Immunoglobulin G4 (IgG4)-related disease is a newly recognized fibroinflammatory condition that is characterized by dense infiltration of T lymphocytes and IgG4-positive plasma cells with associated fibrosis [1]. Autoimmune pancreatitis (AIP) involves typical pancreatic lesions caused by this systemic disease, and concomitant bile duct involvement has also been observed [2]. To avoid unnecessary surgery, characteristic findings on magnetic resonance cholangiopancreatography (MRCP) are used to differentiate between AIP and pancreatic cancer [3–6]. Among the noninvasive imaging techniques available, MRCP is unique in that it enables visualization of pancreatic, intrahepatic, and extrahepatic bile duct stenoses. In our case described herein, serial MRI and MRCP investigations demonstrated the progression of untreated IgG4-related disease and reversal of the features after steroid treatment, although the serum IgG4 level remained abnormally elevated.

**CASE REPORT**

The patient was a 50-year-old woman who had a 4-month history of epigastralgia and had jaundice for 2 weeks prior to her first admission. Icteric sclerae were also noted during the physical examination. The initial laboratory tests showed elevated levels of bilirubin (total bilirubin level/direct bilirubin level, 10.9/6.2 mg/dL) and impaired
Figure 1
liver function, with aspartate aminotransferase (AST) and alanine aminotransferase (ALT) levels of 60 and 149 IU/L, respectively. However, the levels of amylase and lipase were within the normal ranges. Contrast-enhanced abdominal computed tomography (CT) results showed a diffusely enlarged pancreas, which was surrounded by a poorly enhanced capsule-like rim (Fig. 1a). AIP was suspected, and MRI was suggested for further evaluation. On MRI, the diffusely swollen pancreatic parenchyma exhibited a hyperintense appearance in the T2-weighted images (Fig. 1b). In the dynamic contrast-enhanced T1-weighted images, delayed pancreatic parenchymal enhancement was detected with a peripancreatic hypointense rim (Fig. 1c, 1d, 1e). Wall thickening and enhancement of the distal common bile duct were also noted in the post-enhanced T1-weighted images. Diffusion-weighted imaging (DWI) showed diffuse high signal intensity in the pancreas due to water restriction (Fig. 1f). The mean apparent diffusion coefficient (ADC) value of the pancreatic head, body, and tail was $1.003 \times 10^{-3} \text{mm}^2/\text{s}$ in our 3.0T MR scanner. Stenosis of the proximal main pancreatic duct and the distal common bile duct was noted on MRCP (Fig. 1g). A diagnosis of AIP was suggested on the basis of these characteristic findings.

The patient was discharged soon after her jaundice was relieved by percutaneous transhepatic gallbladder drainage. She was readmitted 2 months after discharge due to worsening symptoms. Laboratory data showed persistently elevated levels of bilirubin (total bilirubin level/direct bilirubin level, 4.5/2.2 mg/dL) and impaired liver function (AST level, 140 IU/L; ALT level, 208 IU/L). Repeat MRI and MRCP examinations showed a further decrease in ADC value ($0.914 \times 10^{-3} \text{mm}^2/\text{s}$) and multiple progressive biliary stenoses involving the intrahepatic bile ducts (Fig. 2a, 2b).

To rule out the possibility of periampullary malignancy, a pancreatic head biopsy with bypass cholecystodudodenostomy was performed. Histological findings showed periductal infiltrates composed of lymphocytes and plasma cells, with marked IgG4-positive plasma cell infiltration (Fig. 3). Laboratory data showed a substantially increased serum IgG4 level (4150 mg/L). AIP was diagnosed on the basis of the Mayo Clinic histology, imaging, serology, other organ involvement, and response to steroid therapy (HISORt) criteria [7, 8].

After 8 weeks of steroid therapy (prednisolone: 30 mg/day for 6 weeks, followed by 15 mg/day for 2 weeks), the patient’s symptoms improved, although her serum IgG4 level did not decrease (4340 mg/L). Her bilirubin level and liver function improved, with values that were near the normal range (total bilirubin level/direct bilirubin level, 1.5/0.2 mg/dL; AST level, 20 IU/L; ALT level, 29 IU/L). Follow-up MRI and MRCP studies indicated that the size of the pancreas had returned to normal, which was accompanied by a normal ADC value ($1.508 \times 10^{-3} \text{mm}^2/\text{s}$) (Fig. 2c). The stenosis of the proximal pancreatic duct and the multifocal stenoses of the intra- and extrahepatic bile ducts had also improved (Fig. 2d).

