Autoimmune pancreatitis mimicking cancer of the head of pancreas : Report of two cases

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Abstract

Autoimmune pancreatitis has been characterised in 1995, but only a few cases have been published since then, most of them from Japan. This report describes the cases of two Belgian male patients who presented with isolated obstructive jaundice. Radiological imaging studies were highly suggestive of carcinoma of the head of pancreas and both patients underwent uneventful cephalic pancreaticoduodenectomy with portal vein resection. Pathological analysis of the removed tissues suggested an autoimmune process in both cases. Both patients had hyper-gammaglobulinemia and antinuclear antibodies, but failed to show evidence of any other autoimmune disease or cause of chronic pancreatitis. In both cases final diagnosis was autoimmune pancreatitis. Preoperative clinical suspicion of this diagnosis is mandatory and may avoid unnecessary surgery in future cases. (Acta gastroenterol. belg., **2001**, 64, **227-230**).

Key words : pancreas, autoimmune disease, autoantibody, surgery, treatment, pathology, case report.

Introduction

It has been reported that functional and morphologic abnormalities of the pancreas may occur in some autoimmune diseases, such as Sjögren's syndrome and systemic lupus erythematosus (SLE) (1,2). An autoimmune mechanism has been proposed to be involved in the aetiology of these pancreatic changes (3). In 1995 Yoshida *et al.* described isolated autoimmune pancreatitis as a new clinical entity, and proposed diagnostic criteria (Table I) (4). Since this report, a few cases have been reported, mostly coming from Japan. We report herein the cases of two caucasian Belgian patients who presented with isolated obstructive jaundice. In both cases the diagnosis of probable pancreatic adenocarcinoma was made and both patients underwent cephalic pancreaticoduodenectomy. The pathological analysis of the removed tissue suggested an autoimmune process. Both cases fulfilled most of the diagnostic criteria proposed by Yoshida *et al.* (4), and definitive diagnosis was autoimmune pancreatitis involving the head of the pancreas. The aim of this report is to make physicians aware of this possible diagnosis in order to avoid unnecessary surgery as corticosteroids may reverse the pancreatic enlargement and the obstructive jaundice.

Case Reports

Case 1

A 52-year-old man was admitted to another hospital because of obstructive jaundice. There was no history of prior illness or weight loss. The patient's average alcohol consumption was 2 glasses of wine per day. Physical examination revealed abdominal meteorism, and epigastric and right upper quadrant abdominal tenderness. The rest of physical examination did not reveal other abnormal physical signs. The chemical data showed evidence of cholestatic and cytolytic liver dysfunction. Abdominal ultrasonography (US) showed a 45 mm mixed hypo- and hyperechogeneous mass within the head of the pancreas, and a dilatation of the extra- and intra-hepatic biliary system and of the caudal pancreatic duct. There was no evidence of cholelithiasis. A computed tomography (CT) scan of the abdomen confirmed the heterogeneous mass of the head of the pancreas consistent with a carcinoma, and peripancreatic lymphadenopathies. Endoscopic retrograde cholangiopancreatography (ERCP) revealed marked narrowing of the lower third of the common bile duct with upstream dilatation. The pancreatic duct was not visualised. An endoprothesis was placed in the common bile duct for decompression. The patient underwent laparotomy that revealed a mass adherent to the portal vein. Peroperative biopsies and a double bypass procedure with cholecystojejunostomy and gastrojejunostomy were performed. Histologic examination of the biopsies of the head of pancreas and peripancreatic lymph nodes showed severe fibrosis and inflammatory infiltration, without any malignancy. However, the diagnosis of untreatable pancreatic carcinoma was confirmed to the patient. The jaundice improved remarkably, and the patient was discharged without further treatment.

