Cystic dystrophy of the duodenal wall associated with chronic alcoholic pancreatitis

Clinical features, diagnostic procedures and therapeutic management in a retrospective multicenter series of 23 patients

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SUMMARY

Aim of the study — The aim of this retrospective multicenter study was to collect data from patients with chronic alcoholic pancreatitis and cystic dystrophy of the duodenal wall in order to better understand the outcome after medical, endoscopic and/or surgical treatment.

Patients and methods — The data from medical records of 23 patients consecutively seen in ten primary referral centers from January 1990 to July 2004 were studied. Clinical, biological, and endoscopic features as well as imaging findings were recorded. Response to treatment was noted.

Results — Twenty-three patients (20 men), aged 45 years (range: 30-66), with chronic alcohol intake, cystic dystrophy of the duodenal wall, and previously known (N = 14) or simultaneously diagnosed (N = 9) chronic pancreatitis were included. Symptoms most frequently encountered were abdominal pain (N = 22) and weight loss (N = 16). An abdominal ultrasound was available for 10 patients, abdominal computed tomography for 22, upper endoscopy for 18, and endoscopic ultrasonography for 22. Endoscopic ultrasonography enabled diagnosis of cystic dystrophy of the duodenal wall in 19/22 patients. Six patients were symptom-free after alcohol withdrawal. Seven patients received octreotide 200 to 400 µg per day, 5 of whom subsequently underwent surgery (71%). Fourteen patients out of 23 were operated on (61%), 11 of whom underwent pancreaticoduodenectomy and remained symptom-free for 47 months follow-up. Mean follow-up was 56 months (range: 12-108) for surgical patients (61%).

Conclusion — Cystic dystrophy of the duodenal wall complicating chronic alcoholic pancreatitis may be the revealing sign of pancreatitis. Endoscopic ultrasonography is the most reliable imaging method for diagnosis. Pancreaticoduodenectomy is the most frequently employed definitive treatment.

RÉSUMÉ

Dystrophie kystique de la paroi duodénale associée à une pancréatite chronique alcoolique. Aspects cliniques, diagnostiques et thérapeutiques de 23 cas

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Objectif — Le but de cette étude multicentrique rétrospective menée au sein de l’Association Nationale des Hépato-Gastroentérologues des Hôpitaux Généraux (ANGH) était de colliger les observations de dystrophie kystique de la paroi duodénale sur pancréas aberrant chez des malades ayant eu une pancréatite chronique (PC) d’origine alcoolique, en mettant l’accent sur l’évolution clinique après traitement médical, chirurgical ou endoscopique.

Malades et méthodes — Les données provenant des dossiers de 23 malades suivis de janvier 1990 à Juillet 2004 dans dix services d’Hépato-Gastroentérologie de centres hospitaliers généraux, ont été colligées. Les caractéristiques cliniques, biologiques, endoscopiques et d’imagerie d’une part et les modalités thérapeutiques d’autre part sont rapportées. Le suivi après traitement médical, endoscopique et chirurgical est exprimé en médiane [extrêmes].

Résultats — Vingt-trois malades (20 hommes) âgés de 45 ans [30-66], ayant une consommation excessive d’alcool et une pancréatite chronique connue (N = 14) ou diagnostiquée de façon concomitante (N = 9) ont été étudiés. La présentation clinique était dominée par les douleurs abdominales (N = 22) et l’amaisissement (N = 16). Dix malades ont eu une échographie abdominale, 22 un examen tomodensitométrique abdominal, 18 une endoscopie digestive haute, et 22 une echo-endoscopie ayant permis le diagnostic chez 19 malades. Six des onze malades sevrés étaient asymptomatiques. Sept malades ont été traités par octréotide 200 à 400 µg par jour dont 5 (71%) ont dû être opérés secondairement. Parmi les 14 malades opérés (61%) 11 ont eu une duodéno-pancréatéctomie céphalique qui a permis la disparition des manifestations cliniques à long terme dans tous les cas avec un recul de 47 mois [12-108]. Le suivi a été de 56 mois [2-78] chez les malades qui n’ont pas été opérés (9/23) et de 47 mois [12-108] chez ceux qui ont été opérés (14/23).

