Abstract:
A 64-year-old man with a history of diabetes and gallstones was admitted to our institution with suspected pancreatic malignancy. Computed tomography (CT) revealed multiple pancreatic cysts and massive ascites, and endoscopic ultrasonography (EUS) revealed a 28×27-mm hypoechoic mass in the pancreatic head. An EUS-guided fine-needle aspiration biopsy was performed, and there were no malignant findings. Based on the test results and imaging findings, type 1 autoimmune pancreatitis was suspected. The patient was administered 30 mg of prednisolone daily. After 11 days, CT revealed that the pancreatic cysts and ascites had reduced in size.

Key words: autoimmune pancreatitis, pancreatic cysts, ascites, steroids

Introduction
Autoimmune pancreatitis (AIP) is a type of chronic pancreatitis, recognized as an IgG4-related pancreatic disease. Patients with AIP present with symptoms of pancreatic and biliary duct cancer, such as jaundice and abdominal pain (1). Therefore, an accurate diagnosis requires differentiation between pancreatic cancer and biliary duct cancer.

The present patient had massive ascites and a large cyst measuring 73 mm in diameter. Because of the atypical imaging findings of AIP, peritoneal dissemination of pancreatic cancer was initially suspected. Few cases of AIP with massive ascites and cysts have been reported. We herein report a very rare case of AIP with multiple pancreatic cysts and massive ascites that was successfully managed with steroid treatment alone.

Case Report
A 64-year-old man with a history of diabetes was previously admitted to a different institution with suspected pancreatic malignancy. He presented to the referring hospital with a loss of 10 kg in body weight over 12 months and abdominal distension. The patient had no history of acute pancreatitis or allergic diseases. He had drunk approximately 20 g of alcohol per day for 40 years. Computed tomography (CT) revealed a large amount of ascites, multiple pancreatic cysts up to 73 mm in size, splenic vein occlusion, and a soft-tissue shadow around the common hepatic artery and splenic artery (Fig. 1). A cytological examination of the ascites fluid revealed no malignant cells, and the amylase level was as high as 775 IU/L. Although there were no malignant findings in the ascites fluid, the possibility of a malignant tumor, such as pancreatic cancer, could not be ruled out.
Figure 1. Computed tomography (CT) performed at the previous medical institution. CT showed multiple cysts (red arrows) in the pancreas (maximum diameter, 73 mm) and ascites (yellow arrows). Obstruction of the splenic veins was also observed.

Figure 2. Computed tomography (CT) examination at our hospital. CT showed less ascites than the previous examination; however, multiple pancreatic cysts persisted (red arrows).

Therefore, the patient was referred to our hospital for a further investigation.

The patient’s vital signs were normal. A physical examination revealed no tenderness or spontaneous pain in the abdomen. Laboratory tests showed increased amylase (259 U/L) and lipase (256 U/L) levels, despite the absence of other tumor markers, such as carcinoembryonic antigen (CEA) and carbohydrate antigen 19-9 (CA19-9). Enhanced CT at the time of admission showed that the ascites had ameliorated, but the pancreatic cysts remained (Fig. 2). Diffusion-weighted magnetic resonance imaging (MRI) revealed a hyperintense pancreatic mass-like lesion in the pancreatic head, and magnetic resonance cholangiopancreatography (MRCP) revealed dilation of the common bile duct (CBD) with stenosis in the pancreatic head and diffuse narrowing of the main pancreatic duct (MPD) (Fig. 3). Fluorodeoxyglucose-positron emission tomography (FDG-PET)-CT revealed a maximum standardized uptake value of 3.6-4.1, which was not localized in the pancreatic head lesion. No significant accumulation of FDG was observed in other organs (Fig. 4).

