Groove pancreatitis mimicking pancreatic cancer: Case report and literature review

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Abstract

Groove pancreatitis (GP) is a rare form of chronic pancreatitis located in the pancreaticoduodenal groove, hence its name. It is predominant in males with a history of alcohol intake. Making a differential diagnosis between this condition and pancreatic cancer is highly relevant given its location. Advances in diagnostic methods, such as endoscopic ultrasonography, have allowed a more accurate approach. However, no management guidelines are available and therapeutic approaches are still based on similar pathologies.

Keywords

Pancreatitis; Pancreatic cancer.

INTRODUCTION

Groove pancreatitis is a rare form of chronic pancreatitis. Its name derives from its location in the pancreaticoduodenal groove. It occurs predominantly in males with a history of alcohol consumption. Due to its location, considering pancreatic cancer as a differential diagnosis is of great importance. Currently, the advances that have been made in terms of diagnostic methods, such as endoscopic ultrasonography, have allowed a more accurate approach; however, there are no management guidelines and therapeutic interventions are still based on on those used in similar diseases.

CLINICAL CASE

This is the case of a 44-year-old man with a history of acute pancreatitis, alcohol consumption and smoking since he was 20 years old, who visited our health institution due to having experienced epigastric pain during six months, pain that worsened 4 days before admision. Elevated lipase, alkaline phosphatase and bilirubin levels (predominantly direct bilirubin) were reported in the laboratory tests results, so a cholestatic pattern was considered. A hepatobiliary ultrasound showed a 13 mm dilated common bile duct, without evidence of choledocholithiasis. In view of the patient's clinical condition, a magnetic resonance cholangiopancreatography was performed, which showed a concentric diffuse thickening of the walls of the second part of the duodenum with a 24 x 23 x 24 mm irregular exophytic mass on the anterior wall of the duodenum, hyperintense on T1 and hypointense on T2 with restricted diffusion, which was causing the obstruction of both intraand extrahepatic biliary ducts.

Biliopancreatic ultrasonography was then performed using linear Pentax equipment, where changes suggestive of chronic pancreatitis and dilatation of the common bile duct were found; however, in the endoscopic phase of the procedure, edema and erythema of the bulb and the second part of the duodenum were observed with an ultrasound thickening of the duodenal wall surrounding the head of the pancreas and dependent on echolayers 1 and 2, and partially on echolayer 3, i.e. the mucosa and submucosa; but the echostructure of the layers was preserved, without perilesional adenopathies. The puncture did not show any lesions suggestive of neoplasia. Then an endoscopic retrograde cholangiopancreatography (ERCP) was performed where the following findings were observed: a large edema, erythema and erosion in the second part of the duodenum, and edematous major and minor duodenal papillae with dilatation of the common bile duct, but without presence of stones (Figure 1). Biliary stent placement was carried out to ensure adequate drainage. In addition, biopsies were performed due to the clinical suspicion of ampullary lesion, with the following findings informed in the biopsy report: "inflammatory response without evidence of dysplasia or neoplasia". The patient's condition improved satisfactorily and currently he is under periodic clinical follow-up,

undergoing oral enzymatic treatment and following nutritional recommendations, so it was considered that this was a groove pancreatitis case that mainly affected the duodenum and the two duodenal papillae and caused the obstruction of the bile and pancreatic ducts.

DISCUSSION

The duodenal pancreatic groove is a small area located between the head of the pancreas, the duodenum and the common bile duct (**Figure 2**) (1). The term groove pancreatitis refers to a type of chronic pancreatitis that mainly affects this area of the pancreas, while the rest of the organ remains intact (2, 3). It is a rare disease, probably because it is underdiagnosed (4).

