Cystic Dystrophy in Heterotopic Pancreas of Duodenal Wall —A Case Report—


Department of Pathology, Holy Trinity Hospital, Daegu, Korea
Department of Radiology* and Department of Pathology*, College of Medicine, Yeungnam University, Daegu, Korea

—Abstract—

Cystic dystrophy is an uncommon, benign poorly understood disease. It is characterized by the development of cysts in heterotopic pancreatic tissue. A 57-year-old man was hospitalized for abdominal pain for a week. He is a heavy alcohol drinker. There was a cyst at second portion of duodenum on CT. Under the impression of peptic ulcer perforation, Whipple’s operation was performed. Grossly, a cystic space, measuring 3.0 cm in diameter, was noted within the thickened duodenal wall. Microscopically, the cyst was lined by columnar epithelium and granulation tissue and embedded in ectopic pancreatic tissue. The adjacent pancreatic tissue showed focal chronic pancreatitis.

Key Words: Cystic dystrophy, Heterotopic pancreas, Duodenum

Information

Cystic dystrophy in heterotopic pancreas of duodenal wall is an uncommon, benign disease and first described by Poter and Ducret in 1970.1,2 It was characterized by the development of cysts in heterotopic pancreatic tissue localized in duodenal wall.1,3 The development mechanism of cystic dystrophy is poorly understood.3 On account of nonspecific sign, clinical examination and laboratory finding, diagnosis is very difficult. But this lesion show a cyst containing blood-like fluid within the thickened duodenal wall microscopically and is typical at pathology. Computed tomography and endoscopic ultrasono-
graphy are helpful for diagnosis of cystic dystrophy.\textsuperscript{1,2,4} We report an uncommon typical case and briefly review literature.

Case Report

A 57-year-old man came to other hospital for investigation of abdominal pain and nausea for a week. On CT, there is fluid collection at second portion of duodenum. Under the impression of peptic ulcer perforation, he was transferred to our hospital. There was no melena, hematemesis, or hematochezia. In the past he had consumed large quantities of alcohol daily (Soju 3 bottles/15 years) and had been treated pancreatitis at other hospital. Serum amylase and lipase were elevated to 1014 U/L and 1288 IU/L, respectively.

Follow up ultrasonogram and abdominal CT were performed in our hospital. Ultrasound showed a well marginated 3.7 cm sized cystic mass in duodenal submucosal layers (Fig. 1A). Abdominal CT scans showed

Fig. 1A. Transverse abdominal ultrasonogram shows about 3.7 cm sized cystic mass with internal mixed echogenicity in submucosal layer of second portion of the duodenum.

Fig. 1B. On non-contrast CT scan, the cystic mass (arrow) shows high density representing internal hemorrhage.

Fig. 1C. The mass (arrow) is not enhanced on contrast enhanced CT scan. The 2nd portion of duodenum is mildly compressed by the mass.
round cystic mass lesions in submucosal layer of the 2nd portion of the duodenum. This mass lesion was high density on non-contrast CT scan, which represented internal hemorrhage. The mass was not enhanced after contrast injection. (Fig. 1B, C). The duodenum was mildly compressed and displaced by this masses. Dilatation of pancreatic duct was noted. Localized inflammatory changes were found around duodenum and pancreas. Under impression of submucosal tumor with internal hemorrhagic change or of the peptic ulcer perforation with submucosal hematoma, whipple’s operation was performed.

Macroscopically, a congested bulging lesion was found at serosal surface of the duodenum. On serial sections, a cystic space containing blood-like fluid was noted within the thickened duodenal wall (Fig. 2A). The cyst measured 3.0 cm in diameter and was partly lined by columnar epithelium and inflamed granulation tissue. Heterotopic pancreatic tissue was found in the duodenal wall in adjacent to the cystic lesion and showed focal chronic pancreatitis and papillary ductal hyperplasia (Fig. 2B). Finally it was confirmed as a cystic dystrophy in heterotopic pancreas of duodenal wall.

