

Cystic dystrophy of the duodenal wall: A rare but need-to-know disease

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ABSTRACT

Cystic dystrophy of the duodenal wall (CDDW) is a serious but uncommon complication of heterotopic pancreatic tissue characterized by increased duodenal wall thickness associated with intraparietal cystic lesions. It is mainly observed in middle-aged male patients with alcoholic chronic pancreatitis. Clinical symptoms are usually nonspecific and it is important to take them into account in patients with the abovementioned history. Imaging techniques have been useful for diagnosis, especially endoscopic ultrasound (EUS), to visualize cystic lesions in the wall. There is some controversy regarding treatment, because although good results have been obtained with surgical techniques, the recent emergence of EUS-guided drainages has also achieved acceptable results and they are suggested as a good alternative to traditional surgery. Following is our experience in the diagnosis and treatment of four patients with CDDWs; all of whom had a history of alcoholism and smoking, and were studied due to clinical signs of abdominal pain and vomiting. EUS was particularly useful in reaching the final diagnosis.

Key words: Cystic dystrophy, chronic pancreatitis, endoscopic ultrasound scan EUS, heterotopic pancreas

INTRODUCTION

Paraduodenal pancreatitis includes inflammatory phenomena that affect the head of the pancreas, the pancreaticoduodenal groove (anatomical space delimited by the dorsal cranial part of the head of the pancreas, the duodenal serosa, and the bile duct) and the second duodenal section. This includes differentiated entities such as cystic dystrophy of the duodenal wall (CDDW), pancreatitis of the groove, pancreatic hamartoma of the duodenum, and duodenal fibrosis.^[1] CDDW is a serious but

uncommon complication, characterized by the presence of pancreatic tissue in an unusual location such as the inert part of the second portion of the duodenum. It occurs as a result of ongoing inflammation which leads to fibrosis and subsequent wall thickening^[1] associated with the presence of intramural cystic lesions.^[2]

It is more common in Caucasian males, aged around 50 years, and in 50-75% of cases according to the literature it is associated with alcoholic chronic pancreatitis,^[2-4] and is diagnosed by imaging techniques such as endoscopic ultrasound (EUS) in particular, as it allows proper viewing of the cysts and treatment by draining where the cysts are few and large.

We report four cases of patients diagnosed with CDDW using EUS performed at our facilities and their evolution.

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CASE REPORT

Case 1

A 34-year-old male with former alcoholic habit of 20 g ethanol per day and smoking history 10 pack-years over 10 years with a history of acute alcohol-induced pancreatitis, with an episode 9 months ago.

This patient was under observation due to 1-year history of postprandial epigastric abdominal pain with vomiting, and 20 kg weight loss. An upper endoscopy was performed showing, in the second portion of the duodenum an edematous mucosa with nodular appearance bulging into the lumen and reducing its size without obstructing the passage of the endoscope [Figure 1].

Subsequently, magnetic resonance imaging (MR imaging) was performed, confirming a thickening of the duodenal wall, in contact with the pancreas which had normal characteristics, associated with a small cystic image in the outer wall [Figure 2].

In view of the diagnostic doubts, a EUS was requested, evidencing atrophic pancreas with heterogeneous lobular parenchyma suggestive of chronic pancreatitis and edema thickening of the duodenal wall, with loss of layer stratification and the presence of multiple cystic areas inside, all of them less than 2cm, suggestive of CDDW [Figure 3].

A conservative medical treatment was decided with opioids and octreotide showing no clinical improvement after 6 months, so EUS was repeated, evidencing a growth of the intramural cysts to 2.5 cm, therefore surgery (cephalic pancreaticoduodenectomy) was chosen. The surgical specimen confirmed the suspected CDDW and the patient is currently asymptomatic.

Case 2

A 55-year-old male patient with alcoholic habit of 25g ethanol per day and smoking history of 30 pack-years over 35 years under consultation for clinical signs of dyspepsia of 6 months with onset of epigastric abdominal pain, nausea, and vomiting during recent weeks.

