Cystic Dystrophy of the Duodenal Wall in Heterotopic Pancreas with Groove Pancreatitis: A Diagnostic and Therapeutic Challenge

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Rezumat

Introducere: Distrofia chistică a peretelui duodenal dezvoltată pe pancreas ectopic este o afecțiune rară, benignă, caracterizată prin apariția de chiste adevărate în peretele duodenului. Manifestările clinice nespecifice și imagistica greu de interpretat fac ca diagnosticul afecțiunii să fie dificil, mai ales atunci când există și suspiciune de malignitate. Tratamentul chirurgical (duodenopancreatectomia cefalică) oferă cele mai bune rezultate în tratamentul afecțiunii.

Prezentare de caz: Bărbat în vârstă de 48 de ani, consumator cronic de alcool, cu antecedente de pancreatită acută recurentială (episod ușoară), a fost diagnosticat în 2010 cu pancreatită cronică segmentară la nivelul capului pancreasului, complicată cu un pseudochist de 4 cm. Bolnavul abandonează alcoolul, dar episoadele de pancreatită persistă, în timp ce pseudochistul nu se mai evidențiază. În 2012, pacientul este internat în clinica noastră cu un nou episod acut, pe fond de pancreatită cronică dureroasă. Ecografia endoscopică pune diagnosticul de distrofie chistică a peretelui duodenal și pancreatită a „şantului” (groove), și se practică deschiderea endoscopică a chistelor în duoden. EUS evidențiază și calculi veziculare mici, pentru care efectuăm o colecistectomie laparoscopică. După 6 luni de evoluție asimptomatică, durerea și episoadele acute reapper, impunând tratamentul chirurgical (duodenopancreatectomia cefalică). La 5 ani postoperator, pacientul este asimptomatic.
Concluzii: Distrofia chistică a peretelui duodenal pe pancreas ectopic poate intra în discuție la stabilirea etiologiei unei pancreatite acute recurente. Ecografia endoscopică este cea mai eficientă metodă de diagnostic, care oferă și posibilitatea deschiderii chistelor în duoden (fenestrarea). Tratamentul chirurgical (duodenopancreatectomia cefalică) oferă cele mai bune rezultate pe termen lung.

Cuvinte cheie: distrofie chistică a peretelui duodenal, pancreatita cronică, pancreatita acută recurrentă, pancreas ectopic, tratament chirurgical, duodenopancreatectomie cefalică

Abstract

Background: Cystic dystrophy of heterotopic pancreas is a benign, rare disease characterized by development of true cysts into the duodenal wall. Non-specific clinical manifestations and difficult to interpret imaging provide a diagnostic challenge, especially when pancreatic cancer is suspected. Surgical treatment (pancreatoduodenectomy) offer best outcomes.

Case Report: A 48 years-old man, chronic alcohol consumer, with a history of recurrent mild acute pancreatitis episodes, was diagnosed in 2010 with segmental chronic pancreatitis complicated by a 4cm pseudocyst in the pancreatic head. He stopped drinking, but acute episodes continued to reoccur, while the pseudocyst disappeared. In 2012 the patient was admitted to our department with painful chronic pancreatitis and a new acute episode. EUS diagnosed a cystic dystrophy of the duodenal wall with groove pancreatitis, and endoscopic opening of the cysts into the duodenum was performed. Laparoscopic cholecystectomy was imposed by small gallbladder stones seen at EUS. After 6 months of silence, pain and acute episodes reappeared, imposing pancreatoduodenectomy. Patient is now well, without symptoms during 5 years of follow-up.

Conclusions: Cystic dystrophy of heterotopic pancreas can be thought of in case of recurrent idiopathic acute pancreatitis. EUS is the best diagnosis tool, and can provide opening of the cysts into the duodenum. Surgery – pancreatoduodenectomy – offer best results.