Groove pancreatitis was first described in 1973 by Becker, who used the German word Rinnenpankreatitis (5). In 1982 Solte et al. translated it as groove pancreatitis (2). Subsequently, several terms have been used to refer to this disease in the literature such as dystrophy of the pancreas or duodenal dystrophy, which was reported by the French authors Potet and Duclert (6). Other terms include heterotopic cystic dystrophy, duodenal/paraduodenal wall cyst, pancreatic hamartoma of the duodenal wall, myoadenomatosis, Brunner's gland hamartoma and paraduodenal pancreatitis. These terminology differences make it difficult to find information of this condition in the literature (7).

In 1991, Becker and Mischke described two forms of groove pancreatitis: pure and segmental (8). In the pure form, infiltrative involvement or scar tissue affects only the pancreatic groove, while the parenchyma and the main pan-



Figure 1. Edema in the papilla and peripapillary region.



Figure 2. Anatomical relationships of the pancreatic groove.

creatic duct remain intact. In the case of segmental groove pancreatitis, scar tissue extends to the head and body of the pancreas, near the duodenal wall and the main pancreatic duct, which causes stenosis (9, 10). A clear differentiation between both is not always possible (11). Groove pancreatitis is more common in men with a history of chronic alcohol consumption and tends to occur in the fourth or fifth decade of life. Also, some case series have described smoking as a risk factor (12, 13).

The importance of groove pancreatitis is given by its capacity to imitate pancreatic carcinoma (10). In fact, there are cases of coexistence or even masking of this type of cancer (14), so it must be considered in the differential diagnosis of pancreatic masses or duodenal stenosis (15, 16).

CLINICAL PRESENTATION

The clinical presentation of groove pancreatitis consists of epigastric or mesogastric pain associated with postprandial emesis, weight loss (mainly due to altered intestinal motility) and duodenal stenosis (12, 17). The presence of jaundice is usually fluctuating, except when it occurs concomitantly with pancreatic cancer or cholangitis (18, 19). The course of the disease is chronic, and it may last years from the onset of symptoms and the time diagnosis is reached (20).

Regarding its pathophysiology, it is not clearly defined yet. It is believed that groove pancreatitis is related to the interruption of the pancreatic juice drainage through the duct of Santorini, which is located in the body of the pancreas and drains to the duct of Wirsung forming an acute angle; once there, it causes pancreatitis in the groove area due to increased pancreatic intraductal pressure (15). In addition, groove pancreatitis is caused by anatomical alterations in the minor duodenal papilla or by dysfunction of the papilla (3, 21). Tumors that occlude the minor papilla and the duct of Santorini (7), an obstructed duct of Santorini, pancreas divisum and heterotopic pancreas in the duodenal wall are among said alterations (15). Functional causes are associated with precipitating factors. Some of these include Brunner's glands hyperplasia and chronic excessive alcohol use or smoking, since they lead to dysfunction of the minor duodenal papilla and increased amounts of proteins in the pancreatic juice (3, 22). In the case reported here, alcohol consumption and smoking were considered precipitating factors of groove pancreatitis.

Macroscopic differentiation of this type of pancreatitis occurs in two scenarios: the cystic type, with 10 mm to 10 cm cysts in the duodenal mucosa beyond the ampulla of Vater region; and the solid type, characterized by the presence of <10 mm cysts associated with a significant thickening of the duodenal wall (23). Additionally, scarring causes changes in the common bile duct, leaving a smooth

surface with homogeneous hyalinization changes (7). In early stages of the disease, it slightly affects the head of the pancreas and produces some scars or retractions. However, as it progresses, it produces important fibrosis that affects the head of the pancreas in its entirety (23).

Microscopically, histological findings include dilatation of the pancreatic ducts, the formation of pseudocysts in the duodenal wall, fibrosis in the duodenal submucosa that extends to the neck of the pancreas and Brunner's glands variable hyperplasia, which causes a thick layer around the smooth muscle with myofibroblast proliferation (24). Cystic changes are observed in the duodenal wall, going through the muscularis propria and superficially reaching the pancreatic parenchyma. Myofibroblastic proliferation can enclose areas of cystic changes with very thick secretions corresponding to spindle cells (13). Additionally, the dilated ducts and columnar epithelium can erode and transform into a pseudocyst-like fibrosis that constitutes the cyst type mentioned here (13, 25).

