Journal of Korean Medical Science
Vol. 9, No. 4, 351-356 AUG. 1994

Duodenal Ectopic Pancreas Complicated by Chronic Pancreatitis and Pseudocyst Formation

— A Case Report —

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Ectopic pancreas is no longer a rare clinical condition, but its unusual clinical manifestations, locations or complications are of clinical interest. We experienced a case(a 48 year-old male patient) of duodenal ectopic pancreas complicated by chronic pancreatitis and pseudocyst formation of which preoperative imaging findings mimicked a large duodenal submucosal tumor with cystic degeneration such as a leiomyosarcoma. Simultaneous chronic pancreatitis was also demonstrated in the isotopic pancreas of the patient postoperatively. Herein we report a rare clinical condition occurring in an ectopic pancreas with a brief review of the literature.

Key Words : *Ectopic pancreas, Chronic pancreatitis, Pseudocyst.*

INTRODUCTION

Ectopic pancreas(heterotopic pancreas, aberrant pancreas, accessory pancreas) is defined as pancreatic tissue found in other than its usual location and having no anatomic or vascular connection with the pancreas itself(Martinez et al., 1958). Ectopic pancreas is no longer a rare clinical condition. The reported surgical incidence of ectopic pancreas is one case in every 500 upper abdominal explorations(Barbosa et al., 1946), and incidence estimates have ranged from 0.55% to 13.7% in various autopsy analyses(Barbosa et al., 1946 ; Feldman and Weinberg, 1952). This condition is more common in men(Barbosa et al., 1946 ; Nakao et al., 1980 ; Armstrong et al., 1981) and the most common sites are the stomach, the duodenum, and the jejunum(Krieg, 1941 ; Barbosa et al., 1946 ; Pearson, 1951 ; Dolan et al., 1974). Most patients with an ectopic pancreas are not symptomatic and thus ectopic pancreas is of no significant clinical importance(Barbosa et al., 1946 ; Dolan et al., 1974 ; Armstrong et al., 1981 ; Lai and Tompkins, 1986). However, ectopic pancreas is subject to various pathological changes occurring in the pancreas itself ; namely, cysts, pancreatitis, hemorrhage, necrosis, and neoplastic change(Barbosa et al., 1946). Although several cases of acute pancreatitis(Qizilbash, 1976 ; Green et al., 1977 ; Fam et al., 1982 ; Benbow, 1988) occurring in heterotopic pancreatic tissue have been reported, we could not find any reports regarding chronic pancreatitis and pseudocyst occurring in an ectopic pancreas.

We report a case of duodenal ectopic pancreas complicated by chronic pancreatitis and pseudocyst formation with a brief review of the literature.

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CASE REPORT

A 48 year-old male patient was admitted to Yongdong Severance Hospital on December 17th, 1992 because of a right upper quadrant(RUQ) abdominal pain of 2-year duration. He had consumed 1 bottle of Soju(a kind of Korean distilled liquor, 45gm of alcohol) and 2 packs of cigarettes almost everyday for 30 years. He had been relatively well until 2 years previously when he had felt RUQ abdominal pain. The pain was felt intermittently, dull in character and aggravated by ingestion of food. Whenever he felt the abdominal pain, he took medicine prescribed by pharmacists to no avail. Eight months prior to admission, he had had an appendectomy at another general hospital under the diagnosis of chronic appendicitis. However, the right upper quadrant abdominal pain persisted despite the appendectomy. One month prior to admission, he had visited a private clinic and had had an abdominal computerized tomography(CT) and an UGI series, the findings of which were suggestive of duodenal mass or pancreatic head mass. He was recommended to visit a general hospital for further evaluation and proper treatment. Past history was not pertinent except for a lens extraction operation in the right eye due to cataract. Family history was unremarkable. He complained of indigestion, but denied any nausea, vomiting, weight loss, steatorrhea or bowel habit change.

On examination, the blood pressure was 130/90 mmHg, the body temperature 36.2℃, the pulse rate 72/min and the respiratoty rate 16/min. He appeared well(body weight 58kg, height 169cm), and the sclerae were not icteric. Examination of the abdomen revealed only mild direct tenderness in the RUQ abdomen. On laboratory examinations, complete blood count, urinalysis, stool examination and blood chemistry revealed no abnormalities. Serum amylase was 86 U/L, and lipase 73 U/L. The tumor markers including CEA, CA19-9 and pancreatic oncofetal antigen were all normal(1.9 ng/ml, 3.8 U/ml, and 4.8 U/ml, respectively).

An abdominal ultrasonography(US) showed a 5.7 X4.2X4 cm sized, central anechoic mass lesion on the right side of the pancreas head(Fig. 1). These findings suggested that the mass was a duodenum-originated large submucosal lesion with central necrosis. An abdominal CT scan showed a 4 cm sized soft tissue mass density with central low density area on the right side of the pancreas head(Fig. 2).

