

Case Report

Autoimmune pancreatitis masquerading as pancreatic cancer: case report of a Chinese man

Chu Chu^{1*}, Jianjuan Lou^{2*}, Meiling Bao^{3*}, Shuxian Yang¹, Jianfang Lou¹

Departments of ¹Laboratory Medicine, ²Radiology, ³Pathology, The First Affiliated Hospital of Nanjing Medical University, Nanjing, China. *Co-first authors.

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Abstract: Autoimmune pancreatitis (AIP) is a type of chronic pancreatitis with an autoimmune basis, characterized by infiltrating lymphocytes and plasma cells and fibrosis. Imaging examination revealed pancreatic enlargement and irregular stenosis of the pancreatic duct, and laboratory inspection showed elevated serum IgG4 level. Effectiveness of glucocorticoids (hormone) management is a remarkable feature of this disease. It is reported that both AIP and pancreatic cancer show a marked predilection in older men, and AIP is easily misdiagnosed as pancreatic cancer which leads to unnecessary surgery. Today we present a case of type I AIP in a 64-year-old Chinese old man.

Keywords: Case report, autoimmune pancreatitis, immunoglobulin G4

Introduction

Autoimmune pancreatitis (AIP) is a special type of rare chronic pancreatitis characterized by pancreatic enlargement and irregular stenosis of the pancreatic duct that is related to autoimmune mechanisms. AIP cases have a favorable prognosis with primary steroid treatment; however, due to lack of understanding of AIP in the past, AIP is usually misdiagnosed as pancreatic cancer and may cause unnecessary overtreatment [1].

According to varying pathologic features, autoimmune pancreatitis can be divided into two categories: type I and type II. Type I AIP is usually accompanied by elevated serum IgG4 levels and positive-IgG4 plasma cell infiltration. However, type II shows a prominent idiopathic neutrophil infiltration in the pancreatic duct epithelial tissue and central pancreatitis without elevated serum IgG4, positive autoantibodies, pathologically rare IgG4-positive plasma cell infiltration, and little involvement outside the pancreas, features which are different from the typical Type I AIP, and it is mostly found in Europe and America [2].

Case report

A male, 64 years old, was admitted to the hospital because of “pancreatic head involvement for 1 week” with abdominal pain, and distension, without nausea or vomiting. The signs included scleral icterus and severe mucocutaneous jaundice. In the course of the disease, the patient had a poor appetite and weight loss. In order to further diagnosis and treatment, the patient was admitted for hospitalization with initial diagnosis of “head of the pancreas mass” at that time. Several laboratory examinations were undertaken for further diagnosis and treatment. Results of tumor markers showed: alpha-fetoprotein (AFP): 2.23 ng/mL, carcinoembryonic antigen (CEA): 5.76 ng/mL, carbohydrate antigen 19-9 (CA19-9): 89.83 U/mL and carbohydrate antigen 72-4 (CA72-4): 2.17 U/mL. Other routine detection results were all within normal limits.

CT scanning demonstrated that the pancreatic head was enlarged, showing a bulging contour. After contrast material administration, an area with relatively low density was found in the left portion of the pancreatic head in the arterial phase, measuring 2.8 cm*1.4 cm by enhanced