An upper endoscopy was performed, showing a second duodenal portion with thickened folds of uncertain infiltrative appearance (absence of malignancy on the biopsy), and subsequently confirmed upon completion of a barium meal transit test [Figure 4].

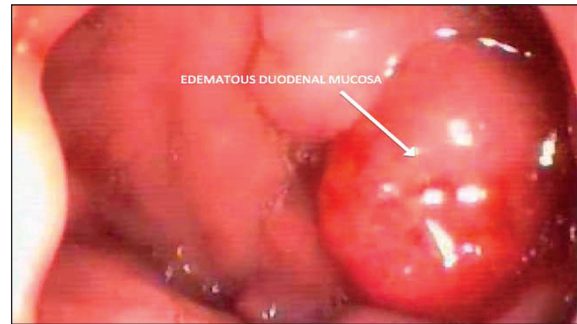


Figure 1. Congestive and edematous duodenal mucosa of nodular appearance with slight narrowing of the lumen size

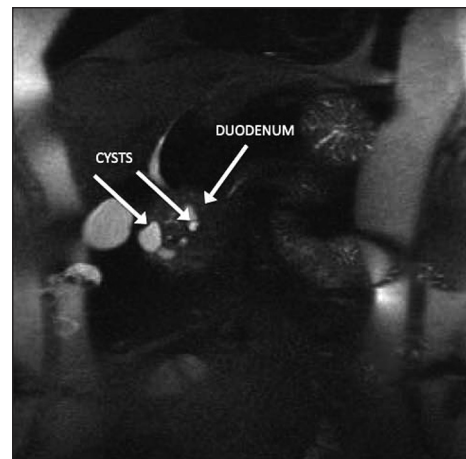


Figure 2. MRI: Thickening of the duodenal frame with associated cystic area. MRI = Magnetic resonance imaging

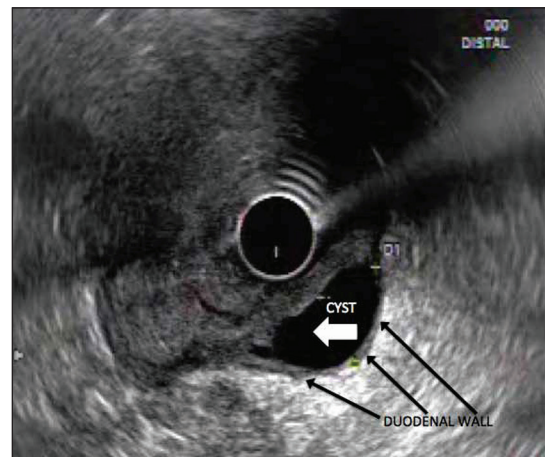


Figure 3. EUS: Thickening of duodenal folds of inflammatory aspect and cystic areas inside the duodenal wall. EUS = Endoscopic ultrasound

An abdominal and pelvic computed tomography (CT) showed a multilocular cystic image adjacent to pancreatic head with no infiltration of vascular structures, encompassing the second duodenal portion and causing stenosis, with peripancreatic lymphadenopathies [Figure 5].

An EUS was performed, showing the duodenal wall circumferentially occupied by an intramural cystic formation with septa inside (multilocular), nondependent on a pancreatic parenchyma with 50 × 41 mm diameter and incipient changes of chronic pancreatitis (indeterminate findings as per Rosemont classification) [Figure 6].

An EUS-guided biopsy of the cyst using a 19-G needle was performed, which achieved almost complete aspiration (amylase 20,000U/L, normal range up to 80) [Figure 7], after which the patient progressed well with significant reduction in the duodenal wall thickness, evidenced by CT; and a further control EUS 3 months later [Figure 8]. Eighteen months later the patient is asymptomatic without treatment.