Conclusion — La dystrophie kystique de la paroi duodénale compliquant la pancréatite chronique d’origine alcoolique en est parfois révélatrice ; l’examen diagnostique le plus performant est l’échoendoscopie.
Introduction

Cystic dystrophy of the duodenal wall in relation to heterotopic pancreas is a very uncommon condition described for the first time by Potet and Duclet in 1970 [1]. The characteristic feature is the presence of true cysts in the duodenal wall which develop from foci of heterotopic pancreatic lying close to the anatomic pancreas. The disease is generally disclosed by signs of luminal obstruction affecting the upper digestive tract (vomiting, delayed transit...) or by signs of acute pancreatitis. Most of the symptoms are also compatible with chronic alcoholic pancreatitis which is associated in the majority of patients [2]. Cystic dystrophy of the duodenal wall has also been observed in young patients with a healthy pancreas [3, 4].

Imaging explorations, particularly endoscopic ultrasonography (EUS) demonstrate the presence of cystic formations in a thickened duodenal wall [5-7]. Pancreaticoduodenectomy is usually proposed for symptomatic patients [1, 8-10]. Certain authors have however proposed a medical approach using octreotide [6, 11, 12] or endoscopic treatment [13] for small series or individual patients.

The purpose of this retrospective multicentric study was to collect data on consecutive cases of cystic dystrophy of the duodenal wall in patients with chronic alcoholic pancreatitis in ten primary referral centres participating in the French national association of general hospital hepatogastroenterologists (ANGH, l’Association Nationale des Hépato-Gastroentérologues des Hôpitaux Généraux). The goal was to describe the clinical, biological, and endoscopic features and collect data on imaging findings as well as therapeutic practices and mid-term response to treatment.

Patients and methods

Patients

All consecutive patients presenting diagnostic criteria for chronic alcoholic pancreatitis associated with cystic dystrophy of the duodenal wall with heterotopic pancreas hospitalized between January 1990 and December 2002 in ten hepatogastroenterology units of ANGH-participating hospitals were included in this study. During this period, all patients with cystic dystrophy of the duodenal wall with heterotopic pancreas seen in these units also had chronic alcoholic pancreatitis.

Diagnosis of chronic pancreatitis

Several criteria were used to establish the diagnosis of chronic pancreatitis [14]:

— presence of calcifications in the pancreatic region on plain x-rays of the abdomen (anteroposterior or oblique view);
— presence of pancreatic calcifications and/or characteristic morphological changes visualized by abdominal ultrasound, computed tomography, endoscopic retrograde cholangiopancreatography or EUS;
— pathological diagnosis on a surgical specimen.

An alcoholic etiology of chronic pancreatitis was retained when alcohol intake was ≥ 60 g/d during the preceding five years [8] and no other cause of chronic calcifying pancreatitis could be identified (metabolic disorder, ductal obstruction, prior radiation of the abdomen, familial history of chronic pancreatitis...).  

Diagnosis of cystic dystrophy of the duodenal wall

The diagnosis of cystic dystrophy of the duodenal wall was retained on the basis of:

— morphological explorations alone (N = 12 patients): 1) EUS demonstrating presence of cystic formations of variable size in the fourth hypoechogenic layer potentially associated with duodenal wall thickening [5, 6]; 2) abdominal computed tomography [15] in one patient who did not undergo EUS.

- pathological examination of surgical specimen of the duodenum (N = 11); gross aspect showing cystic dilatations within the duodenal wall, generally in the muscularis propria and sometimes the submucosa in the vicinity of the ampulla; histologically, cysts lined with pancreatic secretory epithelium or pseudocysts lined with a polymorphous inflammatory granuloma; presence of heterotopic pancreas near the cysts which could thus be interpreted as dystrophic emanations of ectopic pancreatic tissue [1].

Methods

Clinical and biological data as well as imaging findings and pathology reports were recorded for each patient. Pertinent data on treatment and course following treatment were also collected from the medical files by the physician in charge of the patient.

Clinical data included age, gender, alcohol intake, history of acute pancreatitis, date of diagnosis (chronic pancreatitis and cystic dystrophy of the duodenal wall), presence of acute pancreatitis at diagnosis of cystic dystrophy of the duodenal wall, symptoms (weight loss, vomiting, abdominal pain, jaundice), presence of diabetes and treatment with insulin or not, and presence or not of pancreatic exocrine insufficiency treated with pancreatic enzymes.

The results of laboratory tests were also noted: C-reactive protein, fibrinogen, erythrocyte sedimentation rate, creatinine, electrolytes, total bilirubin, alkaline phosphatase, gamma-glutamyl-transferase.