Key words: cystic dystrophy of the duodenal wall, groove pancreatitis, recurrent acute pancreatitis, heterotopic pancreas, surgical treatment, pancreaticoduodenectomy

Introduction

Cystic dystrophy of the duodenal wall in heterotopic pancreas (CDHP) was first described in 1970 by Potet and Ducler (1). This relatively rare entity is associated with a segmental chronic pancreatitis (inflammation and fibrosis) localized in the C-loop of the duodenum, around the pancreatic head (1,2,3). Stolte et al (4) defined in 1982 this area as a “groove”, and proposed the term of “groove pancreatitis” (GP), which was well accepted. Later, in 1991, Becker and Mischke classified it into a pure form, and a segmental form (5). Actually, CDHP is not always associated with GP, as Pezzilli et all (6) had shown in 2011 in a systematic review. Aiming to unify the nomenclature, CDHP associated with GP was named “paraduodenal wall cyst”, term introduced in 1982 (7). Finally, Adsay and Zamboni (8) proposed the term “paraduodenal pancreatitis”, which include all 3 previous names.

No matter what nomenclature we use, the disease is associated with male gender 40 to 50 years old, alcohol consumption and smoking. Most patients are admitted with abdominal pain, vomiting, weight loss, and abdominal imaging shows cystic lesions in the pancreatic head region. Differentiation from pancreatic cancer can be difficult in the absence of the cysts, and if it is not clear, surgery is indicated (9,10). Conservative and
endoscopic treatment may be effective in almost 50% of patients, while surgery is still the best option (10). A stepwise approach, starting with conservative treatment, followed by endoscopic and finally surgery seems a logical option.

We herein present a case of CDHP with GP clinically masquerading as a recurrent acute pancreatitis, which was treated by a stepwise approach. We aim to illustrate all steps of the diagnosis and treatment with appropriate images, characteristic for the disease.

Case Report

A 48 years-old Caucasian male, chronic alcohol consumer, smoking 30 cigarettes /day for more than 25 years, with a 2 years history of recurrent mild acute pancreatitis episodes, was admitted to our department with the classical symptoms of acute pancreatitis. The patient described his recurrent upper abdominal pain, a weight loss of 14 kg in the last 2 years, and provided documents for previous confirmed episodes of pancreatitis. A mild acute episode was diagnosed and treated conservatively. Contrast-enhanced CT (performed 8th day from onset), described a pancreas with signs of chronic pancreatitis, thickening of the duodenal wall, and a 4 cm cystic lesion in the pancreatic head (Fig. 1) in contact to D2 and D3, compressing the Wirsung duct which was dilated to 3-4mm in the body and tail. The lesion was interpreted as a pseudocyst. Upper endoscopy showed extrinsic compression of the duodenum (D2 and D3), without stenosis.

After discharge from hospital, the patient respected the low-fat diet, stopped alcohol consumption, and had no more symptoms for 6 months. Ultrasound performed at 3 months follow-up could not see any cystic lesion. Painful episodes reoccur, with a shorter duration (1-2 days), and a monthly frequency. A readmission to hospital was imposed by a new acute pancreatitis episode, 2 years after the previous one. CT demonstrates a 3.9 cm cystic lesion with liquid content in the pancreatic head, intimal adherent to D2 (Fig. 2). As CT was not able to say if the recurring cyst is a pseudocyst, a walled-off necrosis, or a serous cystadenoma (normal CA19-9 and CEA), we scheduled the patient to EUS (endoscopic ultrasound), which clearly described in the submucosa and muscular layer of D2 three pseudocysts (5 mm, 2 cm and 2.6 cm), moderate changes of chronic pancreatitis in the pancreatic head, and small gallbladder stones. Cysts liquid analysis showed slightly elevated amylase content, normal markers, and no malignant cells at cytology. The EUS operator also performed an opening of the cysts into the duodenum (cysts fenestration). Laparoscopic cholecystectomy was performed, and the patient was discharged, with the diagnosis of cystic dystrophy of the duodenal wall.