Since the findings were atypical for pancreatic cancer, and serum immunoglobulin G subclass 4 (IgG4) was elevated (869 mg/dL [normal range, 5-117 mg/dL]), endoscopic ultrasonography-guided fine-needle aspiration (EUS-FNA) was performed for a definitive diagnosis. EUS revealed a 28×27-mm enlarged hypoechoic pancreatic head compressing the distal CBD. Because there were blood vessels around the lesion, EUS-FNA was performed using a 25-gauge needle (Fig. 5A, B). Pathology and cytology of EUS-FNA specimens demonstrated only mild inflammatory cells infiltration of the stroma without plasma cells or malignancy cells. (Fig. 5C).

Endoscopic retrograde cholangiopancreatography (ERCP) showed narrowing of the MPD in the body (red arrows). No disruption of the MPD or communication between the cyst and MPD could be identified. (Fig. 6). Since the narrowing of the MPD was very severe, we decided that endoscopic nasopancreatic drainage or pancreatic duct stenting would be technically difficult. ERCP also showed smooth stricture in the distal CBD (yellow arrows); therefore, a drainage tube was inserted. Biliary tract and pancreatic fluid cytology revealed no malignancy. A biopsy of the major duodenal papillae also demonstrated no plasma cell infiltration or malignancy.

AIP was suspected because of the enlarged pancreatic head, narrowing of the MPD on ERCP and MRCP, elevated serum IgG4 levels, and no malignant findings on EUS-FNA. Subsequently, we administered steroids (30 mg prednisolone daily) to the patient with his informed consent. Eleven days after the start of steroid therapy, CT showed shrinkage of the pancreatic cyst and a decrease in ascites. A capsule-like rim structure was observed around the pancreatic tail (Fig. 7).

Based on the pre-treatment images, pathological findings,
Figure 3. Abdominal magnetic resonance imaging (MRI) and magnetic resonance cholangiopancreatography (MRCP). A, B: MRI showed a hyperintense area on the head of the pancreas with diffusion-weighted imaging (DWI) and a hypointense area with an ADC map (red arrows). C: MRCP shows diffuse narrowing of the main pancreatic duct. In the caudal cysts, penetration between the expanded pancreatic duct and the cyst was observed (yellow arrows). Stenosis of the distal bile duct was also observed (green arrows).

and good response to steroid treatment, type I AIP was established as the final diagnosis, according to the International Consensus Diagnostic Criteria for Autoimmune Pancreatitis (1). The serum IgG4 levels decreased during steroid treatment, and the pancreatic cysts and ascites completely disappeared on CT two months later. ERCP performed two months later showed improvement in the MPD and CBD narrowing (Fig. 8). There has been no recurrence of AIP for three years following steroid administration.

Discussion

AIP is pathologically characterized by a high degree of lymphocytes and plasma cell infiltration with fibrosis and usually responds dramatically to steroid treatment. Typical imaging findings in AIP include speckled/dotted enhancement and capsule-like rim structures on contrast-enhanced CT and MRI and narrowing of the MPD on MRCP and ERP (1). In addition, approximately 80% of AIP cases have bile duct stenosis, making it necessary to differentiate them from pancreatic and bile duct cancers (2). Imaging findings of AIP often do not provide a typical picture and can show diverse characteristics, including localized swelling of the pancreas and pseudocysts in 19.6% of AIP patients and calcifications in the pancreatic parenchyma in 18% of AIP patients (3). In our case, bile duct dilatation with stenosis of the pancreatic head, a large amount of ascites, and multiple pancreatic cysts were observed. Therefore, although AIP was the primary entity suspected, we considered the possibility of pancreatic cancer or cholangiocarcinoma in the differential diagnosis. A series of examinations were atypical for pancreatic cancer and cholangiocarcinoma and suggested the possibility of AIP. The serum IgG4 levels were elevated to more than twice the normal value, diffuse pancreatic duct stenosis was observed by MRCP and ERP, and EUS-FNA showed no malignancy. A steroid trial was performed with the patient’s informed consent, which resulted in the improvement of pancreatic swelling, reduction of cysts, and improvement of ascites. Based on the various findings and clinical course, we finally diagnosed the patient with type I AIP.