Subsequently the patient came to our institution asking for further evaluation. An arterio-CT scan confirmed the mass in the pancreatic head, and the liver appeared

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Table I. — **Comparison of the characteristics of autoimmune pancreatitis adapted from Yashida (4), and chronic pancreatitis**

	Autoimmune Pancreatitis	Chronic Pancreatitis
Increased γ-globulinemia or IgG	yes	no
Antinuclear antibodies	yes	no
Enlargement of the pancreas	yes	yes
ERCP findings	Narrowing of the main pancreatic duct with an irregular wall	Narrowing and dilatation of the main duct and its branches
Previous pancreatic symptoms	no	yes
Cholestasis	yes	yes
Pancreatic calcifications	no	yes
Pancreatic cysts	no	yes
Interstitial fibrosis with acinar atrophy	yes	yes
Preserved Langerhans islets	yes	yes
Portal vein involment	yes	no
Chronic inflammation	yes	yes
Diffuse lymphoplasmacytic infiltration extending to the contiguous soft tissue	yes	no
Occasional association with other autoimmune diseases	yes	no
Effectiveness of steroid therapy		

normal. Considering the young age of the patient and his good general condition, an exploratory laparotomy was decided with the intention to perform a curative cephalic pancreaticoduodenectomy. Intrapancreatic portion of the portal vein was surgically resected and replaced by an internal jugular vein graft. Pathological examination of the pancreas showed a fibrous chronic pancreatitis with lymphoplasmocytic infiltrate consistent with an autoimmune aetiology. The same features were found in the common bile duct and gallbladder. Retrospectively, the patient had hypergammaglobulinemia and antinuclear antibodies (ANA). Salivary glands biopsies performed two weeks later did not show any autoimmune features. The postoperative course was uneventful. The patient is alive and well four years later.

Case 2

A 50-year-old man was admitted to the emergency room at our institution because of jaundice. His past medical history was negative for any significant prior illness. The patient's average alcohol consumption was 2 glasses of wine per day. In two weeks his weight dropped 10 kg. On examination, the patient was jaundiced. There was epigastric tenderness and enlarged cervical, axillary and inguinal lymph nodes were found. Abdominal US and CT studies showed a heterogeneous mass at the head of the pancreas (Fig. 1). A CT scan guided biopsy showed chronic fibrous and inflammatory process. ERCP examination showed narrowing of the lower common bile duct and of the pancreatic duct

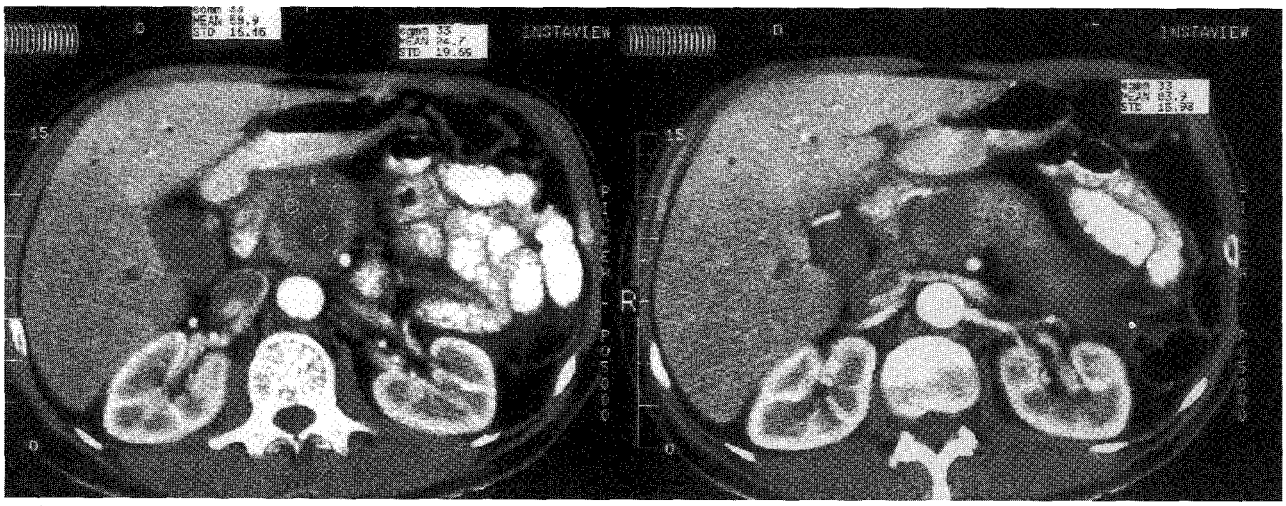

Fig. 1. — Abdominal CT scan of patient 2, showing a heterogeneous mass in the head of the pancreas (left side) and an enlargement of the pancreatic body and tail (right side).