After 6 months of wellbeing, without any symptoms, the patient had 2 readmissions for
new acute pancreatitis episodes (at 1 month interval). The cysts in the duodenal wall were present again (Fig. 3) despite the previous endoscopic treatment, which made us decide for surgery. We performed a pylorus preserving pancreaticoduodenectomy. While preoperatively reviewing the CT images, we found an aberrant right hepatic artery originating from the superior mesenteric artery (not reported, but confirmed by the CT operator) (Fig. 4), which we were able to dissect and preserve during surgery (Fig. 5). Macroscopic pathology examination of the surgical specimen described the presence of 3 cysts (1.6 to 2.4 cm) in the thickened duodenal wall, containing small intra-cystic calcifications, and the groove pancreatitis (Fig. 6). The cysts do not communicate with the duodenum, main biliary duct or Wirsung’s duct. On microscopy, cysts are situated in the muscular layer of the duodenum, and have a cuboid ductal-type epithelium, mostly ulcerated (Fig. 7). In the duodenal wall, in the vicinity of the cysts, fragments of heterotopic pancreas can be seen (Fig. 8). Pathologic examination confirms the diagnosis of CDHP with GP.
During 5 years of regular follow-up visit, the patient had no symptoms, gained weight (7 Kg), and has no signs of endocrine or exocrine pancreatic insufficiency.

Discussion

Paraduodenal pancreatitis (CDHP + GP) is a rare condition, with an incidence of 0.4-14% in autopsy studies (11). The real incidence is not known, but the disease can be observed in at least 3% of patients undergoing a pancreaticoduodenectomy (12). As pathogenesis, heterotopic pancreatic lobules have ducts not connected to the pancreas ductal system. Accumulation of pancreatic juice can produce recurrent episodes of acute obstructive pancreatitis, leading to development of retention cysts (12,13).

The patients have usually a late diagnosis, as there are no specific symptoms, and the nature and location of the cysts can be accurately described only by EUS or MRI (14,15). Diagnosis can be delayed from months to years (9), as it was 2 years in our patient. If doubt persist, and suspicion of malignancy exists, surgical treatment is advised (9,16).

Once the diagnosis is established, management must be conducted in a stepwise manner, starting with conservative treatment (stop alcohol and smoking, low fat diet, analgesics).

According to the systematic review (9), this treatment can be final in 39% of patients. Endoscopic treatment, with cysts drainage into the duodenum is successful in almost half of patients. In our patient, result of endoscopic fenestration of cysts was good for 6 months, followed by recurrence of cysts and symptoms. Surgical treatment was reported in 60% of patients, with good results (16). The procedure of choice is pancreaticoduodenectomy, which resect the duodenum where the inflamed
heterotopic pancreas had become cystic, and the pancreatic head, which is the “pacemaker” of pain and inflammation in chronic pancreatitis. Our patient is well, without any symptoms after 5 years of follow-up.

We performed a pylorus preserving pancreaticoduodenectomy, preserving the aberrant right hepatic artery (ARHA) originating from the superior mesenteric artery. ARHA is the most common variation of the common hepatic artery, with an incidence of 11% to 26.5% in the literature (17,18). This variation may be a problem in pancreaticoduodenectomy, due to its course near the vascular margin, especially near the superior mesenteric artery. Any intra-operative damage of ARHA can produce bile duct and/or right hepatic lobe ischemia, resulting in fistula of the hepatico-jejunal anastomosis, liver necrosis, abscesses and patient death. Preoperative careful examination of CT images is mandatory to prevent incidental discovery of ARHA, usually after a lesion has been produced.

Conclusions

Cystic dystrophy of heterotopic pancreas was clinically manifested in our case as a recurrent acute alcoholic pancreatitis, which produced a 2 years delay in diagnosis. So, CDHP with GP can be thought of in case of recurrent idiopathic acute pancreatitis. EUS is the best diagnosis tool, and can provide cyst liquid analysis, and opening of the cysts into the duodenum. Surgery (pancreaticoduodenectomy) offer best, long-term results.

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References