



CASE REPORT

A variant form of autoimmune pancreatitis successfully treated by steroid therapy, accompanied by von Meyenburg complex

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Abstract

Diagnostic criteria for autoimmune pancreatitis (AIP) have been proposed and used clinically because, despite its unique clinicopathological features, AIP does not have disease-specific serological tests for confirmation. However, diagnosis of a patient with pancreatic lesions mimicking cancer who deviates from these diagnostic criteria is still difficult. We present herein a patient with a variant form of AIP successfully diagnosed by fine-needle biopsy, whose response to steroid therapy was excellent. A 55-year-old Japanese man was admitted to hospital because of jaundice and pancreatic head mass. AIP was considered as one of the differential diagnoses; however, as the patient showed neither pancreatic duct narrowing nor immunological abnormalities, he did not meet the Japanese diagnostic criteria for AIP. Histopathology of the pancreatic mass demonstrated abundant infiltration by lymphocytes and interstitial fibrosis, which suggested AIP. Immunoreaction to IgG4, which is supposed to be specific to AIP, was not observed; however, response to subsequent prednisolone therapy was good, with dramatic pancreatic head mass regression. Aside from the pancreatic head mass, diffusely spreading small lesions were observed throughout the liver. The likelihood of a potential

association with extrapancreatic lesions of AIP was considered and led us to carry out a liver biopsy, which revealed biliary hamartoma, also called von Meyenburg complex (VMC). As IgG4-positive plasma cell infiltration was not demonstrated in the hamartomatous regions, the hepatic condition was thought to have occurred incidentally; however, to the best of our knowledge, this is the first report in which the association between AIP and VMC was investigated and discussed.

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Key words: Autoimmune pancreatitis; Diagnostic criteria; IgG4; Biliary hamartoma; von Meyenburg complex

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INTRODUCTION

Clinically unique chronic pancreatitis, in which autoimmune mechanisms are supposed to be involved, has been focused upon and investigated intensively over the last decade^[1-3], and is now recognized as a clinical entity worldwide, autoimmune pancreatitis (AIP)^[4-6]. Since disease-specific serological tests for determining AIP remain unavailable, diagnostic criteria for AIP have been proposed by incorporating characteristic clinical aspects of the disease^[7-11]. Japanese criteria were established first in the world in 2002 by the Japan Pancreas Society (JPS)^[7]; however, with the accumulation of clinical investigations of AIP, the JPS criteria were modified in 2006 by the Research Committee of Intractable Pancreatic Diseases (RCIPD)^[9], because the concept of AIP has changed gradually with the detection of additional new clinicopathological aspects,

such as a high concentration of serum IgG4^[12], a variety of extrapancreatic involvement relevant to IgG4-positive lymphoplasmacytic infiltration^[13-15], and morphologically atypical localized AIP^[16-19]. On revision, these new clinical findings were included in the criteria, enabling more cases to be diagnosed. However, because Japanese criteria mandate first identification of characteristic imaging, namely, narrowing of the main pancreatic duct demonstrated by endoscopic retrograde cholangiopancreatography (ERCP) and enlargement of the pancreas, in order to exclude absolutely pancreatic or biliary cancer rather than picking-up suspicious AIP patients^[9], some AIP patients remain undiagnosed as a result of being unmatched with the Japanese criteria for AIP. Yet, because AIP has a favorable prognosis if steroid therapy is administered appropriately^[20], we should also pay attention to atypical forms of AIP before a patient undergoes an operative procedure.

We describe herein a case of the variant form of AIP diagnosed by fine-needle aspiration biopsy, and the subsequent successful response to steroid therapy, whose primary presentation was obstructive jaundice caused by the pancreatic head mass alone, without other specific features outlined in the Japanese diagnostic criteria for AIP. In addition, the patient was found to concomitantly have biliary hamartoma (von Meyenburg complex; VMC), the initial clinical appearance of which first led us to consider them as possible extrapancreatic lesions of AIP.

