

Non-alcoholic Duct-destructive Chronic Pancreatitis: Recognition before Definitive Treatment

Jun Pyo Chung¹, Sang Kyu Na¹, Young Nyun Park², Jeong Sik Yu³, Dong Sup Yoon⁴, Se Joon Lee¹, Si Young Song¹, Kwan Sik Lee¹, Jae Bock Chung¹, Sang In Lee¹, and Jin Kyung Kang¹

Abstract

Non-alcoholic duct-destructive chronic pancreatitis is a new entity that differs morphologically and pathogenetically from alcoholic chronic pancreatitis. Some clinical and imaging features of this entity resemble those of pancreatic cancer, and hence most of the reported cases underwent pancreatic resections including an invasive pancreaticoduodenectomy. Recognition of this new entity before a definitive treatment is therefore important to avoid an unnecessary pancreatic resection. Recently, we experienced a case of non-alcoholic duct-destructive chronic pancreatitis in an 80-year-old man presenting with obstructive jaundice and whose radiologic features were characteristic as originally described. Recognition of this new entity before definitive treatment enabled us to manage this patient optimally. In addition, the relation between non-alcoholic duct-destructive chronic pancreatitis and chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct is discussed.

Key Words: Non-alcoholic duct-destructive chronic pancreatitis, pancreatic cancer, obstructive jaundice, pancreatic resection

INTRODUCTION

Non-alcoholic duct-destructive chronic pancreatitis is a new entity characterized histologically by a conspicuous periductal inflammation that apparently causes duct obstruction and focal duct destruction. Radiologically, it is typified by the absence of parenchymal atrophy and significant ductal dilatation proximal to the site of stenosis, the absence of extrapancreatic spread, and clear demarcation of the lesion.^{1,2}

Some clinical and imaging features of this entity resemble those of pancreatic cancer, and hence most of the reported cases underwent pancreatic resections including an invasive pancreaticoduodenectomy.^{1,2} Recognition of this new entity before any definitive treatment is therefore important to avoid an un-

necessary pancreatic resection.

Recently, we experienced a case of non-alcoholic duct-destructive chronic pancreatitis in an 80-year-old man presenting with obstructive jaundice, and whose radiologic features were characteristic as originally described. Recognition of this new entity before definitive treatment enabled us to manage this patient optimally.

CASE REPORT

An 80-year-old man was admitted to the hospital because of a 20-day history of jaundice. Past medical history revealed that he had suffered from left tuberculous pleurisy during his teens, had had operations for rectal prolapse in 1995, for glaucoma in 1996, and for benign prostatic hyperplasia in 1997. He had smoked for about 50 years, but drunk alcohol minimally.

On admission, he complained of general weakness, easy fatigue, anorexia, nausea, and weight loss (7 kg/2 months), but denied abdominal pain, pruritus, or fever. His vital signs were stable and he appeared chronically ill. The skin and sclerae were icteric and an ill-defined non-tender mass was palpable on the epigastrium.

Received May 20, 1999

Accepted August 6, 1999

Departments of ¹Internal Medicine, ²Pathology, ³Radiology, and ⁴General Surgery, Yonsei University College of Medicine, Seoul, Korea.

Address reprint request to Dr. J. P. Chung, Department of Internal Medicine, Yongdong Severance Hospital, Yonsei University College of Medicine, Yongdong P.O. Box 1217, Seoul 135-270, Korea. Tel: 82-2-3497-3310, Fax: 82-2-3463-3882, E-mail: chungjp@yumc.yonsei.ac.kr

Laboratory findings included a hemoglobin of 10.7 g/dL, leukocyte count of 8,440/mm³, and platelet count of 239,000/mm³. The total bilirubin was 20.5 mg/dL, direct bilirubin 18.1 mg/dL, alkaline phosphatase 1,130 U/L, γ -GT 1,200 U/L, AST 252 IU/L, ALT 342 IU/L, total protein 6.7 g/dL, albumin 3.2 g/dL, total cholesterol 260 mg/dL, amylase 186 U/L, and lipase 953 IU/L. CA19-9 was 197.5 U/ml. ANA (antinuclear antibody), HBsAg, and anti-HBs were negative. A chest x-ray revealed fibronodular densities

on both upper lung fields and left pleural thickening. Direct smears of the sputum were positive for acid-fast bacilli and he was put on anti-tuberculous medication (isonizid, rifampicin, and ethambutol) after his serum total bilirubin decreased.