AIP associated with complications, such as massive ascites and multiple cysts, is rare. In general, ascites due to pancreatitis are caused by inflammation in the abdominal cavity, rupture of pancreatic cysts, or collapse of the pancreatic duct (4). In cases with a chronic pancreatitis background, internal fistulas tend to form due to a lack of inflammation around the pancreas, and incomplete encapsulation of pancreatic juice leakage caused by pancreatic duct disruption may result in massive ascites (5). However, AIP can reportedly cause stenosis of the surrounding vessels due
Figure 5. A, B: An endoscopic ultrasonography (EUS)-guided fine-needle aspiration biopsy (EUS-FNA). EUS revealed a 28×27-mm hypoechoic pancreatic head. EUS-FNA was performed on the lesions using a 25-gauge needle. C: Pathology and cytology showing fibrosis and infiltration of inflammatory cells but no plasma cell infiltration or malignancy (Hematoxylin and Eosin staining, ×400).

Figure 6. Endoscopic retrograde cholangiopancreatography (ERCP). ERCP showed narrowing of the main pancreatic duct (MPD) in the body (red arrows) without disruption of the MPD or communication between the MPD and cyst. ERCP revealed smooth distal common bile duct (CBD) stricture (yellow arrows).

to the spread of inflammation to the surrounding area. According to Ishikawa et al. (6), approximately 44% of patients with AIP have vascular lesions, and most of them have splenic vein stenosis. The large amount of ascites in our patient may have been due to collapse of the pancreatic duct or cyst in response to the increased pressure in the MPD. There may also have been some effect of portal hypertension due to obstruction of the splenic vein.

Steroid treatment is commonly used for type I AIP. It is effective even for the treatment of pseudocysts associated with AIP. According to Kubota et al. (7), although steroid treatment appears to be beneficial in patients with type 1 AIP with cyst diameters <55 mm, those with large cysts (≥55 mm in diameter) tend to be refractory to steroid treatment. Considering the above reports, patients with giant cysts should be considered for not only steroid therapy but
Computed tomography (CT) 11 days after the start of treatment. CT showed disappearance of the pancreatic cysts and improvement in ascites. A capsule-like rim structure was observed around the pancreatic tail.

Figure 8. Endoscopic retrograde cholangiopancreatography (ERCP) (two months after the start of treatment). ERCP performed two months later showed improvement in MPD and CBD narrowing.

AIP occasionally resolves spontaneously. In our patient, CT at the previous institution showed massive ascites, but CT at the time of admission showed a decrease in ascites. This course is characteristic of AIP in that it resolved spontaneously. In contrast, according to Kubota et al. (8), elevated serum IgG4 levels and the presence of jaundice were independent factors predicting recurrence in AIP patients with AIP. In our case, the serum IgG4 levels were elevated and subjective symptoms were exacerbated; therefore, steroid treatment was administered. Why steroid treatment alone was successful is unclear, but a possible reason may be that steroid treatment was administered at a stage when the AIP was in spontaneous remission.

In our case, cysts and ascites were dramatically reduced by steroid therapy alone. Only one case of AIP complicated by ascites and multiple cysts has been previously reported (9). This case was complicated by ascites due to pancreatic fluid leakage, which required steroid therapy and pancreatic duct stenting. As mentioned above, although steroids can be used for the treatment of AIP with ascites and cysts, additional endoscopic or surgical treatment should be considered at an early stage if conservative treatment seems ineffective.

The authors state that they have no Conflict of Interest (COI).

Contributing Authors
Koyo K contributed to the conception and design of the study, analysis, and collection and assembly of the data drafting of this article. Reiko Y contributed to the critical revision of the article for important intellectual content. All authors critically revised the report, commented on the drafts of the manuscript, and approved the final report.
References


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