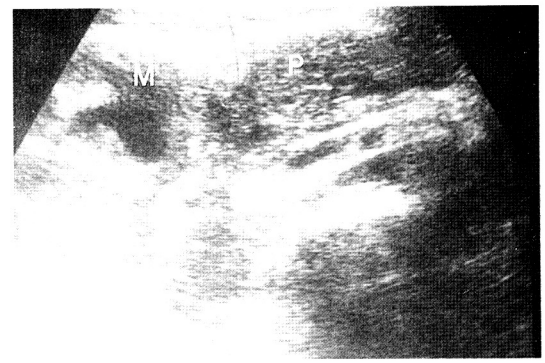

Fig. 1. Abdominal ultrasonography(midline transverse section) shows a 5.7X4. 2X4cm sized, central anechoic mass lesion(indicated with a letter 'M') on the right side of the pancreas head(indicatd with a letter 'P')

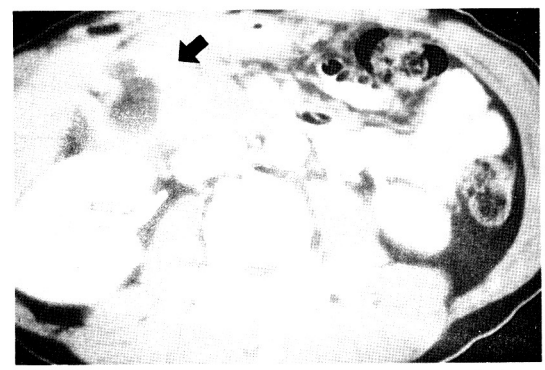

Fig. 2. Abdominal CT scan shows a 4cm sized soft tissue mass density with central low density area on the right side of the pancreas head(arrow). Air density was noted in the right periphery of the mass. Neither the pancreatic duct nor the bile ducts were dilated.

Also air density was noted in the right periphery of the mass. Neither the pancreatic duct nor the bile ducts were dilated. There was no demonstrable lymphadenopathy in the abdomen.

An duodenoscopy revealed a narrowing of the suprapapillary portion of the second duodenum due to a 3-4 cm sized, broad-based submucosal mass lesion(Fig. 3). Neither an opening nor an ulcer was found on the mass. The papilla of Vater was normal. On ERCP examination, the bile duct and the pancreatic duct formed a common channel and showed no intrinsic abnormalities. The common bile duct, however, was indented medially(Fig. 4). The result of an endoscopic biopsy of the duodenal

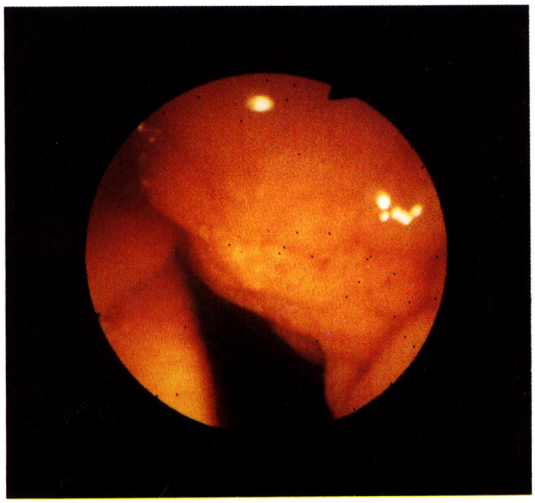

Fig. 3. Duodenoscopic finding shows narrowing of the suprapapillary portion of the second duodenum due to a 3-4cm sized broad based submucosal mass lesion.

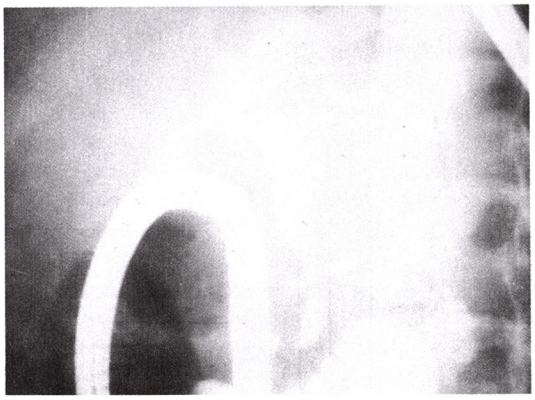

Fig. 4. ERCP shows medial indentation of the distal common bile duct.

mass revealed only chronic non-specific inflammation. Hypotonic duodenography showed a widened duodenal C-loop and a narrowing of the second portion of the duodenum from just distal to the postbulbar portion to just distal to the papilla without mucosal change.

Operative findings showed that a 5X5X6 cm sized mass was palpable at the second portion of the duodenum and densely adhesed to the pancreas head and the distal common bile duct. Pancreaticoduodenectomy(Whipple's procedure) was performed.

Grossly, a yellowish-white nodular fibrotic mass originated from the second portion of the duodenum. When the mass was opened from the duodenal mucosa, a greyish-white fibrotic mass and a cystic structure in the mass were demonstrated(Fig. 5A). On a cut section along the longitudinal axis, the mass was not clearly demarcated from the surrounding structures due to severe fibrosis, but the isotopic pancreas was found on the right side of the mass(Fig. 5B). Microscopically, pancreatic tissues composed of acini, ducts and islets of Langerhans were demonstrated on the submucosal and proper muscle layers of the duodenum(Fig. 6A, 6B). These ectopic pancreatic tissues showed atrophy of the acini and fibrosis and this finding was compatible with chronic pancreatitis.

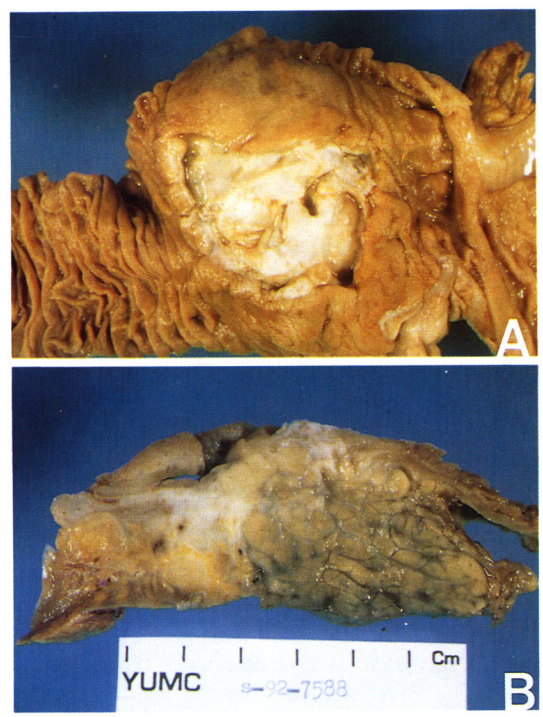

Fig. 5. Gross findings of the resected specimen. A) A yellowish-white nodular fibrotic mass was originated from the second portion of the duodenum. When the mass was opened from the duodenal mucosa, the greyish-white fibrotic mass and the cystic structure in the mass were demonstrated. B) On cut section along the longitudinal axis, the mass was not clearly demarcated from the surrounding structures due to severe fibrosis, but the isotopic pancreas was found on the right-inferior aspect of the mass.

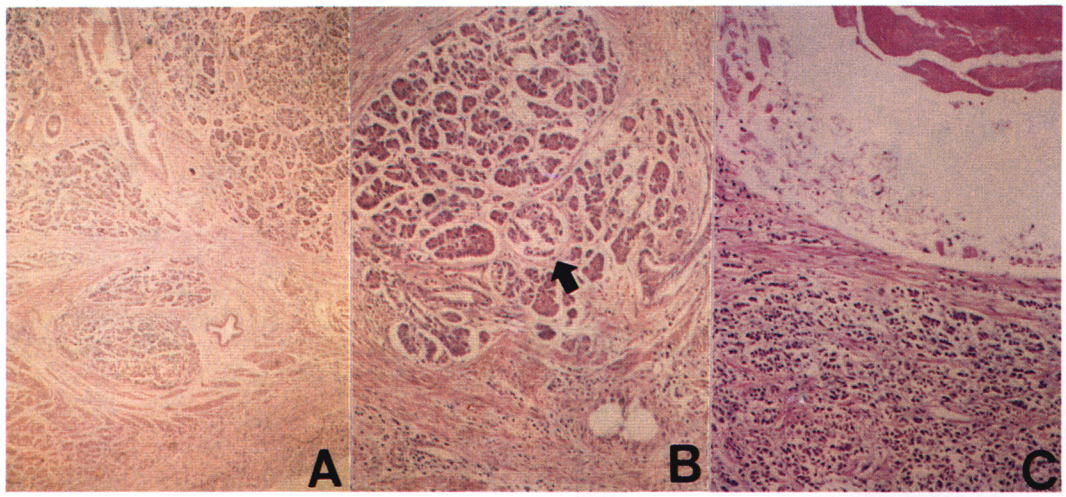

Fig. 6. Microscopic findings of the resected specimen. A) Pancreatic tissues composed of acini and ducts were demonstrated on the submucosal and proper muscle layers of the duodenum. These ectopic pancreatic tissues showed the atrophy of the acini and fibrosis(H & E, ×40). B) Islets of Langerhans(arrow) were also shown(H & E, ×200). C) The cystic portion of the mass revealed no lining epithelium and contained eosinophillic material(H & E, ×200).

The cystic portion of the mass revealed no lining epithelium and contained eosinophillic material(Fig. 6C). Therefore, this cystic structure was considered as a pseudocyst. The isotopic pancreatic tissue also showed atrophy of the acini and fibrosis.

After the operation, the patient complained of postprandial epigastric pain, but gradually improved with conservative management. He was discharged on the 27th hospital day and has been followed up for 5 months till now.

DISCUSSION

Ectopic pancreas is not rarely encountered during upper gastrointestinal tract endoscopy, surgery or autopsy examinations. However, diagnosis of ectopic pancreas is difficult even with the improvement of radiologic, endoscopic, and routine biopsy procedure. Generally, most patients with an ectopic pancreas are symptom-free or show non-specific symptoms such as abdominal pain, epigastric discomfort, nausea and vomiting, bleeding and others-(Kaneda et al., 1989). Specific symptoms of ectopic pancreas can be manifested by either a mass effect or an underlying pathology. The mass effect can cause intussusception of the bowel(Carleton and Ackerbaum, 1976), obstructive jaundice(Daniel et al., 1983 ; Tsunoda et al., 1990 ; Lee et al., 1992), and pyloric obstruction(Krieg, 1941 ; Anseline et al., 1981) depending on the site of an ectopic pancreas. As to the underlying pathology in an ectopic pancreas, almost all of the changes occurring in the pancreas itself may develop in an ectopic pancreas. Pancreatic cancer(Goldfarb and Bennet, 1963), acute pancreatitis (Qizilbash, 1976 ; Green et al., 1977 ; Fam et al., 1982 ; Benbow, 1988) and abscess formation(Kaneda et al., 1989) developed in the exocrine part of ectopic pancreatic tissues have been reported. Also, various islet cell tumors arising from the endocrine part of an ectopic pancreas such as insulinoma(Rose et al., 1980), gastrinoma (Barrocas et al., 1973) and growth hormone-secreting tumor with acromegaly-(Melmed et al., 1985) have been reported. Theoretically, as Busard and Walters(1950) mentioned, chronic pancreatitis might be developed in an ectopic pancreas. We, however, have not been able to find any reports regarding the occurrence of chronic pancreatitis and pseudocyst in an ectopic pancreas.

Our case presented here had complained of right upper abdominal pain for 2 years. The preoperative imaging studies suggested a large duodenal submucosal tumor with central necrosis such as a leiomyosarcoma, so a pancreatico-duodenectomy was performed. Pathologic examination of the re-

sected specimen showed chronic pancreatitis and pseudocyst formation in an ectopic pancreas located in the wall of the second portion of the duodenum with simultaneous chronic pancreatitis in the isotopic pancreas. Although Barbosa et al.(1946) suggested that an ectopic pancreas tissue seemed more subject to malignant change than the pancreas itself for some reasons, ectopic pancreas is believed to be a benign condition, therefore, only local excision, if needed, is sufficient for symptomatic cure. We did not perform a frozen biopsy of the lesion during the operation. If we had diagnosed the ectopic pancreas with a frozen biopsy, we could not have performed a local excision because of severe fibrosis and adhesion. The cause of simultaneous chronic pancreatitis in an ectopic pancreas and the anatomic pancreas of the present case seemed to be heavy alcohol consumption. However, the cause of the pseudocyst formation only in an ectopic pancreas is not clear. We assume that because we were not able to find any draining duct of the ectopic pancreas, either grossly or microscopically, this ectopic tissue might have been more liable to develop pancreatitis. This could be a possible explanation for the patient's localized RUQ abdominal pain, which was relieved by the operation.

Heinrich(1909) classified ectopic pancreas into three histological types. Type I designates a typical pancreatic tissue with acini, ducts and islet cells like a normal pancreas. Type II ectopic pancreatic tissue is composed of a pancreatic tissue with a large number of acini and a few ducts lacking islet cells. Finally, type III designates a pancreatic tissue with a lot of ducts and a few acini without islet cells. It is often difficult to make a distinction between Heinrich's third group and adenomyoma which is defined as a pancreatic tissue consisting of dilated ducts only and unstriped muscle fibers in its stroma. In order to differentiate ectopic pancreas from adenomyoma, Nakao et al.(1980) suggested that histochemical stain for acid phosphatase be used since the ductal epithelium of Brunner's gland showed a strong activity of acid phosphatase whereas that of the pancreas did not. According to the literature review of Nakao et al.(1980) and their own report, Heinrich's second group was the most frequently encountered type and type I was next. Our case fell into the category of Heinrich's first group.

Although several cases of acute pancreatitis occurring in an ectopic pancreas have been reported, we believe that our case is the first clearly demonstrated report on chronic pancreatitis and pseudocyst formation arising from a duodenal ectopic pancreas of which preoperative imaging studies misled the diagnosis to a duodenal sarcoma with cystic degeneration.

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