative procedures, such as subtotal excision or digestive derivation are required due to extensive size or location. Here, a rare case of DDC, communicating with the main pancreatic duct is presented in which the treatment was subtotal excision with intra-duodenal cystoduodenostomy. Our objective in presenting this case of DDC is to increase awareness among physicians of this rare cause of recurrent pancreatitis, particularly in young patients, which is a surgically remediable cause of pancreatitis.

Case Report.

A 13-year-old girl was treated for 3 episodes of acute pancreatitis in a local hospital, with no etiology ascertained. She was referred to our institution for further evaluation of any underlying cause of recurrent pancreatitis. General and abdominal examination were unremarkable except for mild epigastria tenderness. Her biochemical and hematological profiles were normal.
Duodenal duplication cyst … Al-Qahtani

Abdominal ultrasonography (US) showed a cystic lesion between the second part of the duodenum and the head of the pancreas with no evidence of gallstones or bile duct stones. An abdominal CT scan showed a cystic lesion of 4.8 cm x 2 cm in diameter between the second part of the duodenum and the head of the pancreas (Figure 1). An ERCP revealed a normal biliary tract, mildly dilated main pancreatic duct, and a large protrusion into the second part of the duodenum, immediately distal to the major papilla. The main pancreatic duct was communicating with the cyst as the cyst distended with injection of the contrast in the main pancreatic duct (Figures 2a & 2b). At surgery, a duodenotomy was made, and the cyst was found just distal to the ampulla. Cystotomy was performed by incising the mucosa over the cyst. The common wall between the duodenum and the cyst was lined with mucosa on both sides. A clear fluid was seen draining into the cyst from the wall adjacent to the pancreas, thus, the diagnosis was DDC communicating with the main pancreatic duct. The wall between the cyst and duodenum was excised, and the margins were sutured to form a cystodudenostomy. Histopathology confirmed the diagnosis by identifying mucosa with its own muscularis mucosa on each side of the common wall between the cyst and duodenum (Figure 3). She remains asymptomatic at 14 months follow-up.

Discussion. Gastrointestinal tract duplications are rare congenital anomalies that can occur anywhere from the mouth to anus. A DDC constitutes 5-7% of all gastrointestinal duplications and its etiology is as yet unknown. It can present as a cystic or tubular mass, which can be communicating or non-communicating. They are generally located at the medial border of the first and second parts of the duodenum, which may extend to the anterior or posterior sides. The DDC in this patient was cystic and located at the second part of the duodenum on the mesenteric side. It was communicating with the main pancreatic duct. Intrapancreatic variants of DDC communicating with the main pancreatic duct have also been described.

The clinical manifestations are related to the location, size, and type of the DDC (communicating or non-communicating). Patients may present with abdominal pain, palpable abdominal mass, or signs of intestinal obstruction. Bleeding or perforation due to peptic ulceration and jaundice, and pancreatitis...
caused by biliary and pancreatic duct obstruction may also be the clinical manifestations of DDC.\textsuperscript{1,2,4,5} This patient presented with recurrent attacks of acute pancreatitis. Radiological methods, although helpful, rarely establish the diagnosis of DDC preoperatively.\textsuperscript{2} Abdominal US revealed an anechoic, double-walled, bi-lobed cystic lesion containing debris in the pyloro-duodenal region.\textsuperscript{6} An abdominal CT scan provides a better delineation of the location, size, and type of DDC.\textsuperscript{7} Technetium pertechnetate scintigraphy can be used to detect ectopic gastric epithelium in cases that present with bleeding. Fiberoptic duodenoscopy and ERCP help in detecting structural anomalies of the pancreatico-biliary ductal system in relation to the DDC.\textsuperscript{8} In the present case, ERCP successfully demonstrated the communication between the cyst and the main pancreatic duct by filling of the cyst during pancreatogram. With the increased availability of magnetic resonance cholangiopancreatography (MRCP) and endoscopic ultrasound, a diagnosis of duodenal duplication can be reached nowadays without the need for diagnostic ERCP.\textsuperscript{9} The differential diagnosis of DDC should include choledochocele, pancreatic pseudocyst, and intraluminal diverticulum. In this patient, intra-operative findings confirmed the presence of a communication between the DDC and the main pancreatic duct. Histopathology confirmed the diagnosis of DDC by identifying mucosa with its own muscularis mucosa on each side of the wall between the cyst and the duodenum. The management of DDC is determined by the size, type, location, and its relation to the duodenal wall, biliary, and pancreatic ducts. Total resection is the ideal surgical procedure for a non-communicating DDC. However, if it is not feasible, partial resection or internal derivation should be performed.\textsuperscript{2} In a series reported by Antaki et al.,\textsuperscript{9} endoscopic incision and marsupialization of the DDC performed by using a variety of endoscopic tools appears to be a safe and effective technique resulting in excellent long-term outcomes. However, the natural history of DDC remains uncertain in the long term, and malignant changes in DDC have been reported in the literature.\textsuperscript{10}

In conclusion, in patients with recurrent acute pancreatitis with a cystic lesion in the duodeno-pancreatic region in radiological studies, DDC should be considered in the differential diagnosis. In my opinion the ideal surgical treatment of the DDC is complete resection, which will remove the risk of potential complications associated with DDC. Partial resection and/or internal diversion can be carried out if complete excision is not feasible due to the proximity to biliopancreatic ducts.

References