DIAGNOSIS

Groove pancreatitis diagnosis is made based on a set of biochemical tests, imaging studies and, in some cases, biopsies. Pancreatic enzyme levels (amylase and lipase) may be elevated (26). In addition, since inflammatory presentation, instead of the neoplastic one, occurs in most cases, elevated levels of tumor markers of the gastrointestinal tract (CA19-9 and carcinoembryonic antigen) are not usually found (10). Regarding imaging studies, abdominal ultrasound shows a hypoechoic lesion that thickens the duodenal wall and causes both a stenosis in the second part of the duodenum and the obstruction of the common bile duct (22). The disease can be documented by means of ultrasound in several stages: in the early stage, fibrosis predominates and it is possible to observe a hypoechoic band over the pancreatic grooves, which is associated with a thickening of the duodenal wall; on the other hand, e in the late stage, once fibrosis has been established, hyperechogenicity in the duodenal wall is observed as a result of hypertrophy of the submucosa due to Brunner's glands hyperplasia (27).

IMAGING FINDINGS

Abdominal computed axial tomography (CT) scan shows a hypodense laminar lesion between the head of the pancreas and the duodenum, close to the minor duodenal papilla; this lesion is made up of scar tissue (28, 29). In the case of contrast enhanced CT scans, there is a delay in the uptake of the groove due to a circulation alteration secondary to fibrosis (10). In the case of segmental groove pancreatitis, a hypointense lesion can be seen in the pancreatic head, close to the duodenal wall. The main pancreatic duct may show a discrete dilatation towards the body and tail of the pancreas (30). Peripancreatic vascularization is usually preserved, without signs of thrombosis, even in the presence of extensive involvement (31). In the most advanced stages of the disease, a narrowing of the lumen in the duodenum, together with the presence of edema, erythema and polypoid mucosa can be observed in upper endoscopy. Biopsies of the duodenal mucosa usually report an inconclusive result or an active inflammatory response without any evidence of neoplastic lesions (24).

ERCP is a technically difficult procedure since the positioning of the duodenoscope in the presence of duodenal edema limits the visualization of the papilla. The distal common bile duct shows a stenosis without involvement of the main pancreatic duct. In addition, the duct of Santorini obstructed by mucus plugs or simply by the large perilesional edema (4, 32). In the case of our patient, endoscopic findings showed an edema of significant size in the duodenum and the peripapillary region (**Figure 3**).



Figure 3. Endoscopic ultrasound image of edematous duodenal wall.

Endoscopic ultrasound (EUS) has been proposed as the diagnostic technique of choice due to its greater sensitivity (86%) and the added value of making it possible to take samples for histological analysis purposes (33). Imaging findings in EUS show chronic pancreatitis changes based on the Rosemont's criteria, which allow it to have a good diagnostic performance (34). As it happens in other imaging studies, a thickening of the second part of the duodenum and heterogeneity of the dorsocranial portion of the pancreatic head findings have been reported with this imaging technique. In addition, several authors have described a pathognomonic finding using EUS in advanced

stages of groove pancreatitis: hyperechoic thickening of the duodenal wall in the groove area with anechoic ductal structures extending towards the head of the pancreas. This hyperechoic lesion correlates with myoadenomatous proliferation and adjacent fibrosis of the pancreas (10, 27). In our patient, a significant edema in the duodenal wall and dilatation of the common bile duct were observed in the EUS (**Figure 3**).

Magnetic resonance imaging (MRI) offers the advantage of allowing the assessment of the different pathological aspects of the disease. The characteristic lesion between the head of the pancreas and the duodenum is shown as a hypointense lesion on T1 weighted imaging sequence, while on T2 sequence it can be hypo-, iso- or hyperintense depending on the time of evolution of the disease. In acute stages it is hyperintense due to the large edema, while in chronic stages it is hypointense due to fibrosis (28, 29).