Case 3

A 50-year-old male smoker with 20 pack-year history over 20 years, with a drinking habit of 30 g ethanol per

day, diagnosed with pancreatic head pseudomass using abdominal ultrasound in view of abdominal pain and constitutional symptoms. For this reason, a EUS was requested, showing parenchymal changes of chronic pancreatitis with pancreatic head pseudomass, probably of inflammatory origin and fine-needle aspiration (FNA) negative for malignancy.

The patient returned to hospital 3 years later, presenting with recurrent vomiting and postprandial fullness, with endoscopic findings of edema and hyperemia at the inner part of the second portion of the duodenum, causing puckering, and reducing the lumen without obstructing the passage of the endoscope, with findings confirmed in a barium meal transit test.

A subsequent abdominal CT identified a cystic tumor measuring 42 × 57 mm in diameter at the duodenobilio pancreatic crossover, imprinting on the duodenum, causing a gastric retention [Figure 9].

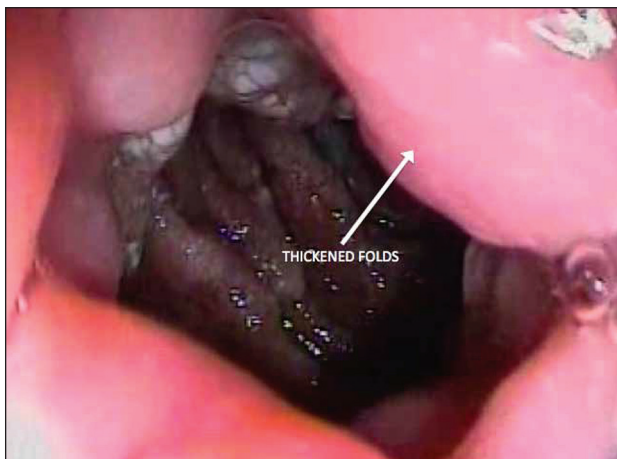


Figure 4. Thickened folds of uncertain infiltrative appearance observed in gastroscopy

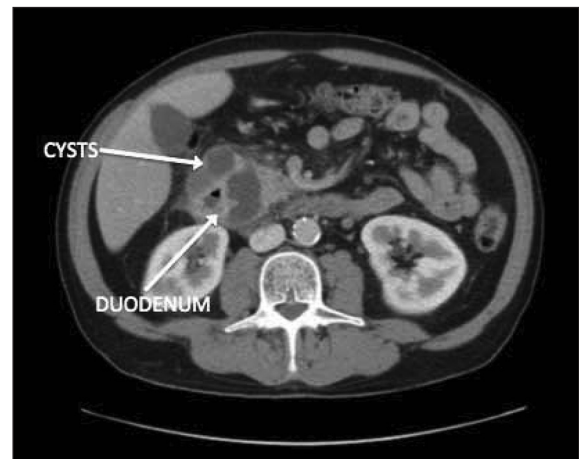


Figure 5. CT: Multilocular cystic area adjacent to the uncinate process. CT = Computed tomography

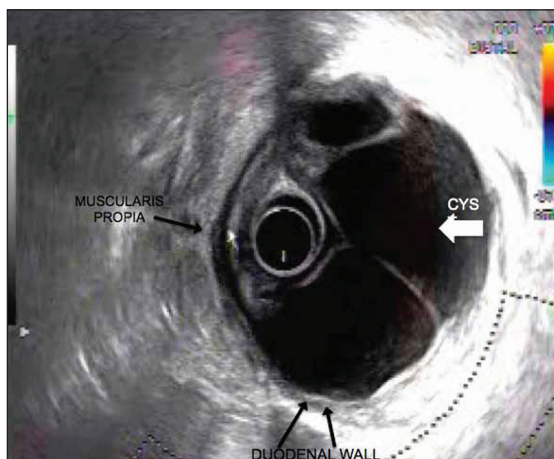


Figure 6. EUS: Circumferential occupation of the duodenal wall due to multilocular cystic collection with septa

