Cystic dystrophy of heterotopic pancreas in duodenal wall

Gandhi Minesh B, Panjvani Sahil I, Shah Cherry K, Shah Nailesh R

SMT. NHL Municipal Medical College, Department of Pathology, Ahmedabad-Gujarat-India

Submission Date: 18-11-2012; Accepted Date: 26-11-2012

ABSTRACT

Cystic dystrophy in heterotopic pancreas is a rare, poorly understood benign disease of gastric and duodenal wall, which was described for the first time by the French authors in 1970, who reported the presence of focal pancreatic disease localized in an area comprising the C-loop of duodenum & the head of pancreas. We report a case of 33 years old male patient with complaint of recurrent abdominal pain with the probable diagnosis of groove pancreatitis on the clinoradiological findings. Whipple surgery was done. Diagnosis of cystic dystrophy in heterotopic pancreas in duodenal wall with chronic pancreatitis was given histopathologically.

Keywords: Cystic dystrophy, heterotopic pancreas, benign, groove pancreatitis, Whipple surgery.

INTRODUCTION

Cystic dystrophy in heterotopic pancreas of duodenal wall is very uncommon entity which was described for first time by Potet & Duclert in 1970. These authors as well as other French authors have described this disorder as the presence of focal pancreatic disease localized in an area comprising the C-loop of duodenum & the head of pancreas. This area was designated as a “groove” by Becker & Mischke in 1991 & they pointed out that it acts as a bed for the large blood vessels, lymphnodes, CBD and main pancreatic duct. The term “groove pancreatitis” was established where associated pancreatitis was present.

Classification of groove pancreatitis was done as “pure groove pancreatitis”, segmental pancreatitis of head and chronic pancreatitis with groove pancreatitis. Recently, the term “paraduodenal pancreatitis” was proposed in patients classified as having “cystic dystrophy of heterotopic pancreas” or “paraduodenal wall cyst” or “groove pancreatitis by Adsay and Zamboni. Moreover additional two types of pancreatitis were established one with cystic changes & the other with solid lesions. The aim of this case report is to briefly describe clinical features, diagnosis, differential diagnosis and pathological features of this rare case of cystic dystrophy of duodenal wall with chronic pancreatitis.

CASE REPORT

A 33 year old male patient was admitted in V.S. General Hospital in Sept 2012 with complaint of acute abdominal pain, nausea and vomiting. He had similar complaints in past. Physical examination revealed mild tenderness in epigastric region. His S. Lipase was 428 U/L (range: 30–210 U/L) and S. Amylase was 126 U/L (range: 30–110 U/L). USG revealed normal gall bladder, dilatation CBD and multicystic lesions in periampulary region.

A CECT of abdomen with pelvis was performed. It showed atrophied pancreatic parenchyma in region of head and proximal body of pancreas. MPD was dilated measuring 12 mm in body region. Circumferentially
enhancing wall thickening was noted in pylorus part of stomach, second part of duodenum and hepatic flexure. Prominent central IHBR and CBD (11 mm) was noted up to distal end with no evidence of calculus within. No evidence of any contrast leak from bowel loops.

A clinical diagnosis of groove pancreatitis was made and Whipple surgery was done. The specimen was sent to histopathology department.

Specimen consisted of panreatoduodenectomy with portion of pylorus. Loop of duodenum measured 20 cm long. Pylorus measured 3 cm. Portion of pancreas measured $4.2 \times 2.4$ cm in size. On cut opening of the duodenum, there was presence of sessile polypoidal lesion in the 2nd part of duodenum measuring $3 \times 2.8 \times 1.8$ cm on pancreatic side of duodenum at and above the Ampulla of Vater. The mucosa overlying the polypoidal lesion was intact. On cut section of polypoidal lesion, multiple cysts containing blood like fluid material were seen. The largest cyst measured $3 \times 2$ cm in size. Cut surface of pancreas was whitish. No evidence of stone was identified. Total 12 lymphnodes were identified from pancreatoduodenal junction.