Cystic lesions in the groove or in the duodenal wall are more evident in the T2 sequence and an increased thickening or duodenal wall stenosis are frequent findings. The common bile duct may show variable stenosis. In the segmental form of the disease, the head or the entire pancreas may appear hypointense on the T1 sequence, with atrophy of the parenchyma and ductal dilatation, showing the progressive loss of glandular cells that in turn are replaced by fibrous tissue (29, 35). ERCP reports the relationship between the pancreatic duct system and cystic changes. In most cases, a widening of the space between the pancreatic ducts, the distal common bile duct and duodenal lumen, due to the space-occupying lesion located in the groove, is found, as well as a marked thickening of the duodenal wall (28). In the case reported here, dilatation of the bile duct, caused by the edema of significant size located in the peripapillary region, was evidenced on magnetic resonance cholangiopancreatography (Figure 4).

The anatomical location of the pancreatic groove means that any lesion in this area must be studied histologically or using imaging techniques. Differential diagnoses include pancreatic head carcinoma, periampullary cancer, neuroendocrine tumor, cystic dystrophy of the duodenum and acute pancreatitis with plastron presence in the groove. Differentiating groove pancreatitis from adenocarcinoma is particularly difficult as both conditions may have similar findings (36, 37). An important feature of groove pancreatitis is the absence of vascular involvement. In addition, it has been reported that the gastroduodenal artery is displaced to the left in this type of pancreatitis, whereas, in the case of adenocarcinoma, it is located between the lesion and the duodenum (38, 39). Also, cystic groove lesions are more common in groove pancreatitis than in adenocarcinoma cases (40). Assessment of the duodenum may also help differentiate groove pancreatitis from pancreatic



Figure 4. Magnetic resonance cholangiopancreatography showing a large edema in the second part of the duodenum.

cancer, as strictures are less common in pancreatic head tumors (35). Likewise, in groove pancreatitis, the presence of stenosis distal to the intrapancreatic portion of the bile duct is observed during magnetic resonance cholangiopancreatography, instead the abrupt and irregular stenosis seen in pancreatic adenocarcinoma patients (10). Periampullary cancer has a clinical presentation similar to that of ductal adenocarcinoma of the pancreatic head. Periampullary carcinomas typically occur in older adults, together with jaundice and weight loss; these tumors are usually sclerosing adenocarcinomas with high fibrotic tissue with low intensity on T1 and T2 sequences that cause stenosis of the common bile duct or abrupt termination at the tumor level that usually shows the shoulder sign, instead of the long and uniform narrowing seen in groove pancreatitis (41, 42). Gastrinoma is the neuroendocrine tumor most frequently located in the groove. These tumors can be differentiated from groove pancreatitis due to the hypervascularity observed in images obtained using contrast enhanced imaging studies, with enhancement of the peripheral ring after the use of gadolinium (10, 43).

Cystic dystrophy of the duodenal wall is characterized by the presence of cysts within the duodenal wall originating from ectopic pancreatic tissue. Imaging findings in cystic dystrophy are very similar to those of groove pancreatitis. So far whether groove pancreatitis and cystic duodenal dystrophy are different conditions or part of the same disease spectrum remains uncertain. Therefore, the term paraduodenal pancreatitis, which includes groove pancreatitis, cystic dystrophy of the duodenal wall, and paraduodenal wall cysts, is used in the literature (36).

CONCLUSION

Groove pancreatitis is a rare form of chronic focal pancreatitis. CT and MRI constitute the diagnostic imaging studies of choice; however, the introduction of biliopancreatic ultrasonography has allowed assessing the pancreas without exposing the patient to radiation and contrast mediums. This case report is intended to highlight the occurrence of groove pancreatitis as a differential diagnosis in acute episodes of pancreatitis that could actually be exacerbations of a chronic condition. Knowing the characteristics of groove pancreatitis favors the physician to make a correct diagnosis using less invasive diagnostic tests and reducing potential risks for the patient.

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