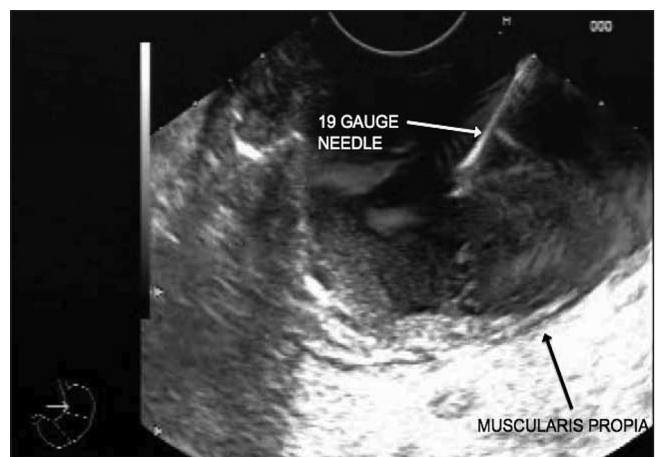


Figure 7. Draining of the cystic collection by EUS.

A EUS was performed, showing a concentric thickening of the duodenal wall at the expense of the submucosa (1.8 mm thick) in the bulb and the beginning of the second portion with several anechoic collections inside the duodenal wall, the largest measuring 26 ´ 42 mm, all suggestive of CDDW [Figure 10].

Aspiration of the cyst was performed guided by EUS (amylase 58, 197U/L normal carcinoembryonic antigen (CEA)) despite which gastric retention persisted; so it was finally decided to opt for bypass surgery by gastrojejunostomy with successful outcome and disappearance of symptoms.

Case 4

A 59-year-old smoker with 30 pack-year history over 30 years without drinking habits presenting for abdominal pain and postprandial vomiting.

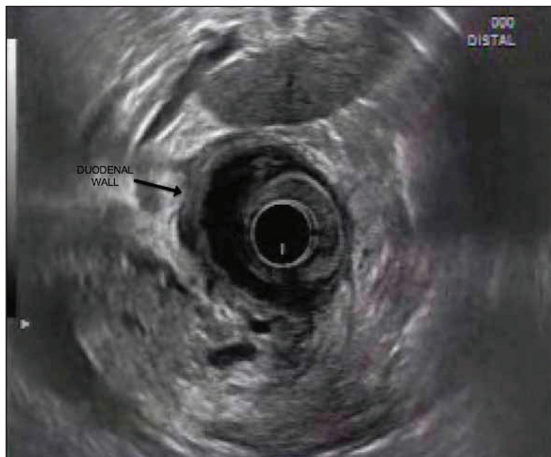


Figure 8. Post-drainage EUS control of intramural cystic collection

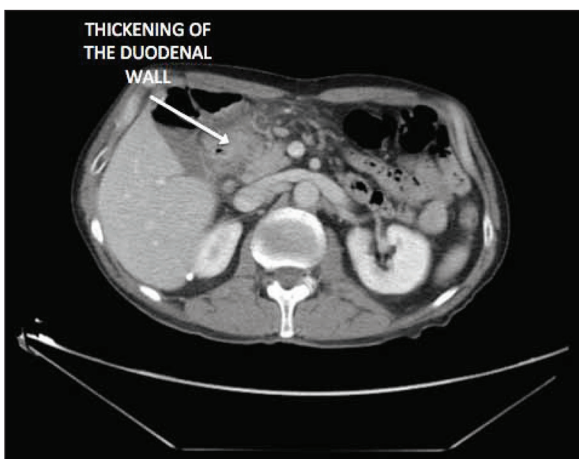


Figure 10. EUS: Duodenal intramural cystic collection

A CT was performed that identified thickening of the wall of the first portion of the duodenum, with trabeculation of duodenal-pancreatic crossover fat [Figure 11]. An EUS was requested, showing findings consistent with chronic pancreatitis according to Rosemont criteria along with a nonstenotic thickened duodenal mucosa with lower intramural cystic area of less than 1 cm [Figure 12].

A conservative treatment was selected, and the patient is currently asymptomatic without treatment.