The results of upper gastrointestinal endoscopy, abdominal ultrasound and computed tomographic explorations as well as EUS were noted. The corresponding documents were re-read by the clinicians and radiologists in charge of the patients.

Treatments noted included: alcohol withdrawal, drug inhibition of gastric secretion, use of somatostatin analogues (octreotide: Sandostatin®), nutritional support, endoscopic or surgical procedures.

The clinical course after treatment was noted to July 2004. Duration of follow-up after starting treatment was noted as well as the course of pain, vomiting, weight loss and blood glucose level.

Results were expressed as median with range.

Results

Twenty-three patients with cystic dystrophy of the duodenal wall with heterotopic pancreas associated with chronic alcoholic pancreatitis were included in this study.

The diagnosis of cystic dystrophy of the duodenal wall was established on the basis of EUS findings in 19 patients and on the basis of abdominal computed tomography findings in one. Histological examination of the operative specimen after pancreaticoduodenectomy confirmed the diagnosis in eight patients. In three patients, the diagnosis of cystic dystrophy of the duodenal wall was missed by EUS and correctly established at histological examination.

Clinical presentation and laboratory results (table I)

Twenty men and three women were included in this study. Median age was 45 years (range: 30-66) at diagnosis of cystic dystrophy of the duodenal wall. Excessive alcohol intake was noted in all 23 patients. Chronic pancreatitis preceded the diagnosis of cystic dystrophy in 14 (61%) patients for a duration of 36 months on average (range: 12-120). Five of these patients had non-calculifying chronic pancreatitis. The diagnosis of both chronic pancreatitis and cystic dystrophy of the duodenal wall was simultaneous in nine patients (39%).

All patients were symptomatic at diagnosis. Abdominal pain predominated and was noted in 22/23 patients (95%). Weight loss (mean of 13 kg, range: 4-35 kg) was noted in 16 patients (70%), vomiting in 10 (43%). Six patients (27%) had one symptom and 17 had two or three.

Cholestasis without jaundice was observed in six patients (26%), an inflammatory syndrome in three (13%), and functional renal failure subsequent to vomiting in two patients (9%). Seven patients (30%) presented with acute pancreatitis at the time of diagnosis.
Table I. – Clinical and biological features, and imaging findings.

Caractéristiques cliniques, biologiques et morphologiques.

<table>
<thead>
<tr>
<th>Clinical features</th>
<th>Details</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>20 M − 3 F</td>
</tr>
<tr>
<td>Age at diagnosis of cystic dystrophy of the duodenal wall</td>
<td>45 years (30-66)</td>
</tr>
<tr>
<td>Diagnosis of chronic pancreatitis known at time of diagnosis of cystic dystrophy of the duodenal wall</td>
<td>14/23 (61%)</td>
</tr>
<tr>
<td>Duration of chronic pancreatitis</td>
<td>3 years (1-10)</td>
</tr>
<tr>
<td>Abdominal pain</td>
<td>22/23 (95.6%)</td>
</tr>
<tr>
<td>Vomiting</td>
<td>10/23 (43.5%)</td>
</tr>
<tr>
<td>Weight loss</td>
<td>16/23 (70.6%)</td>
</tr>
<tr>
<td>Associated acute pancreatitis</td>
<td>7/23 (30.4%)</td>
</tr>
<tr>
<td>Cholestasis without jaundice</td>
<td>6/23 (26.1%)</td>
</tr>
<tr>
<td>Inflammatory syndrome</td>
<td>3/23 (13%)</td>
</tr>
<tr>
<td>Functional renal failure</td>
<td>2/23 (8.7%)</td>
</tr>
</tbody>
</table>

Upper barium study (N = 10)

- Duodenal displacement | 2/10 (20%)
- Duodenal stricture | 4/10 (40%)
- Duodenal compression | 1/10 (10%)
- Thickened duodenal wall | 1/10 (10%)

Upper gastrointestinal endoscopy (N = 18)

- Duodenal stricture | 8/18 (44%)
- Ulcerated bulb | 3/18 (17%)
- Esophagitis | 3/18 (17%)
- Erosive duodenitis | 1/18 (5%)
- Gastritis | 2/18 (11%)
- Waffle-like aspect of the duodenal wall | 2/18 (11%)
- Normal | 1/18 (5%)

Endoscopic ultrasound (N = 22)