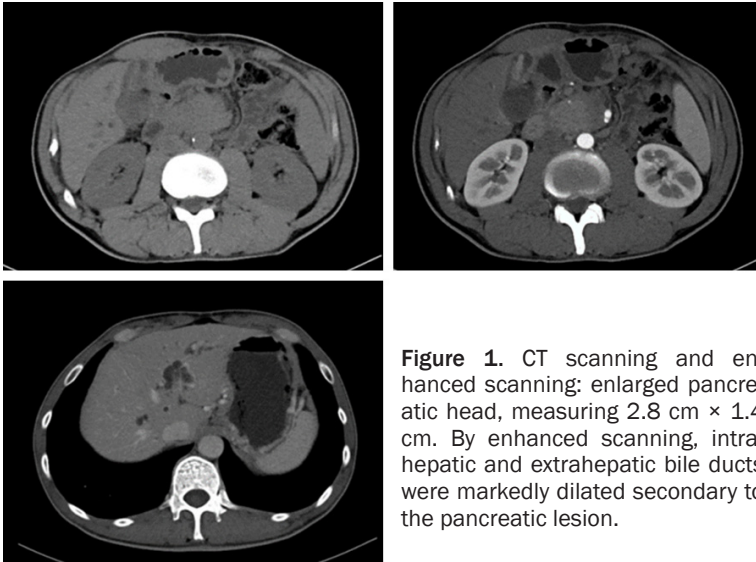


Figure 1. CT scanning and enhanced scanning: enlarged pancreatic head, measuring 2.8 cm × 1.4 cm. By enhanced scanning, intrahepatic and extrahepatic bile ducts were markedly dilated secondary to the pancreatic lesion.

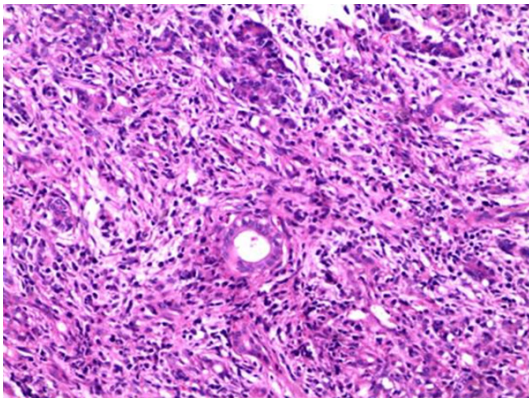


Figure 2. Biopsy of pancreatic tissue (pancreatic head) showing acute and chronic inflammatory cell infiltrate, small abscess formation in a focal area, and hyperplasia of interstitial fibrous tissue (H&E, 20×).

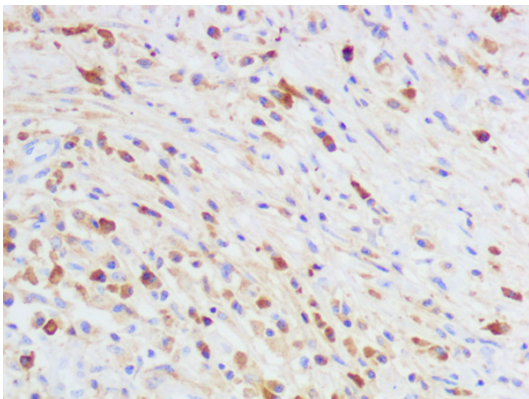


Figure 3. Biopsy of pancreatic tissue (pancreatic head): IgG+ plasma cells/HPF per high power field (Immunohistochemistry, 20×).

scanning. The lesion was ill-defined on the right side, but had a well demarcated right border, abutting the superior mesenteric artery. Intrahepatic and extrahepatic bile ducts were markedly dilated secondary to the pancreatic lesion. The initial imaging diagnosis suggested probable adenocarcinoma arising from the pancreatic head (**Figure 1**).

The patient then underwent a pancreaticoduodenectomy with pylorus preservation and intraoperative section was sent for pathology. Hematoxylin and eosin (H&E) staining of

biopsy of pancreatic tissue (pancreatic head) showed acute and chronic inflammatory cell infiltration, with small abscess formation in a focal area. There were interstitial fibrous tissue hyperplasia, islet cell proliferation, small duct hyperplasia, and focal moderate ductal epithelial atypical hyperplasia. In addition, bile duct margin, duodenal margin, pancreas, and choledochal margin all showed chronic inflammation (**Figure 2**). As well, two and six lymph nodes were involved, in the 8th group and peri-pancreatic, respectively. Immunohistochemistry showed: (pancreatic head) lymphocytes: CD20 (+), CD3 (+), plasma cells: CD38 (+), CD138 (+), Ig κ (+), Ig λ (+), Ki-67 (+). 5 high power fields were counted in the hot spot, showing that the number of IgG4+ plasma cells/HPF was about 18~30 (average: 23) (**Figures 3, 4**). The ratio of IgG4/IgG was about 10%~20% (mean: 15%). Combined with the results of H&E stained biopsy, diagnosis of chronic pancreatitis with focal ductal epithelial moderate dysplasia, interstitial fibrosis, thickened fibrosis of blood vessel wall, with plasma cells and lymphocyte infiltration, and serum IgG4 level was suggested. Finally, IgG4 quantitative assay showed that immunoglobulin G4 level was 5.560 g/L. The patient was eventually diagnosed with autoimmune pancreatitis.