DISCUSSION

Although cystic dystrophy of heterotopic pancreas was described as early as the 70s, it is still a rare and under diagnosed condition. It almost exclusively affects the duodenal wall, and is characterized by the development of cysts in the submucosal heterotopic

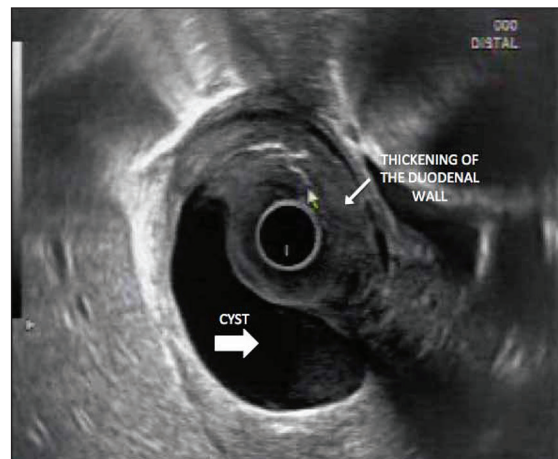


Figure 9. CT: Duodenal-pancreatic crossover fluid collection with compression of the duodenal lumen

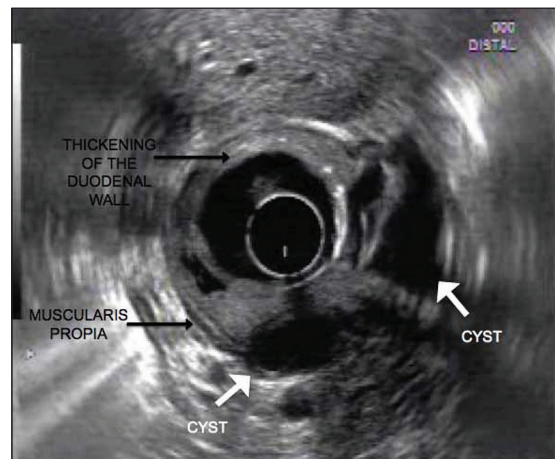


Figure 11. CT: Thickening of the wall of the first duodenal portion, with duodenal-pancreatic fat trabeculation

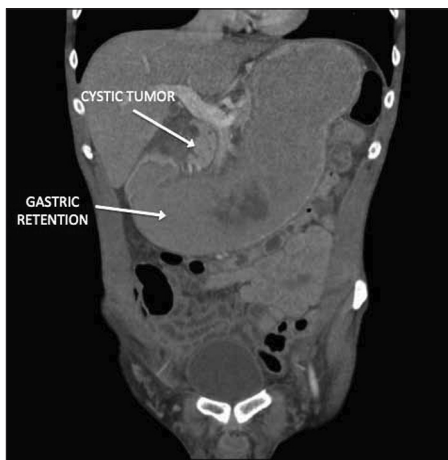


Figure 12. EUS: Intramural cystic image of less than 1 cm

pancreatic tissue located in the duodenal wall, which are surrounded by fibrotic pancreatic tissue, and shows signs of inflammation. It is usually associated with alcoholic chronic pancreatitis in 50-75% of cases, and is diagnosed mostly in middle-aged males, like our patients.^[2-6] During the last decade, the number of cases reported has increased; thanks to the appearance and development of new techniques that allow us to observe in detail the wall of the duodenum.^[2,3,5,6]

Clinical signs are diverse and nonspecific in many cases, going from asymptomatic patients to patients showing signs of intestinal obstruction. It is usually associated with epigastric pain, nausea, vomiting and weight loss, which are symptoms of chronic pancreatitis and are caused by inflammation and fibrosis of the heterotopic pancreatic tissue.^[2,7]