An abdominal ultrasonography revealed dilated extra- and intrahepatic bile ducts and an ill-defined, 2×2 cm sized mass on the pancreatic head (Fig. 1). An abdominal computed tomography (CT) showed enlarged pancreatic head with a lobulated outer margin (Fig. 2A) and enlarged body and tail of the pancreas with mild dilatation of the main pancreatic duct (Fig. 3A). An endoscopic retrograde cholangiopancreatography (ERCP) taken on the 4th hospital day showed a 2 cm-long irregular stenosis of the main pancreatic duct at the head and neck, as well as minimal dilatation of the upstream main pancreatic duct (Fig. 4). The distal common bile duct was also found to be stenosed. A selective cannulation of the bile duct was tried, but it failed, so a percutaneous transhepatic biliary drainage was carried out immediately. Dynamic magnetic resonance imaging (MRI) was performed on the 6th hospital day to define a mass in the pancreatic head. The pancreatic head was enlarged in T1- and T2-weighted images without a signal difference. After injection of gadolinium, the enlarged pancreatic head was less enhanced than the rest of the pancreas during the arterial and delayed phase while it was not different during the portal phase. Image-guided gun biopsies were performed



Fig. 1. An abdominal ultrasonographic finding on admission. An ill-defined 2.2×2.2 cm sized mass is delineated at the pancreatic head.

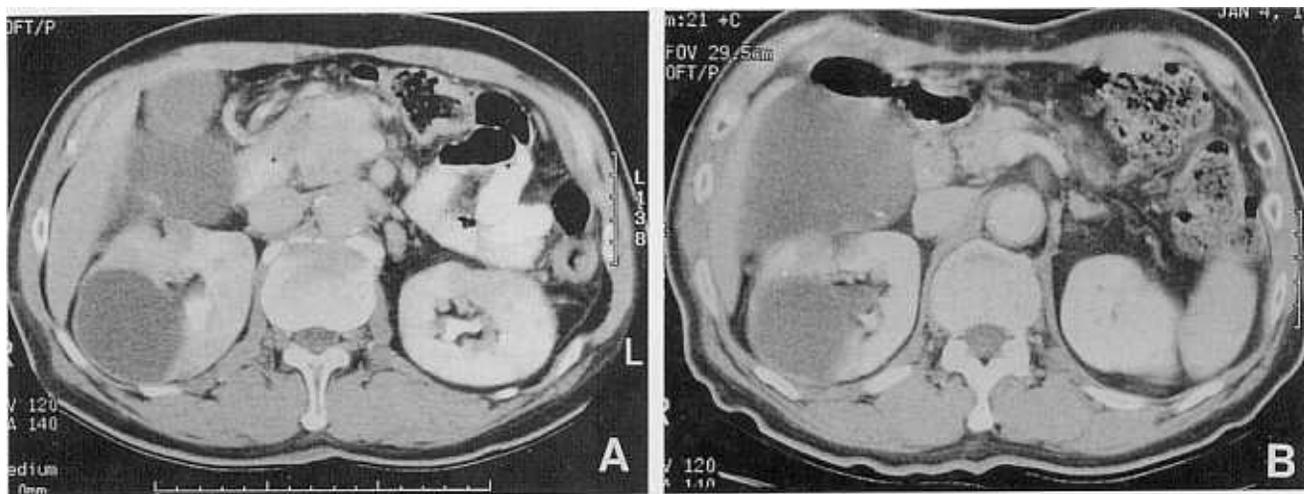


Fig. 2. An abdominal computed tomography at the level of the pancreas head. (A) The pancreatic head is enlarged and shows a lobulated outer margin without a definite mass density. (B) Six months later, the bulging contour of the head was not noted.