Discussion

AIP is a rare chronic pancreatic disease induced by autoimmune mechanisms, increasingly recognized in recent years. Due to a clinical behav-

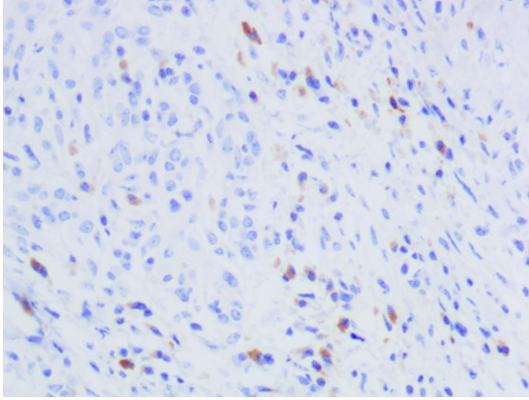


Figure 4. Biopsy of pancreatic tissue (pancreatic head) showing about 18~30 (average: 23) IgG4+ plasma cells/HPF per high power field (Immunohistochemistry, 20×).

ior similar to pancreatic malignancy and painless obstructive jaundice, challenges to the diagnosis and treatment are posed. AIP is pathologically divided into two types: type I and type II reported by Shimosegawa T [3]. Type I, also called IgG4-related AIP, is the most commonly observed AIP in the clinic first described by Sarles [4] with distinctive features including elevated serum IgG4 level and effectiveness of glucocorticoid treatment. Type I has been demonstrated to occur mostly in middle- and elderly-aged men [5]. To date, reports about AIP occurring in the pancreatic head is rare [6]; currently we report Type I AIP in the pancreatic head.

From this case, we can see that the patient had a pancreatic head mass with abdominal pain, distension, severe jaundice, a poor appetite and weight loss which is similar to pancreatic malignancy. After enhanced scanning by CT, because of the limitations and similarities of CT diagnosis (often both have diffuse pancreas enlargement, capsule-like rim and delayed homogenous enhancement on CT imaging) [7] between the two diseases, pancreatic malignancy was initially presumed.

Meanwhile, serum CA19-9 level which has always been considered as an important pancreatic adenocarcinoma marker, also showed a marked increase. Similar reports of a significant increase of serum CA19-9 level above normal range has been reported in many AIP patients [8]. In this case, serum CA19-9 level displayed a remarkable increase, that suggested pancreatic malignancy with consideration of

concurrent symptoms such as jaundice and weight loss.

By pathology, the number of IgG4+ plasma cells/HPF was significantly increased by immunohistochemistry analysis in the pancreatic head, with lymphoplasmacytic infiltration. Based on the histomorphology in accordance with type I AIP, the serum IgG4 was suggested for further exploration. Combined with symptoms, imaging, pathology, and increased serum IgG4 level, the final diagnosis was confirmed. After surgical resection and subsequent steroid treatment, the patient was getting better, which verified a correct diagnosis.

Due to the rarity and low incidence rate of all pancreatic diseases, clinicians easily ignore these findings. Chronic inflammatory diseases are usually related to malignancy and chronic pancreatitis has been reported to be highly associated with the occurrence of pancreatic cancer [9]; thus, it cannot effectively distinguish AIP from pancreatic malignancy.

In many cases, CT examinations may have similar imaging features between AIP and pancreatic cancer such as focal density reduction, envelope-like margins, calcification, pancreatic duct truncation, and pancreatic duct stenosis or expansion. Also, imaging appearances of Type I AIP sometimes are similar to that of pancreatic malignant cystic tumor [10]. For some cases that are difficult to identify, clinicians had best save the patient from an extensive surgical procedure.

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Disclosure of conflict of interest

None.

Address correspondence to: Jianfang Lou, Departments of Laboratory Medicine, The First Affiliated Hospital of Nanjing Medical University, Nanjing, China. E-mail: loujianfang@126.com

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