Diagnosis is difficult for several reasons. On the one hand, this condition does not show any typical clinical manifestations, and therefore on diagnosis many of the patients have a history of symptoms dating back many months or years.^[7] And, given the location of this condition, in some cases the differential diagnosis with other pathologies arises, particularly chronic pancreatitis and pancreatic neoplasia, as seen in our patients. On the other hand, the characterization and cytological study of cystic lesions is difficult, although in recent years the emergence of EUS has minimized this problem, and has been a breakthrough for diagnosis.^[2,8] Currently, diagnosis is based on radiological and endoscopic imaging techniques that allow us to observe a thickened duodenal wall with intramural cysts.^[7-9]

CT results analyzed in two retrospective studies of 30 cases^[9,10]; has proved to be a good diagnostic method,

offering the possibility of visualizing the thickened duodenal wall with cysts inside, and has shown a good correlation with the anatomic and pathologic findings. It also allows us to see indirect findings of chronic pancreatitis to enable a differential diagnosis with pancreatic pseudocysts.^[6]

An MRI can provide useful information about the cysts in the duodenal wall, and show heterotopic pancreatic tissue with lower signal intensity than the normal pancreas on T2-weighted images, that after gadolinium show a marked contrast between the pathological ectopic tissue and native pancreas in T1.^[5,6] It also allows us to visualize the biliary and pancreatic tract noninvasively.^[6]

Lastly, EUS is the most accurate technique for diagnosis.^[2,3,6,11] It identifies the duodenal wall cysts as hypoechoic structures dependent on the duodenal submucosa and muscularis propria^[2,8] associated with a thickening of the duodenal wall and an existing network of canals around the cysts.^[9] In addition, the possibility of EUS-FNA cytology aids in the differential diagnosis of this condition.^[2]

In our opinion, the best diagnostic strategy is the combination of MRI with an echoendoscopy. The first allows a noninvasive study of the bile and pancreatic duct (MR cholangiogram) as well as eliminate their communication with the pancreatic duct in cases of differential diagnoses with cystic tumors of the pancreas. The EUS allows not only confirmation of the intraparietal location of the cysts, but also a definitive diagnosis via FNA biopsy (FNAB) and the therapeutic approach of the selected cases.

In our study, the patients presented to a greater or lesser degree associated with chronic pancreatitis. Although the definitive diagnosis was only established through a histology in one case (case 1), in all other cases the presence of cysts could be identified in the interior of the duodenal wall via EUS, which along with elevated levels of amylase in the aspiration biopsy (cases 2 and 3) allowed for the confirmation of the diagnosis of CDDW, and ruled out other potential causes such as periduodenal pseudocysts or cysts with duodenal duplication.

Treatment is controversial, and currently there is no established consensus about the most appropriate treatment.^[2,11] Medical treatment with octreotide

has been questioned in recent years due to its high cost, delayed effects, and variable efficacy in pain control.^[5,11] Surgical procedures are typically used to provide long-lasting results.^[11] Resecting surgery by pancreaticoduodenectomy is the procedure of choice in cases where there is uncertainty with an associated neoplasia;^[2] besides from resecting the area of suspected malignancy, it can confirm the diagnosis. Bypass surgery such as a gastrointestinal anastomosis is the most frequent technique in cases of obstruction, allowing relief of symptoms with lower rates of morbidity and mortality.^[11] Case 1 was treated by resection surgery; whereas, case 3 presented a duodenal obstruction, so bypass surgery was chosen.

Endoscopic therapy is an alternative treatment with low morbidity, but for which there is little literature published. The technique can involve a simple biopsy and aspiration of the cysts, or fenestration with a needle-knife, or drainage via placement of prosthesis in cases of large, sparse, and scattered cysts.^[2,5,11] The endoscopic therapy guided by EUS can eliminate or improve symptoms, but with subsequent relapses according to the published literature.^[2,5,11] Some authors advocate the use of this technique as a good alternative for patients who have not achieved complete alcohol abstinence.^[3]

In conclusion, CDDW remains a poorly understood disease, often under diagnosed, for which EUS has been proven to be the most useful technique for reaching a diagnosis, as occurred in our three patients, and for which the best treatment option has not yet been well established.

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