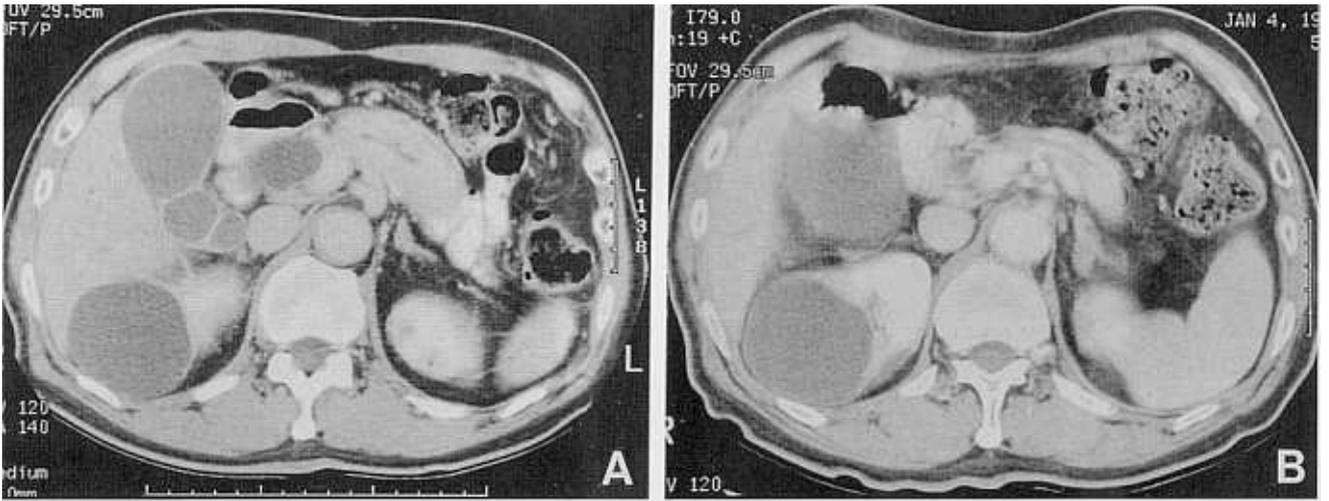


Fig. 3. An abdominal computed tomography at the level of the pancreas body. (A) The extra- and intra-hepatic bile ducts are dilated and the gallbladder is distended. The pancreas appears to be enlarged with mild dilatation of the main pancreatic duct. Incidentally, a huge cyst was found in the right kidney. (B) Six months later, the pancreas decreased in size without interval change of dilatation of the pancreatic duct and the size of the renal cyst.

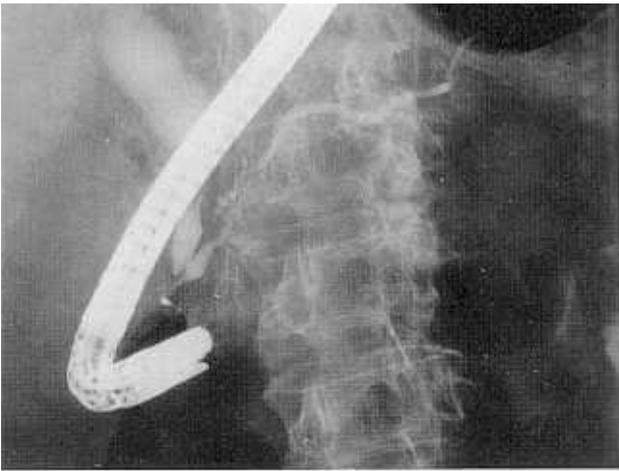


Fig. 4. An endoscopic retrograde cholangiopancreatography. A 2 cm-long irregular stenosis is noted at the pancreatic head and neck area. The upstream of the stenosed pancreatic duct is minimally dilated. The distal common bile duct is also stenosed.

twice, on the 9th and 13th hospital days, and revealed only chronic inflammation and fibrosis of the pancreas. Since non-alcoholic duct-destructive chronic pancreatitis was strongly suspected, an exploratory laparotomy was carried out on the 21st hospital day with the aim of a surgical bypass. On exploration, the pancreas was diffusely enlarged and hard in consistency, but no definite mass was palpated. A frozen



Fig. 5. A microscopic finding of a gun-biopsied specimen. Only glandular atrophy, chronic inflammation and fibrosis are noted without infiltration of malignant cells (H&E stain, ×40).

biopsy of the pancreas head was negative for malignancy. After a wedge biopsy and intraoperative gun biopsy, a cholecystectomy and choledochojejunostomy were carried out. Histological examination of the intraoperatively biopsied specimens also revealed chronic inflammation and fibrosis of the pancreatic parenchyma without infiltration of malignant cells (Fig. 5). Unfortunately, since the pancreatic tissue samples had never included the pancreatic ducts, we were unable to document periductal lymphocytic

infiltration. The patient recovered uneventfully and gained weight after his operation. A follow-up abdominal CT scan done 6 months later showed that the pancreas had decreased in size without evidence of any progression of the malignant process (Fig. 2B and 3B). At present, the patient is in good health (13 months after operation).

DISCUSSION

According to the original description of non-alcoholic duct-destructive chronic pancreatitis by Ectors et al,¹ 11 out of 12 patients underwent pancreatic resections, 9 of whom underwent invasive pancreaticoduodenectomies. Moreover, a total pancreatectomy was reported in a 20-year-old woman.³ The main reason for pancreatic resections has been difficulty in excluding a deadly pancreatic cancer. Some shared features of non-alcoholic duct-destructive chronic pancreatitis and pancreatic cancer are painless jaundice and the presence of a focal or more diffuse mass causing stenosis of the main pancreatic duct or side branches.² Nonetheless, recognition of this special type of chronic pancreatitis is important to avoid an unnecessary pancreatic resection. In this respect, the present case makes a good example. Recognition of this new entity before a definitive treatment led us to manage this patient optimally. Instead of a pancreaticoduodenectomy, a bilioenteric bypass surgery was performed to relieve his obstructive jaundice, which has proved superior to endoscopic stenting in relieving a bile duct stricture due to chronic pancreatitis.⁴ Van Hoe et al. reported that their most recent case was also diagnosed by radiologic findings and CT-guided biopsy, but a detailed description of this patient was unavailable.²