- Chronic pancreatitis | 22/22 (100%)
- Duodenal cyst(s) | 19/22 (86%)
- Thickened 2nd duodenum | 4/22 (18%)

Abdominal computed tomography (N = 22)

- Hypertrophy of the head of the pancreas | 7/22 (32%)
- Chronic calcifying pancreatitis | 10/22 (45%)
- Duodenal thickening | 2/22 (9%)
- Duodenal cysts | 6/22 (27%)
- Cyst(s) of the head of the pancreas | 11/22 (50%)
- Thickened antral wall | 1/22 (5%)

**Endoscopic and imaging findings (table I)**

- Upper gastrointestinal endoscopy was performed in 18 patients (70%). An extrinsic compression of the duodenal wall was observed in ten patients (55%) and prevented further progression of the endoscope in one. Associated lesions were noted in nine patients (50%) including: peptic esophagitis (N = 5), ulceration of the duodenal bulb (N = 2), erosive gastritis (N = 2).
— Upper barium study was performed in ten patients (43%). It was normal in one (10%), could not be interpreted in one (10%) and was abnormal in eight (80%). A widened duodenal arch was noted in two patients, complete duodenal stenosis in one, and incomplete stenosis in four. Thickening of the duodenal folds was noted in one patient.

— Abdominal ultrasound was performed in ten patients. The results could not be interpreted in two patients. The examination demonstrated a mass effect in the pancreatic head in three patients (30%), signs of chronic pancreatitis in five (50%), a hypoechogenic zone in the duodenopancreatic region in three (30%) and a thickened duodenal wall in one (10%).

— Abdominal computed tomography was performed in 22 patients with injection of contrast medium. A thickened duodenal wall was observed in two patients (9%) and cystic images in the duodenopancreatic region in 17 (77%). The diagnosis of cystic dystrophy of the duodenal wall with heterotopic pancreatitis was initially suggested in nine of these 17 patients. Images suggestive of chronic pancreatitis (calcifications, heterogeneous tumefaction of the pancreatic head, dilatation of the main pancreatic duct, dilatation of the common bile duct) were also observed in 17 patients (77%).

— Endoscopic ultrasound (EUS) of the biliary and pancreatic regions was performed in 22 of the 23 patients. Passage beyond the genu superius was not possible in one patient. In two patients (9%) EUS failed to reveal any images suggestive of chronic pancreatitis. In one patient (4%) EUS showed isolated thickening of the second duodenum in addition to signs of chronic pancreatitis. In 19 patients (83%) EUS enabled the diagnosis of cystic dystrophy of the duodenal wall due to the presence of duodenal wall thickening and the presence of hypoechoic cavities within the muscularis propria of the duodenal wall [5]. These data were confirmed histologically on surgical specimens in eight patients.

Treatments

Exclusive medical treatment was given for nine patients (39%) (table II) and medical and surgical treatment in 14 (61%) (table III).

Medical treatment (tables II and III)

Alcohol withdrawal was achieved in 20 patients (87%). These patients were also given minor analgesic drugs (35%) and/or type 2 H₂ receptor blockers (5%) and/or proton pump inhibitors (10%) and/or pancreatic extracts (5%). Three patients (13%) required prolonged morphine. Parenteral nutrition was administered in four patients (17%) as oral intake was not tolerated.

Among the patients abstinent for alcohol, four (20%) did not experience recurrent pain and remained pain-free with no other treatment for 36 to 67 months. Five patients had a morphological control (EUS for four and computed tomography for one) which revealed a reduction in the cystic dystrophy in three (EUS and computed tomography) and its stability in two (EUS).

Alcoholic intake continued in three patients (13%) who remained symptomatic and required analgesics. Surgery was not performed in these three patients.

Octreotide (Sandostatin®) was administered subcutaneously (200-400 µg b.i.d. for three months) in seven patients (35%). Six became abstinent and one was lost to follow-up. One patient remained symptom-free 87 months after treatment onset. Five patients underwent secondary surgery; one month after treatment onset for two and at 8 to 28 months for the other three.

Endoscopic treatment

Two patients were treated by endoscopic fenestration of the cysts. When recurrent symptoms developed 18 and 20 months later, cystojejunal deviation was performed in one and cephalic pancreatico-duodenectomy in the other.