CASE REPORT

A 55-year-old Japanese man was referred and admitted to our hospital for the evaluation of jaundice and a pancreatic head mass on 2 May 2007. One week before admission, the patient had visited another hospital because of body weight loss and a 1-wk history of epigastralgia, and was found to have obstructive jaundice caused by pancreatic head swelling, upon abdominal ultrasonography (US). Upon admission, he was 170 cm tall and weighed 57.0 kg, blood pressure was 112/62 mmHg, and temperature was 37.1°C. There was no history of drug or alcohol abuse.

Physical examination revealed no abnormal findings except for jaundice of the conjunctiva and skin. Laboratory findings on admission are shown in Table 1. Abdominal US showed obstructive jaundice caused by a hypo-echoic pancreatic head mass that measured 43 mm in diameter, which compressed the lower portion of the bile tract, and resulted in intra- and extra-hepatic dilatation of the bile duct. In addition, many irregular hyperechoic spots and comet-like tails were found to spread diffusely in the bilateral liver parenchyma (Figure 1). Upon dynamic contrast-enhanced computed tomography (CT), the pancreatic head tumor was slightly less enhanced than the other portion of the pancreas, together with irregular dilatation of the main pancreatic duct distal to the tumor, which suggested pancreas cancer. However, the distal area of the pancreas was not atrophied (Figure 2A and B).

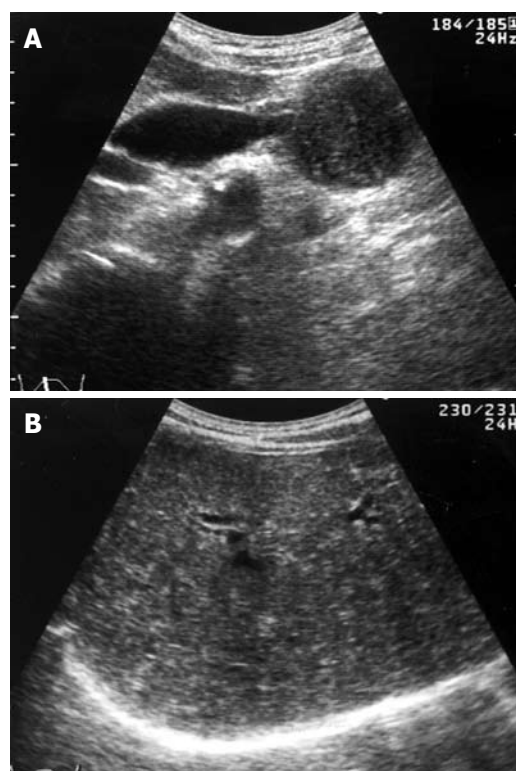


Figure 1 Abdominal US. A: Pancreatic head mass with hypo-echogenicity, 43 mm in diameter, which compressed the lower portion of the bile duct, and resulted in upstream bile tract dilatation; B: Many tiny, diffuse hyperechoic patches or comet-like tails that suggested the presence of certain intrahepatic abnormalities, but the reason could not be elucidated.

Magnetic resonance cholangiopancreatography (MRCP) performed subsequently demonstrated intensive stenosis of the pancreatic head portion and peripheral dilatation of the common bile duct, as well as the skipped stenotic lesions of the main pancreatic duct (Figure 3). Simultaneously, on MRCP imaging, numerous hyperintense round nodules of small diameter, presumably correlating with the US imaging of the liver, were found to be diffusely scattered throughout the liver (Figure 3). We could not perform ERCP because of the lack of an expert ERCP endoscopist in the hospital. Upon MRCP, skipped stenotic lesions of the main pancreatic duct, which was suggestive of AIP was observed; however, irregular narrowing of the pancreatic duct seen in typical AIP was not observed. The levels of tumor markers, carbohydrate antigen (CA 19-9, DUPAN-2) and carcinoembryonic antigen (CEA), were normal. Immunologically, antinuclear antibody was absent and the serum level of total IgG as well as IgG4 was within the normal range (Table 1). As we could not obtain diagnostic confirmation from these sequential atypical clinical features of AIP or pancreatic cancer, we performed US-guided percutaneous aspiration biopsy using a 21G fine needle that targeted the pancreatic head mass on 15 May 2007. Histopathological examination demonstrated marked infiltration of pancreas parenchyma by mononuclear cells and fibrosis adjacent to the lesion, which suggested a diagnosis of AIP (Figure 4A). There were no malignant cells indicating