One remarkable feature of non-alcoholic duct-destructive chronic pancreatitis has been reported to be the patients' young age.² Although non-alcoholic duct-destructive chronic pancreatitis may also occur in older patients, pancreatic cancer has a peak incidence in the seventh decade and is only rarely diagnosed in patients younger than 30 years old.² Another point which may support the diagnosis of non-alcoholic duct-destructive chronic pancreatitis was the presence of associated autoimmune or related diseases, which may be associated in up to 30–50% of the cases.² The present subject, however, was neither young nor

associated with autoimmune or related diseases.

Interestingly, the term 'chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct' has been reported and refers to chronic pancreatitis with characteristic features of the main pancreatic duct: marked stenosis and irregularity of the wall without prestenotic dilatation throughout the gland.^{5,6} Histologically, these cases have been reported to show severe fibrosis with prominent lymphocytic infiltration.⁶ This entity was also reported to be associated with autoimmune diseases.⁶ Therefore, we believe that non-alcoholic duct-destructive chronic pancreatitis and chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct are essentially the same entity. We prefer to use the term non-alcoholic duct-destructive chronic pancreatitis because this term encompasses the etiologic and pathologic features of the disease⁷ and the duct narrowing is not always diffuse as seen in our case.

Wakabayashi et al. classified chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct into three groups in view of etiologic factors: (1) complicated with Sjögren's syndrome or primary sclerosing cholangitis, (2) uncomplicated cases with hypergammaglobulinemia and positive ANA, and (3) uncomplicated cases without abnormal immunological findings.⁶ The first and second groups should be autoimmune in origin and responsive to steroid therapy,⁶ but the third group arises from other unknown causes and needs to be determined whether steroid therapy is effective or not. We think this classification is reasonable and, obviously, our present case belongs to the third group.

According to Wakabayashi's collective review,⁶ swelling of the pancreas in patients with chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct could be resolved or atrophied with or without steroid therapy. In the present case, although we are not sure whether it is resolution or atrophy, the pancreas markedly decreased in size without steroid therapy. This finding may indicate that a bile duct stricture that complicates chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct or non-alcoholic duct-destructive pancreatitis may also resolve with time.

In summary, we presented a case of non-alcoholic duct-destructive chronic pancreatitis in an 80-year-old man, in whom the radiologic findings were so characteristic that recognition before definitive treatment

led us to manage the patient optimally. The relation between non-alcoholic duct-destructive chronic pancreatitis and chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct has been discussed here. Obviously, there is a special type of chronic pancreatitis that differs morphologically and pathogenetically from alcoholic chronic pancreatitis and it is easily mistaken for pancreatic cancer. Recognition of this entity before definitive treatment will save unnecessary pancreatic resections.

REFERENCES

1. Ectors N, Maillet B, Aerts R, Geboes K, Donner A, Borc'hard F, et al. Non-alcoholic duct destructive chronic pancreatitis. *Gut* 1997;41:263-8.
2. Van Hoe L, Gryspeerdt S, Ectors N, Van Steenberg W, Aerts R, Baert AL, et al. Nonalcoholic duct-destructive chronic pancreatitis: imaging findings. *AJR* 1998;170:643-7.
3. Klöppel G, Petersen F, Lankisch PG. Chronic pancreatitis imitating pancreatic carcinoma. *Gastrointest Endosc* 1998;48:231.
4. Born P, Rösch T, Bruhl K, Ulm K, Sandschin W, Frimberger E, et al. Long-term results of endoscopic treatment of biliary duct obstruction due to pancreatic disease. *Hepatogastroenterology* 1998;45:833-9.
5. Toki F, Kouzu T, Oi I, Nakasako T, Suzuki M, Hanyu F. An unusual type of chronic pancreatitis showing diffuse irregular narrowing of the entire pancreatic duct on ERCP. A report of four cases. *Endoscopy* 1992;24:640.
6. Wakabayashi T, Motoo Y, Kojima Y, Makino H, Sawabu N. Chronic pancreatitis with diffuse irregular narrowing of the main pancreatic duct. *Dig Dis Sci* 1998;43:2415-25.
7. Bogomoletz W. Duct destructive chronic pancreatitis. A new insight into the pathology of idiopathic non-alcoholic chronic pancreatitis. *Gut* 1997;41:272-3.