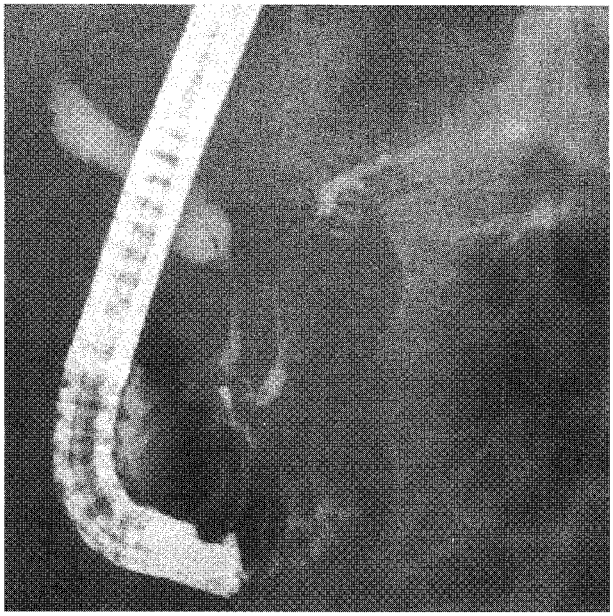

Fig. 2. — ERCP examination of patient 2, showing the narrowing of the lower common bile duct and of the pancreatic duct.

(Fig. 2). Sphincterotomy was performed and an endoprothesis was set in the common bile duct. Malignancy of the pancreatic head was suspected and the patient underwent cephalic pancreaticoduodenectomy. Operative examination showed a mass involving most of the pancreatic head, adherent to the portal vein. The portal vein was resected and replaced by an internal jugular vein graft. Pathological analysis showed a lymphoplasmocytic inflammatory process consistent with autoimmune pancreatitis (Fig. 3). Liver biopsy showed portal hepatitis. Hyper-gammaglobulinemia and ANA were present. Salivary gland biopsy showed signs consistent with, but not specific of Sjögren's syndrome. His postoperative course was uneventful and he was discharged 13 days later. Three years later the patient is alive and well.

Discussion

We report the cases of two Caucasian Belgian patients with a lymphoplasmocytic inflammatory process involving the pancreatic head. Two men in their fifties presented obstructive jaundice and exhibited radiological findings highly suggestive of pancreatic carcinoma. Operative gross appearance of the pancreas showed an enlargement of the pancreatic head compatible with carcinoma. Histologic findings were characterised by diffuse lymphoplasmocytic infiltration with interstitial fibrosis and involvement of the portal vein (Fig. 3). In one of the two patients, similar inflammatory processes involved the bile duct and the gallbladder, comparable with lesions described in primary sclerosing cholangitis. These two cases meet most of the criteria for