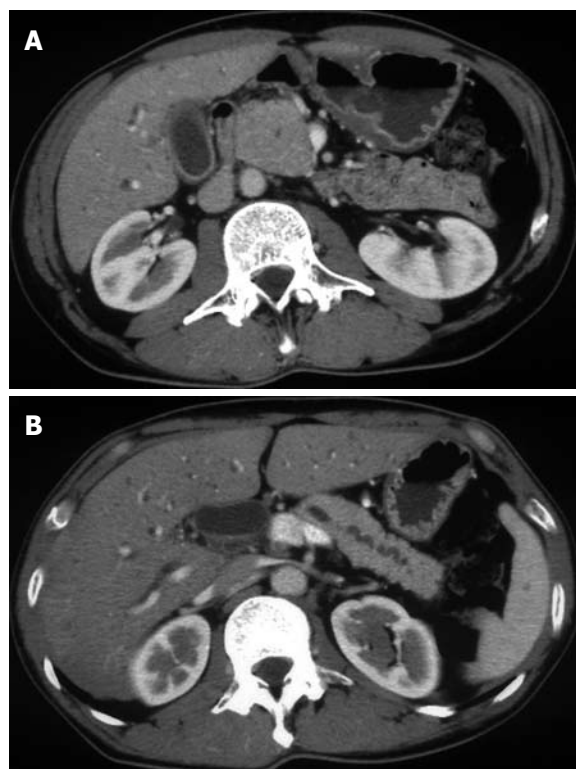


Figure 2 Abdominal enhanced CT imaging with contrast medium in May 2007. A: Swelling of the pancreatic head like a solid tumor was shown. B: The distal portion of the main pancreatic duct was found to be irregularly dilated; however, the pancreatic body and tail were not atrophied as seen in pancreatic cancer.

pancreatic cancer. These infiltrated cells were revealed to be mainly T lymphocytes by subsequent immunohistochemistry (Figure 4B), which was more compatible with AIP. However, plasma cell infiltration was scarce and immunoreaction to anti-IgG4 antibodies was not seen (Figure 4C).

The patient did not fulfill the JPS diagnostic criteria for AIP. However, as the presence of pancreatic cancer cells was histopathologically ruled out and the likelihood of spontaneous regression of obstructive jaundice seemed to be poor, we started oral prednisolone (PSL) therapy on 18 May 2007. Once PSL at an initial daily dose of 30 mg was introduced, prompt regression of the pancreatic head mass and improvement of bile and pancreatic duct dilatation was achieved (Figure 5A and B).

Apart from the pancreatic head mass, the presence of intrahepatic anomalies was suspected from the US and MRCP imaging. As AIP is known to show frequently a variety of extrapancreatic involvements, we carried out liver biopsy in order to elucidate the identity of the diffusely spreading aberrant echoes on US and small nodules seen on MRI, in addition to investigating the possible association with AIP. Histopathological examination of the hepatic parenchyma revealed aggregation of irregular-shaped cystic dilated bile ducts embedded in fibrous stroma, with minimal inflammatory reaction (Figure 6), and biliary hamartoma, also called VMC. Additionally, we performed immunohistochemistry using anti-IgG4 antibody, which resulted in negative interactions in those areas.

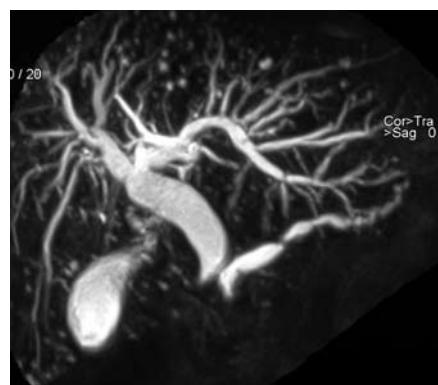


Figure 3 MRCP showed proximal stenosis of the bile duct as well as the main pancreatic duct; however, narrowing of the pancreatic duct, as seen in AIP, was not demonstrated at all. A simultaneous finding was that there were many round high-intensity nodules scattered throughout the liver.