Surgical treatment (table III)

Fourteen patients underwent surgery (61%). Surgery was performed early in three patients within three months of diagnosis. Half of the operated patients (N = 7) had received octreotide (N = 5) or endoscopic (N = 2) treatment. Mean time between medical management and surgery in patients who were not given a specific treatment prior to surgery was 12 months (range: 1-13).

Gastroenterostomy was performed in one patient followed later by cephalic pancreatico-duodenectomy. A biliary and digestive bypass was performed in one patient who remained symptom-free for 55 months. A cysto-jejunal bypass using a Y-loop was fashioned in three patients, one of whom later underwent cephalic pancreatico-duodenectomy. Surgery relieved symptoms in the other two patients, completely in one who died from lung cancer at 14 months and partially in the other who died from uterine cancer at 62 months.

<table>
<thead>
<tr>
<th>Patient — treatment</th>
<th>Follow-up (months)</th>
<th>Alcohol withdrawal</th>
<th>Efficacy</th>
<th>Exploration during follow-up</th>
</tr>
</thead>
<tbody>
<tr>
<td>N° 3 Analgesics</td>
<td>36</td>
<td>Yes</td>
<td>Yes</td>
<td>EUS</td>
</tr>
<tr>
<td>N° 4 Analgesics + anafranil</td>
<td>56</td>
<td>Yes</td>
<td>Yes</td>
<td>CT</td>
</tr>
<tr>
<td>N° 5 Analgesics</td>
<td>24</td>
<td>No</td>
<td>No</td>
<td>EUS</td>
</tr>
<tr>
<td>N° 6 Analgesics</td>
<td>78</td>
<td>Yes</td>
<td>No</td>
<td>—</td>
</tr>
<tr>
<td>N° 13 Analgesics</td>
<td>67</td>
<td>Yes</td>
<td>Yes</td>
<td>EUS</td>
</tr>
<tr>
<td>N° 14 Analgesics + Octreotide</td>
<td>87</td>
<td>Yes</td>
<td>Yes</td>
<td>EUS</td>
</tr>
<tr>
<td>N° 16 Anti H2 + PPI</td>
<td>44</td>
<td>No</td>
<td>Non</td>
<td>—</td>
</tr>
<tr>
<td>N° 17 Anti H2 + PPI</td>
<td>48</td>
<td>Yes</td>
<td>Yes</td>
<td>EUS</td>
</tr>
<tr>
<td>N° 20 Analgesics + Octreotide</td>
<td>2</td>
<td>No</td>
<td>No</td>
<td>CT</td>
</tr>
</tbody>
</table>

Anti H2: Anti-histaminic H2; PPI: Proton Pump Inhibitor; EUS: Endoscopic ultrasound; CT: computed tomography.
This type of presentation may however be underestimated since symptoms of cystic dystrophy can readily be attributed to chronic pancreatitis. Examining series of chronic pancreatitis patients shows that the frequency of cystic dystrophy of the duodenal wall is to the order of 25% [15, 19, 25].

Characteristic features of our population are similar to those reported by others [2, 7, 18, 30]: strong male predominance (sex ratio: 6.6), mean age 45 years (range: 30-66), abdominal pain as principal symptom (95%), and frequent weight loss (70%) and vomiting (43%). Fourteen of the 23 patients (61%) were known to have chronic pancreatitis at the time of diagnosis of cystic dystrophy of the duodenal wall. The proportion of simultaneous diagnoses was not reported by Rebours et al. [18], but it was probably high since chronic pancreatitis had been known for a median three months at the time of diagnosis of cystic dystrophy of the duodenal wall.

Upper EUS was performed in 22/23 patients in this series and established the diagnosis in 19 of them (86%). Considered probable before surgery in the operated patients, the diagnosis was confirmed in all eight at pathological examination of the operative specimen. The three patients whose EUS failed to establish the diagnosis underwent pancreaticodudenectomy. These results confirm the predominant role of EUS for the diagnosis and its potential contribution to treatment [27]. Biliopancreatic EUS is currently the exploration of choice to search for cystic dystrophy of the duodenal wall [5, 6]. The diagnosis is confirmed in the presence of tissue thickening, generally observed on the inner border of the second portion of the duodenum, associated with cystic formations found predominantly in the fourth hypochoicogenic layer of the duodenal wall and/or a ductal network sometimes leading to the intestinal lumen.

Computed tomography, proposed by certain authors [28], also enables an evaluation of pancreatic lesions. The diagnosis of cystic dystrophy of the duodenal wall was not however retained in nine patients in our series, confirming early reports...

