CASE REPORT

Cystic dystrophy of the duodenal wall in heterotopic pancreas: A case report

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Abstract Cystic dystrophy of the duodenal wall is a rare benign disease characterized by development of multiple cysts in heterotopic pancreatic tissue in the duodenal wall. A forty-five-year-old male came to the emergency department of our hospital complaining of severe abdominal pain and vomiting. Computerized tomography (CT) scan of the chest, abdomen and pelvis had been done. The patient was treated by pancreaticoduodenectomy.

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1. Introduction

Cystic dystrophy of heterotopic pancreas was first described by the French authors Poter and Duclert in 1970 (1). It is characterized by development of cystic changes in heterotopic pancreas located in the duodenal wall. The pathogenesis of the development of these cystic changes is not fully known (2). CT scan may help in the diagnosis but usually the radiologic diagnosis is difficult (3). A MEDLINE search of papers published between 1966 and 2010 was carried out by Pezzilli et al. (4) and 59 papers were considered for the study; there were 19 cohort studies and 40 case reports. Chronic pancreatitis involving the entire pancreatic gland was present in half of the patients with cystic dystrophy of the duodenal wall and, in the majority of them; the pancreatitis had calcifications (4).

2. Case Report

A forty-five-year-old male came to the emergency department of our hospital with upper quadrant pain and severe projectile vomiting. He also gave history of recurrent attacks of milder form of vomiting and progressive weight loss (about 6 kg in the past two months) with recurrent epigastric pain.

On physical examination, severe epigastric tenderness was present but the abdomen was not distended. A round mass with ill defined borders and transmitted pulsation was also felt in the epigastric region.

The laboratory examination at the time of admission to the hospital showed elevated lipase; 1178 unit/litre (the normal range is 147–310 unit/litre) and elevated CA19.9; 46.7 IU/ml (the normal range is 0–37 IU/ml).

CT scan showed cystic lesion completely encircling the 2nd part of the duodenum. In addition, there was a solid component representing pancreatic tissue with mildly dilated ducts which open into the duodenum. The lesion leads to marked...
stenosis of the duodenum with subsequent marked dilatation of the stomach. The pancreas shows prominent head and borderline pancreatic duct. The liver shows minimal intrahepatic biliary radicle dilatation (Fig. 1).

The gross specimen consisted of distal part of the stomach, duodenum, pancreas and gall bladder. The gallbladder and the duodenum were unremarkable. The duodenum shows mucosal thickening with circumferential cystic lesion encircling the duodenum at the site of the mucosal thickening filled with clear fluid. Step sectioning of the duodenal wall reveals ectopic pancreatic tissue related to the wall of the cystic lesion.

Microscopically; the stomach, gall bladder and pancreas were free, apart from chronic gastritis and pancreatitis. Heterotopic pancreatic tissue was identified within the muscular layer of the duodenal wall. Dilated ducts of variable size were identified within the heterotopic pancreatic tissue. Sections taken from the main cyst that surrounds the duodenal wall show eroded lining, with minute stretches of reactive columnar epithelium.

The final diagnosis was cystic dystrophy of the duodenal wall in heterotopic pancreas, negative for malignancy.

3. Discussion

Pancreatic heterotopia is defined as the presence, outside its usual location, of pancreatic tissue that lacks anatomical and vascular continuity with the pancreas proper. The most common locations are the stomach, duodenum and jejunum. Cystic dystrophy is an uncommon but serious complication of heterotopic pancreas. It is characterized by the presence of cysts in the duodenal or gastric wall surrounded by inflammation and fibrosis, intermingled with pancreatic ducts and lobules. Cystic dystrophy of the duodenal wall is a rare disease and rarely published in the literatures. Due to the low frequency of the disease, there are a limited number of cohort studies (No. = 19) and a large number of case reports (No. = 40) (5). The cause of the cystic and inflammatory changes observed in the heterotopic pancreas is unknown. Although heterotopic pancreas is localized in the duodenal wall in 25% of cases, cystic dystrophy in heterotopic pancreas occurred in this site in all the patients done by Leger et al. (6) but one. The lesions could be the result of local conditions with repeated attacks of acute pancreatitis. These episodes may be determined by obstruction of minor excretory ducts of ectopic

Fig. 1  Axial CT scan image (A) shows cystic lesion (black arrow heads) encircling the second part of the duodenum (long thin white arrow). The stomach is markedly distended (short thick white arrow). Coronal reformatted CT scan image (B) confirms the location of the cystic lesion (white arrows) around the second part of the duodenum (black arrow). Axial CT scan image (C) at a lower level than (A) shows ectopic pancreatic tissue (white arrow head) with dilated ducts (white arrow) in the duodenal wall. Coronal reformatted CT scan image (D) shows clearly the cystic lesion (black arrow head) and the ectopic pancreatic tissue (long white arrow) encircling and narrowing the second part of the duodenum (black arrow). The pancreas shows mildly dilated pancreatic duct (short thick white arrow).
pancreatic exocrine lobules. The dilated ducts that were observed in all patients in the study done by Leger et al., favour this hypothesis (6).