Multiple sections from polypoidal lesion in the 2nd part of duodenum showed presence of cysts in submucosa and muscular wall of duodenum. The lining epithelium of the cyst was partially eroded and focally lined by columnar epithelium and surrounded by inflammatory granulation tissue, stromal cell proliferation and fibrosis [Figures 3, 4, 5]. At places ectopic pancreatic tissue around the cyst was seen [Figure 6]. There was also presence of marked Brunner's gland hyperplasia [Figure 3].
Section from pancreas revealed changes of chronic pancreatitis. No evidence of pancreatic intraepithelial neoplasms or malignacy was seen. Sections from lymph nodes showed features of reactive node.

**DISCUSSION**

Cystic dystrophy is a disease which is characterized by presence in duodenal or gastric wall of cysts surrounded by inflammation and fibrosis intermingled with pancreatic ducts and lobules. It may be considered as an unusual complication of heterotopic pancreas. Cystic dystrophy in heterotopic pancreas of duodenal wall is an extremely uncommon benign disease. Heterotopic pancreas is defined as presence, outside its usual location, of pancreatic tissue, that lacks anatomic and vascular continuity with the pancreas proper. The incidence rate ranges from 1–14% on necropsy examination, and the most common locations are stomach, duodenum and jejunum. The disease usually affects young male. The characteristic feature is presence of cyst formations & fibrosis in duodenal wall with or without associated pancreatitis.

Underlying mechanism of cystic dystrophy of pancreas is not fully understood. According to Fékété F et al., there are two different types. First is cystic dystrophy in heterotopic pancreas without chronic pancreatitis. It is due to obstructive pancreatitis of heterotopic pancreas by active secretion of pancreatic tissue and progressive duct compression. Second is cystic dystrophy associated with chronic pancreatitis. It is seen in association with chronic alcoholism or chronic pancreatic cell insults. Presence of cystic or solid types is dependent on size of cyst. Easy recognition of cystic type is due to presence of cystic lesions (>1 cm) located within the thickened duodenal wall, while presence of small cysts (<1 cm) within the fibrous thickening of duodenal wall is seen in solid type of cystic dystrophy.

The disease is commonly associated with signs of upper gastrointestinal tract obstruction i.e. abdominal pain, nausea and vomiting, jaundice and weight loss. Sometimes signs of acute pancreatitis are also seen. In majority of the patients, symptoms are compatible with chronic alcoholic pancreatitis with which it is associated. Young patient with healthy pancreas are also be affected with this disease. There is often a chance of misdiagnosis or diagnosis at late stage due to these non specific clinical manifestations. The diagnosis relies on radiologic and endoscopic procedures. Until recently, CT has been used for diagnosis of chronic pancreatitis or adenocarcinoma associated with cystic dystrophy of duodenal wall.

CT and MRI reveal same findings. An important fact of MRI over CT is that it can be followed by MRCP with advantage of no morbidity, which is a drawback in ERCP. MRI is superior to CT in evaluating biliary ducts in para-duodenal pancreatitis as well as in groove carcinomas. Recently, EUS has become a useful technique for accurate evaluation biliary pancreatic structure in diagnosing pancreatic disease. Differential diagnosis of cystic dystrophy includes a pseudocyst.

Surgical resection is the mainstay of treatment with the best result. A pancreatoduodenectomy remains the most effective surgical treatment, as it removes the affected tissue.

In summary, cystic dystrophy in heterotopic pancreas in duodenal wall is a rare lesion. The diagnosis of cystic dystrophy in heterotopic pancreas in duodenal wall is made accurately by specific histopathological features coupled with clinical and imaging features. As a pathologist awareness about this entity is critical in differential diagnosis of cystic lesions of duodenum and in patients present with groove pancreatitis.

**ABBREVIATIONS**

CBD—common bile duct, USG—ultrasonography, CECT—contrast enhanced computed tomography, MPD—main pancreatic duct, IHB—infrahepatic biliary radicles, GB—gall bladder, MRI—magnetic resonance imaging, MRCP—magnetic resonance cholangiopancreatography, ERCP—endoscopic cholangiopancreatography, EUS—endoscopic ultrasonography.
REFERENCES