Cystic dystrophy of the duodenal wall
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Section: Abdominal imaging
Area of Interest: Abdomen Case Type: Clinical Cases
Authors: Arora A, Mukund A, Thapar S, Jain
Department of Radiodiagnosis, Institute of Liver and Biliary Sciences, New Delhi, India
Patient: 30 years, male

Clinical History:
A 30-year-old male patient presented with history of recurrent episodes of upper abdominal pain in the past and mild gradually progressive jaundice. He had no history of smoking or alcohol abuse.

Imaging Findings:
Unenhanced axial CT sections of abdomen revealed changes of chronic calcific pancreatitis with dilated suprapancreatic common bile duct. Additionally cystic lesions along the medial duodenal wall were seen, indenting the duodenal mucosa. Intrinsic contour bulge was seen along the luminal aspect of D-2 duodenum, however, there was no suggestion of upstream bowel dilatation. Remaining abdomen was unremarkable.

The imaging findings favored cystic dystrophy of the duodenal wall with chronic calcific pancreatitis. In lieu of clinical details, the patient was suspected to have genetic pancreatitis, as he had no predisposing factors (no history of smoke or alcohol abuse). Accordingly the patient was referred to a higher centre for relevant genetic screening tests (SPINK), which resulted negative. The patient is being treated as a case of idiopathic chronic pancreatitis.

Discussion:
Cystic dystrophy of duodenum is a sub-type of paraduodenal pancreatitis. Paraduodenal pancreatitis is an umbrella term used to encompass three entities with overlapping clinico-radiological features including: “groove pancreatitis”, “cystic dystrophy of the duodenum”, and “para-duodenal wall cyst” [5].

Cystic dystrophy of duodenum is characterised by the development of true cysts within the heterotopic pancreas located in the gut wall [3, 4]. Most commonly this heterotopic pancreatic tissue is seen along the duodenum in close vicinity to the pancreas. However, cystic dystrophy has also been reported in the gastric and jejunal walls. Histopathology of the involved segment of gut reveals cystic changes within the thickened submucosa or muscularis layer. The cysts are usually lined with cuboid epithelium similar to those lining pancreatic ducts [3, 4]. Additionally, ectopic pancreatic tissue with signs of chronic pancreatitis is seen.

Although cystic dystrophy has been rarely reported in young patients with healthy pancreas; most of the patients have underlying chronic calcific pancreatitis [1]. It is predominantly seen in male patients, 40-50 years old, with a history of chronic alcohol abuse [6]. These patients usually seek medical attention for complains pertaining to chronic pancreatitis. However, progressive scarring fibrosis and luminal compromise of the duodenum can present with features of gastric outlet obstruction. Obstructive jaundice due to distal common bile duct compression has also been reported.

Endoscopic ultrasonography (EUS) is considered the preferred imaging modality for the diagnosis of cystic
dystrophy of pancreas [1, 2]. EUS excellently delineates the intramural location of the cysts within the thickened duodenal wall of the first and the second part of duodenum. Associated chronic pancreatic parenchymal changes and main pancreatic duct dilatation can also be well visualised at EUS. CT and MRI are also very useful in demonstrating the presence of cysts in a thickened duodenal wall [2, 3, 4]. MRCP can aid in the diagnosis by displaying cystic changes along the duodenal wall and in the pancreatico-duodenal groove. Duodenal cystic changes can be associated with groove pancreatitis i.e. simultaneous fibrotic-tissue and cysts within the pancreatico-duodenal groove (the space between the duodenum, pancreatic head and the common bile duct). It is still debatable whether groove pancreatitis and cystic dystrophy of the duodenum are distinct entities or part of the same spectrum.

Pancreatoduodenectomy is considered the preferred treatment especially in patients with duodenal fibrosis and narrowing, and those in whom medical treatment or endoscopic drainage has failed [6, 7].

Differential Diagnosis List: Cystic dystrophy of the duodenal wall in chronic pancreatitis, Cystic neoplasm of pancreatic head, Duodenal duplication cyst

Final Diagnosis: Cystic dystrophy of the duodenal wall in chronic pancreatitis

References:


Egorov VI et al (2010) Pancreas-Preserving Duodenal Resections with Bile and Pancreatic Duct Replantation for Duodenal Dystrophy. Two Case Reports. JOP. J Pancreas (Online)11:446-452
Figure 1

Description: Unenhanced CT of abdomen reveals multiple calcific specks in the pancreatic parenchyma consistent with chronic calcific pancreatitis. Origin:
Description: CT findings are consistent with chronic calcific pancreatitis. Origin:
Description: CT depicts hypoattenuating cysts along the medial wall of duodenum with mild duodenal wall thickening. Origin:
Description: CT depicts hypoattenuating cysts (arrows) along the medial wall of duodenum with mild duodenal wall thickening. Origin:
**Description:** Coronal CT displays a cyst embedded in the medial wall of duodenum causing intrinsic contour bulge and medial displacement of the enhancing duodenal mucosa. **Origin:**
Description: Coronal CT displays a cyst embedded in the medial wall of duodenum causing intrinsic contour bulge and medial displacement of the enhancing duodenal mucosa (arrow). Origin:
**Description:** Few more cysts can be seen embedded in the medial wall of duodenum. **Origin:**
Description: Few more cysts can be seen embedded in the medial wall of duodenum (arrow). Origin:
Description: Intraluminal projection of the cysts and mucosal indentation can be well appreciated on this sagittal image. Origin:
Description: Intraluminal projection of the cysts and mucosal indentation can be well appreciated on this sagittal image (arrows). Origin: