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Endoscopic Retrograde Pancreatographic Findings of Pancreatic Lipomatosis

Pancreatic lipomatosis is characterized by fatty infiltration or replacement of the pancreas, and has been associated with many conditions. We recently experienced two cases of pancreatic lipomatosis in patients with pancreatic pseudocyst and a case of lipomatosis in diabetes mellitus. In these patients, abrupt obstruction of the main pancreatic duct with smooth tapering is a typical endoscopic retrograde pancreatography (ERP) finding of pancreatic lipomatosis and must be differentiated with pancreatic carcinoma.

Key Words: *Lipomatosis; Pancreatic pseudocyst; Cholangiopancreatography, endoscopic, retrograde*

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INTRODUCTION

Pancreatic lipomatosis, or fatty replacement of the pancreas, is a rare benign disease (1). Although the etiology of this condition is unknown, it has been associated with a variety of diseases, including obesity, diabetes mellitus, chronic pancreatitis, hereditary pancreatitis, and obstruction of the pancreatic duct by a calculus or tumor (1-3). The diagnosis requires demonstration of fatty replacement of the pancreas, a typical pathological feature of the disease under computed tomography (CT) and magnetic resonance imaging (MRI) (2, 3). We report two cases of pancreatic lipomatosis in patients with pancreatic pseudocyst and a case of lipomatosis in diabetes mellitus and pancreas divisum. To our knowledge, no such pseudocyst in a patient with pancreatic lipomatosis has ever been reported in Korea. In addition, we evaluated endoscopic retrograde pancreatographic (ERP) findings in these patients.

CASE REPORTS

Case 1

A 57-year-old Korean woman was referred to Korea University Hospital for evaluation of a pancreatic mass. She complained of generalized abdominal pain and loose stool for four days, and had been diagnosed with acute gastroenteritis and cystic mass of the pancreas at a local clinic. Physical examination was normal. Routine laboratory tests, CEA, and CA 19-9 were unremarkable.

CT scan of the abdomen revealed a thin-walled and non-enhancing cystic mass with curvilinear calcification in the head of the pancreas and thread-like linear density in the pancreatic parenchyme of the body and tail. MRI showed a 3.5×2 cm-sized, lobulated, and clearly marginated mass at the pancreatic neck portion in T1-weighted image and increased signal intensity in T2-weight image. In fat suppression image, body and tail portion of the pancreas showed suppressed signal intensity resembling retroperitoneal fat (Fig. 1). Endoscopic retrograde cholangiopancreatography (ERCP) was performed, and pancreatogram showed abrupt obstruction of the main pancreatic duct at the neck portion without distal duct filling or connection with pseudocyst (Fig. 2A).

An exploratory laparotomy was done and the cystic mass was enucleated. Histopathologic finding of the

Fig. 1. MRI shows pancreatic body and tail (arrow) are suppressed in signal intensity similar to retroperitoneal fat and the pancreatic duct (arrow head) is bright in fat suppression image (case 1).

cyst revealed that the cystic wall possessed a clearly defined wall made of fibrous tissue, but no epithelial lining cells. These findings were consistent with a calcified pseudocyst of the pancreas in a patient with lipomatosis. Follow-up ERCP three months later showed no changes in interval of the main pancreatic duct after surgical resection of the pseudocyst (Fig. 2B), but symptoms were subsided.

Case 2

A 30-year-old man was admitted to the Yong-Dong Severance Hospital due to a sudden onset of vomiting and severe epigastric pain radiating to the back after drinking alcohol the night before. About ten years before admission, the patient had had blunt trauma on the abdomen. In addition, the patient experienced severe epigastric pain again which was relieved in several days without treatment four years ago. On admission, he appeared acutely ill and examination of the abdomen revealed direct tenderness on the epigastrium. Liver function test, amylase, CEA, and CA 19-9 were unremarkable except for a total bilirubin of 1.8 mg/dL.

An abdominal CT scan showed a 3×3 cm sized round cystic lesion in the neck of the pancreas and pancreatic duct dilatation within the replaced fatty tissue. ERCP showed stenosed duct of the pancreatic body, non-visualization of the duct of the pancreatic tail, and visualization of the duct of Santorini (Fig. 3). An exploratory laparotomy was done to evaluate the cyst. A large (7.5×5 cm) cystic lesion was found in the head, neck, and body portion of the pancreas, and histologic examination revealed pancreatic pseudocyst.

Case 3

A 64-year-old woman was admitted to Chungang Gil Hospital for evaluation of a pancreatic head enlargement. The patient was incidentally found with a pancreatic

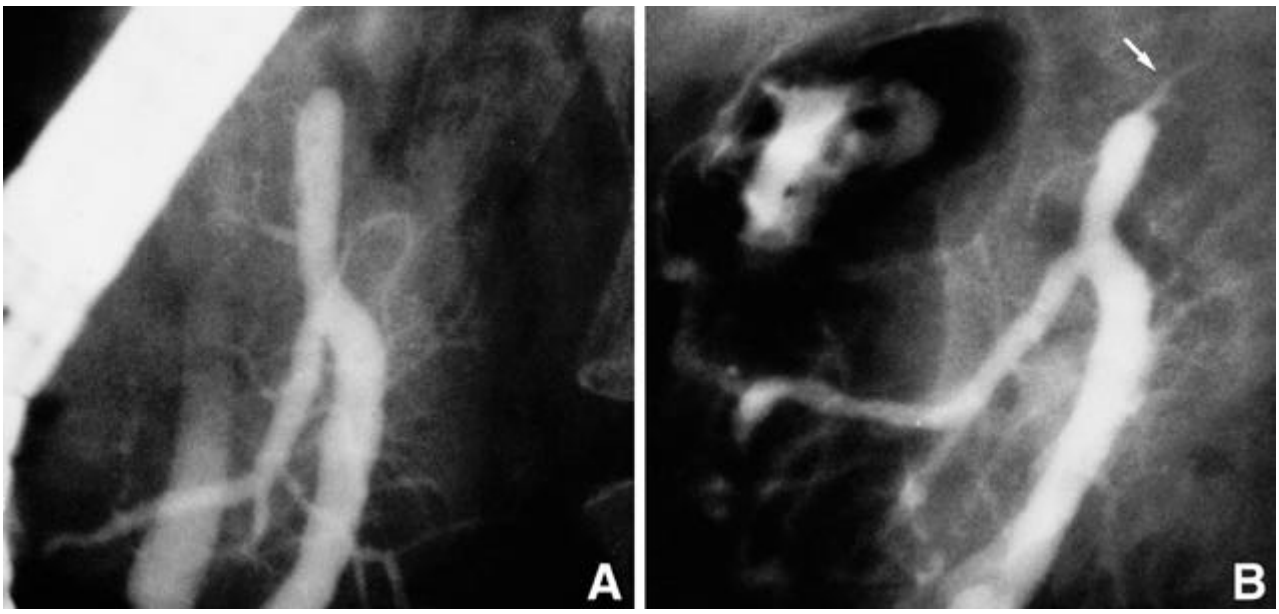

Fig. 2. Initial ERP (case 1) shows abrupt obstruction, which had no connection to the pseudocyst, at the neck portion of the pancreas (A). Follow-up ERP three months later (B), done after cyst resection, shows no remarkable changes in interval of the main pancreatic duct, but with distal minimal stenotic pancreatic duct (arrow).

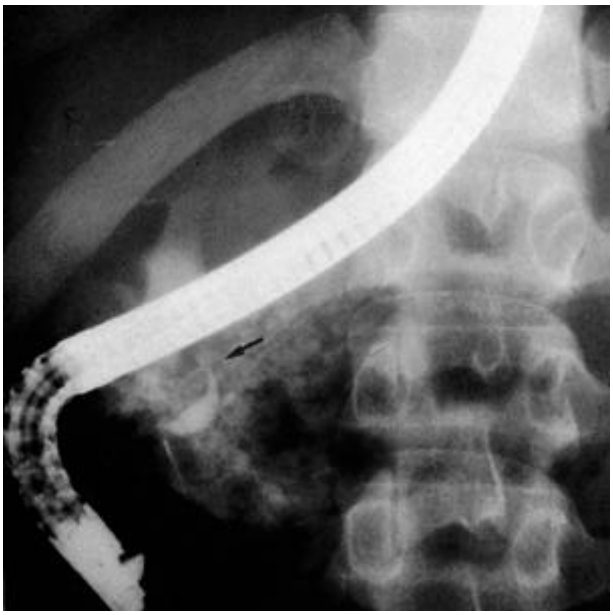

Fig. 3. Findings of the main pancreatic duct on ERP (case 2) show short duct of Wirsung with stenosed and non-visualization of the duct of the pancreatic body and tail (arrow).

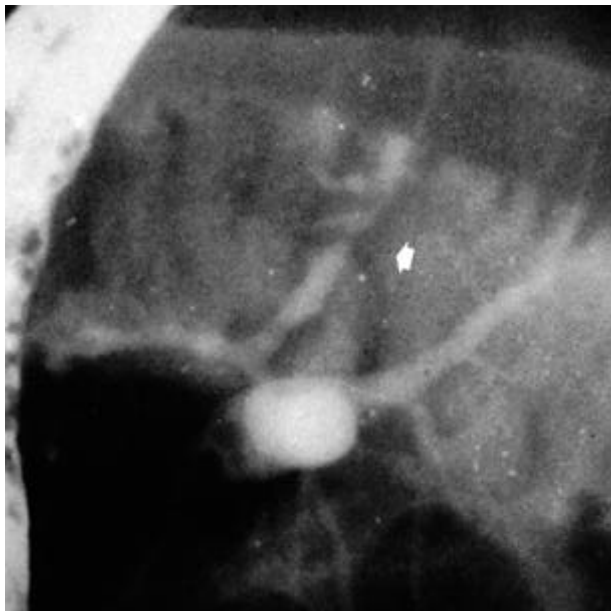

Fig. 4. Pancreatogram (case 3) shows incomplete type of pancreas divisum with abrupt obstruction of the dorsal duct at the neck portion of the pancreas (arrow).

enlargement during general check-up. She had a past history of diabetes mellitus and was treated with oral hypoglycemic agent for ten years. Physical examination and routine laboratory tests were un-remarkable except for a fasting blood glucose of 168 mg/dL.

Abdominal CT scan revealed replaced fatty change at the body and tail portion of the pancreas, and MRI confirmed CT scan finding. ERCP performed through the major papilla, and pancreatogram showed abrupt obstruction of the dorsal duct at the neck portion, shortened ventral duct with small diameter, and communication between the ventral and dorsal duct (Fig. 4). Thus, it was compatible with incomplete type of pancreas divisum and pancreatic lipomatosis.

DISCUSSION

Pancreatic lipomatosis is characterized by fatty infiltration or replacement of the pancreas, but the pathogenesis is not well established (1). No single etiology has been identified for fat replacement of the pancreas, but this condition has been associated with many conditions, including obesity, diabetes mellitus, chronic pancreatitis, hepatic disease, dietary deficiency, viral infection, steroid therapy, obstruction of the pancreatic duct by a calculus, and fibrocystic disease (3-7). According to Olsen (8), age and obesity were significantly correlated to the grade of pancreatic lipomatosis. In our patients, it was difficult to decide whether lipomatosis was caused by ductal obstruction due to a pseudocyst or other factors such as chronic pancreatitis and diabetes mellitus.

Although lesser degrees of fatty replacement are not associated with any clinical symptom, atypical abdominal pain, mass effect of the pancreas, or pancreatic exocrine insufficiency resulting in malabsorption can occur with lipomatosis (1, 2, 5, 9). In these cases, there were no clinical manifestations of the abnormal pancreatic exocrine function, and pancreatic lipomatosis was incidentally detected in radiologic examinations such as in abdominal sonography and CT. Although lipomatosis can be detected by CT and MRI (3, 10-12), MRI is 100% in demonstrating lipomatosis, which gives a very typical signal (3, 12). Abdominal CT shows fat replacement in the pancreatic bed without demonstrable pancreatic parenchyma (14). MRI visualizes normal-sized or enlarged pancreas, a hyper-intense signal on T1- and T2-weighted image, and a null signal on short time inversion recovery-weighted image. As lipomatosis is a rare disease, however, the findings of pancreatogram are not well known. In minimal fat replacement of the pancreas, ERP shows normal appearance of pacreatogram. In severe fat replacement of the pancreas, however, the appearance of pancreatogram is stenosis or abrupt obstruction at the body and tail portions, which is the most common site of ductal changes in pancreatic lipomatosis (13). In this case, only normal ductal finding in pancreatogram was restricted to the pancreatic head without fat replacement. Stenotic main pancreatic duct is typically smooth and has an elongated appearance in pancreatogram of pancreatic

lipomatosis, such as in our cases. In the case of obstruction of the main pancreatic duct, however, it is important to distinguish this obstruction of the main pancreatic duct from obstructions caused by pancreatic dorsal agenesis, chronic pancreatitis, or pancreatic carcinoma. ERP findings of pancreatic carcinoma were stricture, irregular stenosis, or complete obstruction of the main pancreatic duct. In our cases, case 1 and 2 showed obstructive patterns of pancreatogram and thus, differential diagnosis can be made with pancreatic cancer, especially pancreatic head cancer. However, the existence of fat replacement in the pancreas detected by CT and MRI may differentiate between lipomatosis and pancreatic cancer (3). On the other hand, a reliable diagnostic procedure to differentiate pancreatic lipomatosis from dorsal agenesis is selective pancreatic angiography which demonstrates pancreatic blood supply. Angiography showed normal pancreatic blood supply in the case of pancreatic lipomatosis, but not in the dorsal agenesis (14).

In summary, although pancreatic duct obstruction on ERP shows as pancreatic carcinoma, abrupt obstruction of the main pancreatic duct with smooth tapering is a typical ERP finding of pancreatic lipomatosis and must be differentiated with pancreatic carcinoma.

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