**Discussion**

Heterotopic pancreas has been found in 0.5% to 13.7% of autopsies with a male predominance.¹ ⁴ It defined as pancreatic tissue in an extra pancreatic location without

Fig. 2A. Within duodenal wall, a large cyst is present in the submucosa and muscle layers. The cyst wall is denuded of lining epithelium in large part.

Fig. 2B. Pancreatic acinar (left corner) and ductal components are identified in the duodenal wall adjacent to the cyst. Several dilated ducts containing red blood cells are surrounded by chronic inflammatory cells.
continuity the pancreatic itself. Cystic dystrophy in heterotopic pancreas of the duodenal wall is a rare, benign disease affecting young man.\textsuperscript{3-5} It is characterized by the presence of cyst formations surrounded by inflammation and fibrosis in the duodenal wall with or without associated chronic pancreatitis.\textsuperscript{1, 3-5}

The most common symptoms are abdominal pain, emesis, jaundice, nausea, vomiting and weight loss. Pain may be due to inflammation of heterotopic pancreas. Emesis, weight loss and jaundice may be due to duodenal or biliary stenosis.\textsuperscript{3, 6}

The development mechanism of cystic dystrophy of pancreas is poorly understood.\textsuperscript{3} Two distinct types have been reported.\textsuperscript{1, 3} First is cystic dystrophy in heterotopic pancreas without chronic pancreatitis. It is the result of obstructive pancreatitis of heterotopic pancreas by active pancreatic tissue secretion and subsequent cystic enlargement leading compression of pancreatic ducts. Second is cystic dystrophy associated with chronic pancreatitis. It is associated with chronic alcoholism or chronic pancreatic cell insults. Depending on the size of cysts, more frequent cystic and solid types are present. Cystic type cystic dystrophy shows easily recognized cystic lesion (more than 1 cm), located within the thickened duodenal wall. The solid type shows small cysts, (less than 1 cm) within the fibrous thickening of the duodenal wall.

Cystic dystrophy in heterotopic pancreas may be misdiagnosed in clinical practice as a pseudocyst. Several differences exist between cystic dystrophy and psudocyst: The number of cysts is multiple in cystic dystrophy, whereas pseudocyst is solitary. And the cysts of cystic dystrophy are small and round compared with tubular shape of the pseudocyst. The prevalence of calcification in the pancreatic parenchyma is 25\% in cystic dystrophy compared with 100\% of pseudocyst. The other criteria, which favor cystic dystrophy is presence of localized inflammatory changes of the second portion of the duodenal wall and anterior pararenal space. Our case showed a relatively large round to ovoid cyst with localized inflammation.\textsuperscript{1}

Radiologic and endoscopic procedures contribute to diagnosis of cystic dystrophy. Multiple hypo-dense small lesions within a markedly enlarged duodenal wall are seen on CT.\textsuperscript{1, 2} MR imaging shows thickened duodenal area associated with fluid collections and clearly separated from the orthotopic pancreas.\textsuperscript{4} Endoscopic ultrasonography is an useful investigation due to more easily demonstrating the duodenal thickening and tiny intramural cysts less than 1 cm in diameter that are not identified with other imaging modalities.\textsuperscript{4}

Surgical resection is the only means for eradicating cystic dystrophy in heterotopic pancreas. Panreatoduodenectomy is the best treatment for lesion of the duodenum.\textsuperscript{3-7}
요 약

심이지장의 납포성위축증은 특징적으로 이 소성 채장조직에서 발생하는 드문 질환이다. 임상 증상, 이하학적 소견 및 검사실 소견은 특이적이기 전단이 쉽지 않다. 납포성위축증이 발생하는 기전은 아직 명확히 밝혀지지 않았다. 병리학적으로는 심이지장 복에 존재하는 납포성 병변과 이소성 채장조직을 발견할 수 있어 쉽게 진단이 가능하다. 영상학적으로는 두꺼워진 심이지장 복내에 역시 다양한 크기의 납종을 관찰할 수 있다. 근본적인 치료를 위해서는 수술적 절제를 하여야 하며 채심이지장절 제술이 가장 좋은 방법으로 알려져 있다.

References