Table 1 Laboratory data

	Normal value	May 2, 2007	July 14, 2007
WBC (White blood cell)	4000-9000/ μ L	10210	15350
RBC (Red blood cell)	410-530 $\times 10^4$ / μ L	504	471
Hb (Hemoglobin)	14-18 g/dL	15.5	15.0
Plt (Platelet)	12-36 $\times 10^4$ / μ L	33.9	23.4
TP (Total protein)	6.5-8.0 g/dL	6.8	6.9
Alb (Albumin)	3.9-4.9 g/dL	3.5	4.3
IgG (Immunoglobulin G)	870-1700 mg/dL	919	843
IgG4 (Immunoglobulin G4)	4.8-105 mg/dL	20.3	-
AST (Aspartate aminotransferase)	10-33 IU/L	49	19
ALT (Alanine aminotransferase)	4-30 IU/L	57	28
LDH (Lactate dehydrogenase)	100-230 IU/L	146	132
ALP (Alkaline phosphatase)	167-345 IU/L	992	265
γ -GTP (γ -glutamyl transpeptidase)	10-75 IU/L	140	51
TB (Total bilirubin)	0.2-1.2 mg/dL	4.6	0.7
DB (Direct bilirubin)	0-0.4 mg/dL	2.6	0.1
Amylase	30-120 IU/L	105	36
BUN (Blood urea nitrogen)	8-20 mg/dL	13	13
Cr (Creatinine)	0.6-1.1 mg/dL	0.6	0.7
CRP (C-reactive protein)	0-0.4 mg/dL	0.0	0.0

All laboratory abnormalities seen upon admission were normalized by 14 July 2007 (Table 1), and the patient remains in a good condition without recurrence at the present time, under maintenance therapy with 5 mg/d PSL.

DISCUSSION

This case report presents notable clinical information for dealing with AIP patients. First, the existence of a variant form of AIP not meeting the diagnostic criteria was verified by fine-needle aspiration biopsy, together with a subsequent dramatic good response to steroid therapy. Second, concomitant development of AIP and VMC was investigated and discussed for the first time, even though this condition might occur incidentally.

Sets of diagnostic criteria for AIP have been proposed by integrating several known unique morphological, immunological and histopathological characteristics of AIP. Currently, four major sets of diagnostic criteria for AIP have emerged from Japan^[9], Italy^[8], Korea^[10] and the