**DISCUSSION**

IgG4-related disease is a recently described disease entity that is characterized by tissue infiltration of IgG4-positive plasma cells and T lymphocytes into single or multiple organs. AIP is the prototypical form of IgG4-related disease. A study of patients with AIP showed frequent extrapancreatic involvement, including hilar lymphadenopathy (80%), extrapancreatic bile duct lesions

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**Figure 1. a.** Axial contrast-enhanced CT scan shows diffuse enlargement of the pancreas with homogeneous attenuation and the peripheral hypoattenuation halo. **b.** Axial T2-weighted fat-suppressed image shows pancreatic parenchyma appears hyperintense compared with liver parenchyma. Precontrast **c.** arterial **d.** and delayed **e.** axial contrast-enhanced fat-suppressed T1-weighted images demonstrate delayed pancreatic parenchymal enhancement. **f.** The diffusion-weighted imaging (DWI) shows a diffusely swollen pancreas with a high-signal-intensity area ($b = 1000 \text{mm}^2/\text{s}$). **g.** Magnetic resonance cholangiopancreatography (MRCP) shows narrowing of the distal common bile duct and proximal pancreatic duct with upstream biliary duct dilatation.
Unlike acute pancreatitis, severe abdominal pain with elevated serum amylase and lipase in levels is unusual in patients with AIP. Patients with AIP typically present with chronic upper abdominal pain and jaundice [10]. Accurately distinguishing AIP from other forms of chronic pancreatitis and pancreatic carcinoma can not only avoid unnecessary radical surgery such as the Whipple operation but also facilitate timely initiation of steroid therapy.

The diagnostic criteria for AIP proposed by the Mayo Clinic (the HISORt criteria) are commonly used in clinical practice and include the presence of 1 or more of the following: histological features suggestive of AIP, pancreatic imaging features suggestive of AIP, elevated serum IgG4 levels that are ≥2 times the upper limit of the normal range, involvement of organs other than the pancreas, and response to glucocorticoid therapy [7, 8]. Typical CT findings include diffuse or focal enlargement of the pancreas with delayed enhancement with or without a capsule-like rim [11]. Several MRI sequences, including MRCP and DWI, can provide useful findings that are suggestive of AIP. MRCP clearly delineates the irregular narrowing of the main pancreatic duct and the intra- and extrapancreatic bile ducts. DWI, on the other hand, can demonstrate the characteristic high-signal-intensity area of the pancreas, which may persist even after steroid treatment.

**Figure 2.**

- **a.** DWI obtained 2 months later (prior to steroid treatment) persistently shows a diffusely swollen high-signal-intensity area.
- **b.** MRCP shows progressive multifocal strictures of the intrahepatic bile ducts.
- **c.** and **d.** Two months after steroid therapy, DWI indicates that the thickness of the pancreatic parenchyma has returned to normal, with markedly decreased signal intensity. MRCP shows improvement in the multifocal strictures in the extra- and intrahepatic bile ducts and the proximal pancreatic duct.
extrahepatic bile ducts [3]. DWI, a recent technical development in MRI, has been used to evaluate diseases involving abdominal organs. On DWI, AIP is observed as either focal or diffuse high-signal-intensity areas in the pancreas due to water restriction. Only a few DWI-based studies have been performed for distinguishing between AIP and pancreatic cancer [4–6]. Terumi et al. reported that AIP is associated with lower ADC values than pancreatic cancer, and they suggested the establishment of an ADC cutoff value for distinguishing AIP from pancreatic cancer; however, a consensus ADC cutoff value has not yet been achieved [4].

Unlike most other cases, in this case, the serum IgG4 level did not decrease after treatment, despite the improvement in clinical symptoms. Therefore, imaging studies play an increasingly important role in monitoring the response to treatment. Several of the sequences in MRCP are useful for evaluating disease activity; most importantly, MRCP does not expose patients to radiation, unlike CT. Our patient clearly showed progressive multifocal skipped intrahepatic bile duct narrowing prior to steroid treatment. After treatment, the thickness of the pancreatic parenchyma returned to normal. During DWI, the signal intensity in the involved area showed a significant decrease from that before treatment, and the ADC values of the area involved increased to values similar to those obtained from a normal pancreas. In addition, stenoses of the extra- and intrahepatic bile ducts and pancreatic duct disappeared. This reversal in features that occurred after treatment indicate that MRI studies, including the MRCP sequence, are critical and likely superior to serum IgG4 level measurement for evaluating treatment effects and monitoring disease activity.

In conclusion, we report a rare case of IgG4-related disease involving the bile ducts and pancreas. In our case, the serum level of IgG4 was not a reliable marker for reflecting the disease activity following treatment. Therefore, MRI and MRCP may serve as a “one-stop shop” modality for the diagnosis and follow-up of patients with this relatively rare clinical condition.

REFERENCES