Table III. – Outcome after surgical treatment.

<table>
<thead>
<tr>
<th>Patient, prior treatment</th>
<th>Delay (months)</th>
<th>Surgical treatment</th>
<th>Revision procedure</th>
<th>Follow-up (months)</th>
<th>Efficacy</th>
</tr>
</thead>
<tbody>
<tr>
<td>1- Alcohol withdrawal, Creon®, PPI</td>
<td>7</td>
<td>CPD</td>
<td>No</td>
<td>36</td>
<td>Yes</td>
</tr>
<tr>
<td>2- Alcohol withdrawal, octreotide</td>
<td>8</td>
<td>CPD cysto-jejunal drainage</td>
<td>No</td>
<td>68</td>
<td>Yes</td>
</tr>
<tr>
<td>7- Alcohol withdrawal, cystoduodenostomy</td>
<td>18</td>
<td>drainage</td>
<td>No</td>
<td>19</td>
<td>Partial</td>
</tr>
<tr>
<td>8- Alcohol withdrawal, analgesics</td>
<td>13</td>
<td>CPD</td>
<td>No</td>
<td>72</td>
<td>Yes</td>
</tr>
<tr>
<td>9- Alcohol withdrawal, analgesics</td>
<td>1</td>
<td>gastroenterostomy</td>
<td>CPD</td>
<td>25</td>
<td>Yes</td>
</tr>
<tr>
<td>10- Alcohol withdrawal, analgesics</td>
<td>12</td>
<td>cysto-jejunal drainage</td>
<td>CPD</td>
<td>62</td>
<td>Yes</td>
</tr>
<tr>
<td>11- Cystoduodenostomy</td>
<td>20</td>
<td>CPD</td>
<td>No</td>
<td>103</td>
<td>Yes</td>
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<td>12- Alcohol withdrawal, analgesics</td>
<td>12</td>
<td>CPD</td>
<td>No</td>
<td>108</td>
<td>Yes</td>
</tr>
<tr>
<td>15- Alcohol withdrawal</td>
<td>11</td>
<td>CPD</td>
<td>No</td>
<td>12</td>
<td>Yes</td>
</tr>
<tr>
<td>18- Cystoduodenostomy</td>
<td>12</td>
<td>cysto-jejunal drainage</td>
<td>No</td>
<td>14</td>
<td>Yes</td>
</tr>
<tr>
<td>19- Alcohol withdrawal, octreotide</td>
<td>12</td>
<td>double bypass</td>
<td>No</td>
<td>55</td>
<td>Yes</td>
</tr>
<tr>
<td>21- Octreotide</td>
<td>1</td>
<td>CPD</td>
<td>No</td>
<td>47</td>
<td>Yes</td>
</tr>
<tr>
<td>22- Octreotide</td>
<td>28</td>
<td>CPD</td>
<td>No</td>
<td>24</td>
<td>Yes</td>
</tr>
<tr>
<td>23- Octreotide</td>
<td>1</td>
<td>CPD</td>
<td>No</td>
<td>15</td>
<td>Yes</td>
</tr>
</tbody>
</table>

CPD = Cephalic Pancreatico-Duodenectomy.

Discussion

This study of 23 patients with cystic dystrophy of the duodenal wall with heterotropic pancreas is one of the largest in the literature [8, 16-19]. Unlike the former reports, it was not limited to surgical cases [4, 8, 16, 17]. It is also illustrative of the treatments proposed and their mid-term impact (mean follow-up 47 months, range: 2-108 months). Two types of cystic dystrophy of the duodenal wall have been described. The most common presentation is cystic formations found predominantly in the fourth hypochoicogenic layer of the duodenal wall and/or a ductal network sometimes leading to the intestinal lumen.
Magnetic resonance imaging (MRI) of the biliary and pancreatic regions was not performed in our patients but would probably be useful as a non-invasive diagnostic tool [29, 30]. MRI can visualize cystic formations and a band of low-intensity signals on T1 and T2 sequences lying between the second portion of the duodenum and the pancreas and corresponding to heterotopic pancreatic tissue [29]. Diagnostic accuracy, particularly in comparison with EUS, remains to be determined.