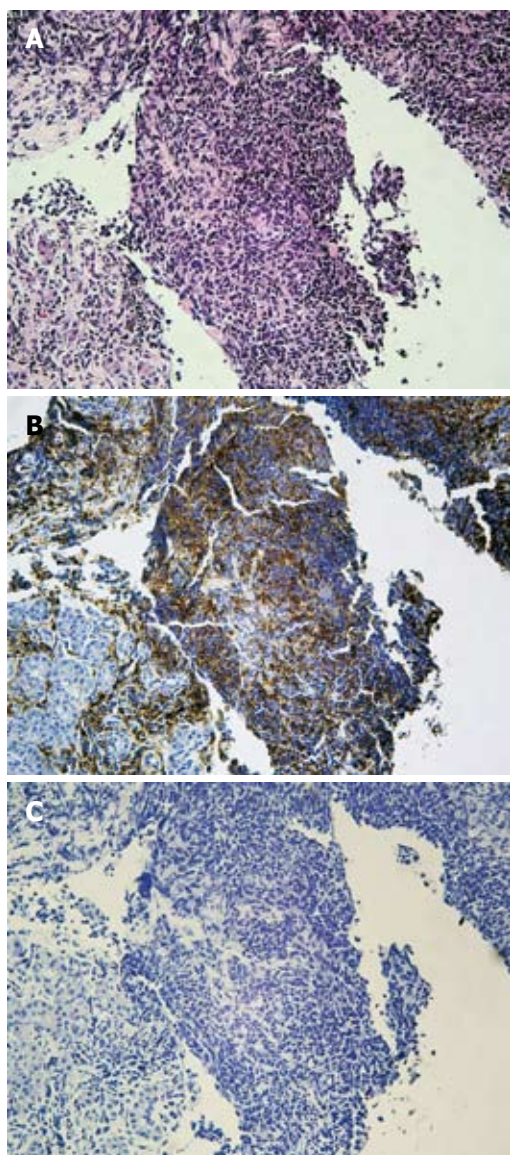


Figure 4 Histopathology of a specimen taken from the pancreatic head mass showed no malignant cells that indicated cancer. A: Marked infiltration of pancreatic parenchyma by lymphocytic mononuclear cells, as well as interstitial fibrosis suggested a diagnosis of AIP (HE, $\times 200$). B: These infiltrated cells were revealed to be mainly T lymphocytes (T-cell staining, $\times 200$). However, the histopathology was not diagnostic without so-called LPSP with scarce plasma cell infiltration. C: Immunohistochemistry using antibody against IgG4 resulted in a negative study (IgG4 staining, $\times 200$).

United States^[11], and have been used clinically in each setting. However, there are some differences between each criterion for AIP that make it difficult to compare data in studies from different centers. Recently in 2008, Asian diagnostic criteria for AIP have been proposed by RCIPD and the Korean Society of Pancreatobiliary Diseases to reach an international consensus, thus enabling more cases to be accurately diagnosed and restraining the routine use of steroids for the diagnosis of AIP^[21]. In the JPS criteria established in 2002^[7], on the basis of the minimum consensus features of AIP required to avoid misdiagnosing malignancy, but not to pick-up suspicious cases of AIP, the demonstration of diffuse swelling of the pancreas and diffuse and irregular narrowing of the main pancreatic duct by

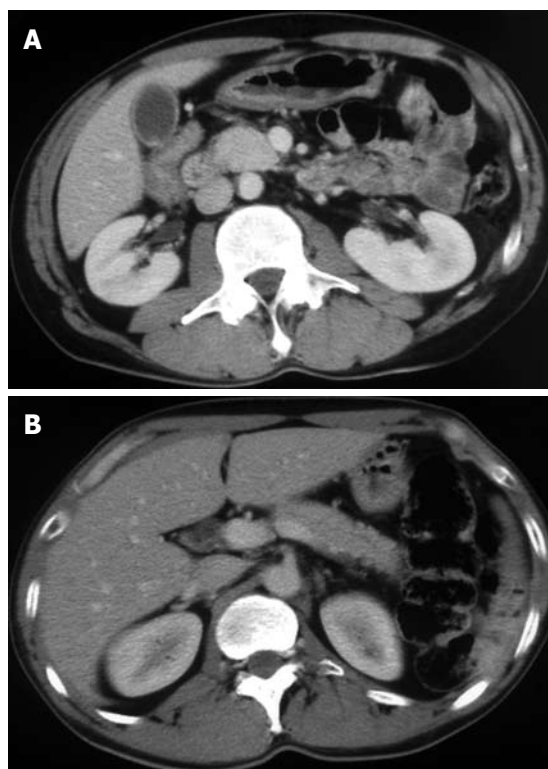


Figure 5 Abdominal enhanced CT scan with contrast medium in July 2007. As compared with CT imaging taken in May 2007, swelling of the pancreatic head had significantly regressed (A). Irregular dilatation of the main pancreatic duct had improved with the whole body bulk being reduced (B).

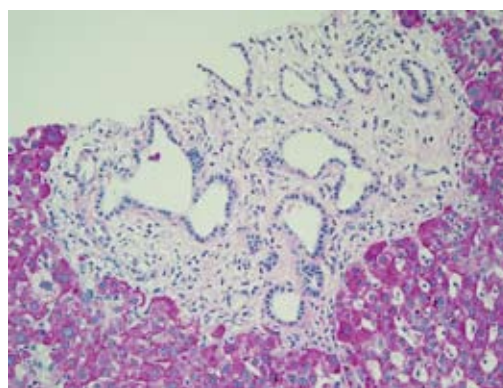


Figure 6 Histopathology of specimens taken from the hepatic parenchyma (PAS, $\times 200$). Distribution of many irregular, angulated duct structures was observed. Ductal epithelium was comprised of monolayered columnar epithelium, morphologically identical to biliary epithelium, surrounded by fibrous connective tissues with minimal inflammatory change. The sequential histopathological features were considered to be bile duct hamartoma (VMC). Subsequent immunohistochemistry of the region by anti-IgG4 antibody was negative.