A recent Italian survey which reviewed the data from 2000 to 2005 reported that the frequency of this entity is higher in males than in females by 5.5% and in alcoholics more than non alcoholics (7). However; in a study done by Flejou et al. (8) two only out of 10 cases were alcoholics. In a surgical setting, the incidence of pancreatitis ranges from 2.7% to 24.5% (9). The mean age of the disease was 47 years (10). Chronic pancreatitis of the entire pancreas was reported in 166 of the 302 (55.1%) patients and there were pancreatic calcifications in 125 of these 166 patients (75.3%) in a study done by Galloro et al. (11). The clinical presentation in most of the studies done was mainly caused by intestinal or biliary stenosis, or both, or secondary to inflammation or fibrosis as abdominal pain, nausea, vomiting, jaundice or weight loss (11).

The imaging diagnosis of dystrophy of the duodenal wall is rarely assessed using a single radiological modality. Even if ultrasound is the first line imaging modality in these patients, it is rarely diagnostic. Endoscopic retrograde cholangiopancreatography (ERCP), which was frequently used in the past, is feasible and in typical cases it demonstrates smooth tubular stenosis at the distal part of the common bile duct (CBD) without abnormality of the main pancreatic duct or, occasionally, with only slight irregularities (12–13).

CT Scan may reveal the presence of duodenal stenosis with wall thickening and cystic lesions in the duodenal wall or in the groove area. The cysts may be tiny and multicellular cystic lesions may be observed. The main pancreatic duct may be mildly dilated above the lesion. Delayed enhancement may also be seen in the thickened duodenal wall (5).

An important diagnostic aspect of magnetic resonance imaging (MRI), which cannot be evaluated by CT scan, is that MRI can be followed by MR cholangiopancreatography (MRCP); this additional evaluation provides images similar to those of the ERCP without the morbidity of this latter technique (14).

Conservative treatment is the main option in the acute phase of the disease. The main therapeutic option for these patients is a surgical approach. The most frequent surgical approach is a pylorus-preserving pancreaticoduodenectomy or a Whipple procedure; in a limited number of patients, a gastrointestinal by-pass, with or without biliary by-pass, has been carried out (15).

A new surgical approach has been recently carried out in two patients of the study done by Egorov et al. (16), who were successfully treated by two modifications of a pancreas-preserving duodenal resection with reimplantation of the bile and pancreatic ducts into the neoduodenum (16).

4. Differential diagnosis

4.1. Diverticulum of the duodenal wall

- Air-fluid, fluid or residual particulate food-filled mass along the medial wall of the 2nd or 3rd portions of the duodenum duplication cyst.
- Thin enhancing wall.
- Communication with the duodenal lumen.
- ± Dilated bile duct and/or pancreatic duct.

4.2. Choledococele

- Cystic mass in the duodenal lumen, at the periampullary level.
- Communicates and continues with the bile and pancreatic ducts.
- Signs of acute or chronic pancreatitis.
- Bile stones.

4.3. Duplication cyst

- Well-circumscribed cystic mass along the mesenteric border of the descending duodenum, high density content (blood clot), mural nodules (carcinoma).
- It can cause mass effect and eventually luminal compression.

5. How to reach the diagnosis of cystic dystrophy of the duodenal wall in heterotopic pancreas

- Clinical history of repeated attacks of vomiting with or without pancreatitis.
- Multiple small cysts in the duodenal wall.
- Remnants of pancreatic tissue in the duodenal showing the same density/signal intensity and pattern of enhancement of the pancreas. Tiny pancreatic duct could be present in this pancreatic tissue.
- Mural thickening of the second part of the duodenum.
- ± Stenosis of the duodenal lumen.

6. Conclusion

Cystic dystrophy of the duodenal wall is a rare benign disease characterized by development of multiple cysts in heterotopic pancreatic tissue in the duodenal wall. The diagnosis can be easily assessed nowadays due to technical advances in the imaging modalities as multislice CT scan, MRI and endoscopic ultrasonography.

The disease is associated with chronic pancreatitis in half of the patients and the pancreatitis has calcifications in the majority of them. Surgery is required in symptomatic patients who improved markedly after surgery.

Conflict of interest

We have no conflict of interest to declare.

References