In this series, medical management was proposed first, followed by surgery if symptoms persisted. Surgery was performed early after diagnosis in only one patient due to the severity of the clinical presentation. Besides non-specific treatment with abstention from alcohol successfully achieved in four patients, management practices included: octreotide, endoscopic fenestration of the cysts, and surgery. Surgery was performed in 61% of patients, 78% of whom had pancreaticoduodenectomy, and provided sustained symptom relief in all.

By analogy with treatments proposed for pseudocysts of the pancreas [31], several authors have tried octreotide with success in patients with cystic dystrophy of the duodenal wall [6, 12, 32]. Others have reported failures with octreotide [33, 34]. Palazzo et al. [11] reported improvement in all seven patients treated with octreotide but with only 90 days follow-up. Rebours et al. reported, in an abstract form, on a series of 109 patients [18] which included nine treated with octreotide but no details on long-term outcome were provided. Among the seven patients in the present series given octreotide, one abstinent patient remained symptom-free for 87 months, one non-abstinent patient developed recurrent symptoms, and five underwent surgery after recurrence of symptoms 0 to 25 months following interruption of the octreotide regimen. At the present time, the probability of response to octreotide cannot be predicted. Frequent association with chronic pancreatitis and the similarity of the symptoms makes it very difficult to identify predictive factors.

Endoscopic treatment by cystogastrostomy has been proposed [35] after failure of medical treatment. This approach is similar to that proposed for pancreatic pseudo-cysts [36] accessible endoscopically. Ponchon et al. [13] treated ten patients with both cystic dystrophy of the duodenal wall and chronic pancreatitis by endoscopic fenestration following injection of contrast agent to opacify the cyst and obtained the following results: immediate failure (N = 1), early failure at 4 and 10 days (N = 2), late recurrence at 14 and 117 weeks (N = 2), and sustained improvement for 108 weeks (N = 5). Beaulieu et al. [27] treated three patients endoscopically, using a pigtail drain for two of them. For the third patient who was treated for recurrence 15 months later, a new drainage procedure with insertion of an internal drain was successful. Cystoduodenostomy was performed endoscopically in two patients in the present series; one underwent surgical cystojejunal bypass 18 months later and the other pancreaticoduodenectomy 20 months later. These results illustrate the risk of recurrence after cystoduodenostomy which does not remove the diseased tissue. These cases of resorting to surgery should not however be considered as treatment failures since further endoscopic sessions for drainage could have been performed. Endoscopy can however only be proposed for selected patients with appropriate morphological characteristics such as one or two very large cysts assumed to be the only cause of the bulging mucosa.

In the past, pancreaticoduodenectomy was widely used for diagnostic and therapeutic management of cystic dystrophy of the duodenal wall, but with improved preoperative imaging, it is being used less frequently as a first-line option. Rebours et al. [18] reported that only 27% of their 109 patients underwent surgery, two-thirds with pancreaticoduodenectomy and the other third for bypass procedures. Surgery was performed in 14 of the 23 patients in the present series, eleven had pancreaticoduodenectomy either as a first intention procedure (N = 4), or as a secondary option after octreotide (N = 4), endoscopic treatment (N = 1), gastroenterostomy (N = 1), or cystojejunal drainage (N = 1). All eleven patients remained symptom-free for a median of 47 months follow-up. A cystojejunal drainage was fashioned in three patients; one later required pancreaticoduodenectomy and pain was only partially relieved in one other. The third patient later had a double bypass which successfully relieved symptoms with a 55 months follow-up. Regarding outcome of the different surgical options, pancreaticoduodenectomy remains the most effective since the diseased tissue is removed. The other procedures are designed to relieve symptoms by draining the cyst or fasting the biliary or intestinal bypass but cannot prevent recurrent flare-ups as was observed in two of our patients.

Nevertheless, pancreaticoduodenectomy is a major operation with high morbidity (risk of diabetes, exocrine insufficiency) and significant mortality (1-5%) [37]. Many thus hesitate to propose pancreaticoduodenectomy for a benign though serious condition, preferring less aggressive options. Administration of somatostatin analogs is proposed by some although there is no way to recognize potential responders. Endoscopic fenestration, preferably with internal drainage, should be proposed initially for patients who have been unable to abstain sufficiently from alcohol intake. In light of the different alternatives proposed, it would be useful to conduct prospective studies in a large number of patients in order to determine the appropriate indications for each. When surgery is needed, pancreaticoduodenectomy should be preferred, preserving bypass procedures for patients with excessive risk. It is also important to recall that remission is inevitably compromised if alcohol withdrawal is not achieved.

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