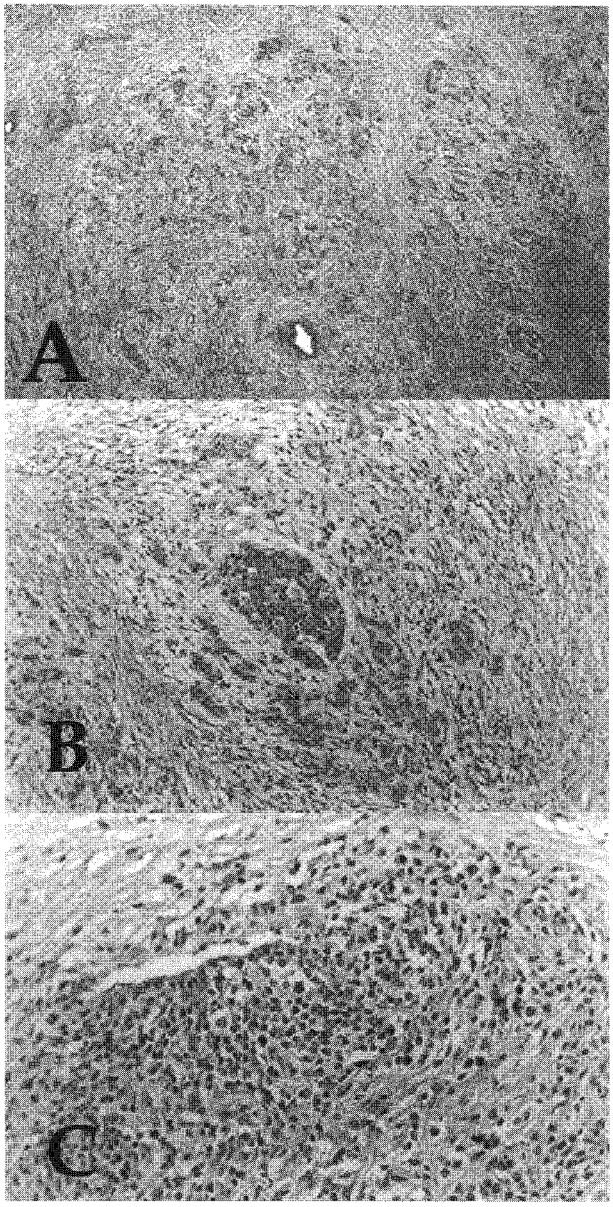

Fig. 3. — Pathology of the resected tissue (hematoxylin-eosin)
A. (× 40). Severe inter- and intra-lobular fibrotic changes, with pseudocirrhotic dissection of the pancreatic parenchyma.
B. (× 100). Conservation of the Langerhans islets.
C. (× 250). Diffuse lymphoplasmocytic infiltrate.

autoimmune pancreatitis proposed by Yoshida *et al.* (Table I), but steroid administration was not initiated since this diagnosis was retrospective and surgical resection had first been performed based on the high suspicion of pancreatic carcinoma.

A particular type of chronic pancreatitis has been reported in association with autoimmune diseases such as Sjögren's syndrome, SLE or primary sclerosing cholangitis (1,2). Pancreatic changes encountered in these diseases have also been shown in patients free of any other autoimmune manifestation (1,3). Yoshida *et al.*

proposed isolated autoimmune pancreatitis as a clinical entity in 1995 (4). Painless jaundice is usually present. In the early stage, serum pancreatic enzymes may increase, but may be depleted in the advanced stage. Radiological findings in autoimmune pancreatitis are not specific : diffusely or locally enlarged pancreas on abdominal US or CT (5), and diffuse or local narrowing and irregularity of the main pancreatic duct on ERCP (1,3). These changes may be related to the inflammatory process and the pancreas may return to normal size with steroid therapy (1,6,7,8). Hyperglobulinemia and ANA are generally present and may lead to the clinical suspicion of the autoimmune process.

In isolated autoimmune pancreatitis macroscopic findings may consist in a diffuse enlargement of the whole pancreas, but localised enlargement of the pancreatic head or tail has also been described (1,6,7,8). This enlargement seems to be secondary to the inflammatory process and may completely reverse with corticosteroid therapy (1,6,7,8). The pancreatic consistency is hard ; its surface is nodular and irregular. Microscopically the pancreatic parenchymal damage consists in a diffuse lymphoplasmocytic infiltrate, as shown in our cases (Fig. 3C). The pancreatic parenchyma is largely dissected by inter- and intra-lobular fibrosis (Fig. 3A). The exocrine tissue is atrophic with a significant decrease in acini number. Langerhans' islets and excretory canals are generally preserved (Fig. 3B). The wall of the main pancreatic duct, the common bile duct and eventually the gallbladder are involved and thickened by the inflammatory process. The soft tissues surrounding the pancreas may also be involved by the inflammatory process that may also infiltrate the wall of the portal vein and induce an obliterating phlebitis (3).

Early diagnosis of autoimmune pancreatitis may avoid unnecessary major surgical procedure and should therefore be proposed in patients with local or diffuse enlargement of the pancreas and with hyperglobulinemia and positive ANA. Chronic pancreatitis due to other known causes should initially be excluded. As opposed to "classical" chronic pancreatitis, alcohol consumption, family history, hypertriglyceridemia and hypercalcemia do not appear to play any role in autoimmune pancreatitis. The effectiveness of steroid therapy has been reported in many cases of autoimmune pancreatitis (1,6,7,8). The steroid therapy may lead to improvement of the jaundice and to the normalisation of the size of the pancreas on CT scan within a few weeks and should be initiated as a diagnostic treatment in case of suspicion of autoimmune pancreatitis.

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