ERCP is stressed and is required first of all. On revision of the JPS criteria in 2006^[9], more cases of AIP have been correctly diagnosed. However, in the present case, since pancreatic swelling was located in only the head portion, which resembled a solid tumor, and there were neither pancreatic duct narrowing nor immunological abnormalities including IgG4, AIP could not be definitely diagnosed even with application of the revised Japanese criteria.

There are an increasing number of reports dealing

with atypical cases of AIP with focal swelling mimicking pancreatic cancer, as well as cases that begin with focal mass and progress to diffuse swelling, which results in a typical AIP appearance^[16-19]. However, the most important issue when dealing with localized mass-forming AIP is that we should differentiate it absolutely from pancreatic or biliary cancer, in order to prevent too ready use of steroids, and leaving cancer untreated under a mistaken diagnosis. With this in mind, we performed US-guided percutaneous fine-needle aspiration biopsy, which targeted the pancreatic head mass and showed massive inflammatory cell infiltration, as well as interstitial fibrosis, and denied the presence of malignant cells. Subsequent steroid therapy resulted in an excellent clinical course of the patient. As AIP is known to show favorable prognosis, as in the present case, when steroid therapy is adequately administered^[20], we should also detect suspicious variant forms of AIP before a patient undergoes invasive surgery.

It is debatable whether the diagnosis of AIP can be made from pancreatic core biopsy specimens, because they often do not show the complete spectrum of diagnostic change of lymphoplasmacytic sclerosing pancreatitis (LPSP) seen in typical AIP by routine histology alone^[22]. In addition, certain types of pancreatic cancer exhibit a reaction that resembles LPSP^[21]. However, we think that the purpose of fine-needle aspiration biopsy is to rule out the presence of malignant cells, and to provide a basis for the introduction of steroid therapy, rather than to establish a diagnosis of AIP, when we encounter a patient with a pancreatic mass, in whom differential diagnosis is difficult. Therefore, when a patient presents with uncertain clinical features of a pancreatic tumor, the presence of AIP and pancreatic fine-needle aspiration biopsy should be considered as one of the diagnostic modalities.

As awareness of AIP increases and more cases are reported and clinically investigated, high serum IgG4 concentration has been recognized as a notable characteristic and a marker of AIP disease activity^[12]. Moreover, as the presence of a variety of extrapancreatic lesions related to IgG4-positive plasma cell infiltration, such as sclerosing cholangitis^[23], sialadenitis^[24] and retroperitoneal fibrosis^[25], becomes evident, AIP has been proposed as one of the clinical aspects of generalized autoimmune IgG4-related sclerosing disease^[13-15]. Therefore, we should consider the presence of extrapancreatic lesions of AIP when encountering unexplained manifestations outwith the pancreas. In the present case, the initial features of diffusely scattered intrahepatic lesions demonstrated on US and MRCP first led us to consider them to be possible extrapancreatic lesions of AIP. However, subsequent liver biopsy showed the presence of a unique group of cystic and dilated bile ducts surrounded by a dense fibrocollagenous stroma, which was revealed to be biliary hamartoma. Biliary hamartomas, also referred to as VMC, are benign liver malformations that histopathologically include a cystic dilated bile duct, typically 0.1-0.3 cm in diameter,

embedded in abundant fibrous stroma^[26]. As a result of its asymptomatic nature, this relatively rare entity is usually detected incidentally in 5.6% of reported autopsies^[27]. In view of the results of histopathological investigation of the liver, including negative IgG4 staining, it seems unlikely that a common pathogenesis underlies AIP and VMC. However, as far as we know, this is the first presentation dealing with both AIP and VMC.

In conclusion, we reported herein a variant form of AIP accompanied by VMC, which did not fulfill the Japanese diagnostic criteria. When we encounter a patient with atypical presentation of a pancreatic disease and find it difficult to make a diagnosis, despite applying diagnostic criteria, fine-needle aspiration biopsy of the pancreatic lesion should be considered as a diagnostic modality.

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