

# A Case of Type 1 Autoimmune Pancreatitis

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## Abstract

Autoimmune pancreatitis (AIP) is a form of pancreatitis with clinical, serological and histological features of an autoimmune process. This is the case of a 32-year-old diabetic man who had been suffering from painless jaundice with pale stools and tea-colored urine for 3 months prior to consultation. An endoscopic retrograde cholangiopancreatography revealed a proximal common bile duct stricture, and magnetic resonance cholangiopancreatography showed a bulky pancreatic head. His serum amylase level was mildly raised, and his serum IgG titer was markedly elevated. The serum alkaline phosphatase and conjugated bilirubin levels were high, suggesting an obstructive jaundice. A diagnosis of AIP was made and treated with steroids. He responded well to the steroid therapy.

## INTRODUCTION

Autoimmune pancreatitis is a rare condition in contrast to other autoimmune disorders. It affects men more often than women (ratio 2:1) and up to 70–80% of patients present with painless jaundice (Buscarinia et al., 2010). Marked cachexia, inability to eat and narcotic-requiring pain are more suggestive of pancreatic cancer (Shimosegawa et al., 2011), on the contrary, but the case is considered autoimmune due to its dramatic response to steroid therapy. Currently, elevated serum IgG<sub>4</sub> levels are considered to be the sole serological hallmark of AIP (Buscarinia et al., 2010). This is a case of autoimmune pancreatitis reported to Universiti Kebangsaan Malaysia Medical Centre (UKMMC) for diagnosis and management.

## CASE PRESENTATION

This 32-year-old diabetic gentleman presented to UKMMC for diagnosis and management of autoimmune pancreatitis. The patient had suffered from painless jaundice for three months prior to hospitalization. The patient's stool was pale with tea-colored urine. There were no symptoms of vomiting or anorexia and no alteration in bowel habits. He had no history of fever. He had no viral hepatitis and had not been admitted to a hospital for pancreatitis before. He was a long-term smoker and alcoholic.

The first radiological investigation was performed by endoscopic retrograde cholangiopancreatography (ERCP) and revealed a proximal common bile duct stricture, possibly secondary to external compression. Following this, a

magnetic resonance cholangiopancreatography was done, and we observed a bulky pancreatic head and a thickened gallbladder wall with a gallstone inside. The blood investigations were performed, and the results are presented in Table 1.

## Figure 1

Table 1: Baseline blood investigation results of patients with type 1 autoimmune pancreatitis

Parameter	Result	Reference range	Flag
Amylase	157 IU/L	< 115 IU/L	Mildly elevated
Albumin	30 g/L	35-50 g/L	Slightly low
Globulin	45 g/L	23-35 g/L	Elevated
Total bilirubin	375 µmol/L	< 17 µmol/L	Markedly elevated
Conjugated bilirubin	275 µmol/L	< 3 µmol/L	Markedly elevated
Alanine aminotransferase	63 IU/L	30 – 65 IU/L	Normal
Alkaline phosphatase	223 IU/L	50 – 136 IU/L	Markedly elevated
Gamma glutamyl transferase	274 IU/L	15 – 85 IU/L	Markedly elevated

Results indicated a mild form of pancreatitis as evidenced by the raised amylase. The globulin level was raised, which indicated that the serum antibody level was higher than usual. The elevated ALP (with a normal ALT) suggests that the grossly raised conjugated bilirubin level in this patient was secondary to an obstruction of the biliary tree rather than hepatitis.

The patient was provisionally diagnosed to have autoimmune pancreatitis. Antinuclear antibody of the patient was negative, in contrast, immunoglobulin G level was found to be 3220 mg/dL.

Abdominal CT scan results revealed that his pancreas was diffusely enlarged, especially at the tail. No focal pancreatic lesions were seen. The pancreatic duct was irregularly narrowed. Hepatosplenomegaly was present and a gallstone was noted in the gallbladder.

He was started on oral prednisolone 30 mg daily for 2 weeks, followed by 25 mg daily for a week and then 20 mg daily for a week. After two weeks on prednisolone, a repeat CT scan of the abdomen showed a reduction in the pancreatic mass. A further reduction in the pancreatic mass size was observed 6 months later.

### Figure 2

Table 2: Radiological findings of pancreas before and after commencing prednisolone therapy

Region of pancreas	Before commencing prednisolone	2 weeks after commencing prednisolone	6 months after commencing prednisolone	Reference range*
Head	3.4 cm	3.1 cm	1.2 cm	1.0 – 2.0 cm
Neck	2.5 cm	1.9 cm	-	-
Body	3.2 cm	2.2 cm	1.8 cm	0.4 – 1.0 cm
Tail	3.6 cm	2.7 cm	1.9 cm	0.8 – 2.2 cm

\* As used by the radiology department of UKMMMC

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During follow up, the IgG level of the patient showed a declining trend as presented below:

### Figure 3

Marker	At diagnosis	6 months after diagnosis	12 months after diagnosis	Reference range
Serum IgG	3220 mg/dL	2220 mg/dL	1850 mg/dL	751 - 1560 mg/dL

Finally, he was put on a maintenance dose of oral prednisolone 5 mg daily.

## DISCUSSION

The term autoimmune pancreatitis (AIP) was first proposed by the Japanese in 1995 (Yoshida et al., 1995) who described a case of steroid-responsive pancreatitis. Type 1 AIP is also known as lymphoplasmacytic sclerosing pancreatitis. It is an uncommon condition, accounting for 2% of chronic pancreatitis cases (Zen et al., 2011). The major symptom at onset is obstructive jaundice due to the enlargement of the pancreatic head or thickening of the bile duct wall. The disease is characterized by elevated serum IgG<sub>4</sub> levels and extrapancreatic lesions (e.g., sclerosing cholangitis, sclerosing sialadenitis, and retroperitoneal fibrosis). However, the clinical diagnosis of type 1 AIP can often be made without the study of histology (Shimosegawa

et al., 2011).

Pancreatic findings on abdominal CT or MRI are often the first clues that raise the suspicion of pancreatic cancer or AIP. Patients with a diffusely enlarged pancreas, but without pancreatic ductal dilatation and without a low density mass on CT/MRI are highly likely to have AIP. Where there is doubt, the patient should be considered as having pancreatic cancer unless the workup for cancer is negative and there is strong collateral evidence for AIP (Shimosegawa et al., 2011).

It is fortunate that this patient had a diffuse pancreatic lesion instead of a focal one, as the latter can complicate diagnosis. The focal form, particularly in the presence of a low-density pancreatic mass may be easily confused with pancreatic cancer (Frulloni, 2011). In patients with appropriate collateral evidence of AIP, response to steroids can confirm a suspicion of AIP (Shimosegawa et al., 2011). Treatment by resection is not recommended for type 1 AIP (Zen et al., 2011). Steroid response is characterised by improvement in imaging abnormalities (e.g. biliary strictures and pancreatic enlargement), a decline in IgG<sub>4</sub> titres (Shimosegawa et al., 2011) and a decrease in CA 19-9 levels (Moon et al., 2008). However, symptomatic improvement and a sense of well-being occur non-specifically in response to steroids and can be seen even in pancreatic cancer patients. Therefore, these parameters should not be used to assess response (Shimosegawa et al., 2011). Relapses occur in up to 54% of patients and recurrences seem to be more frequent in focal AIP than in diffuse AIP (Frulloni et al, 2009).

Although the patient in this case report is relatively young (32 years old), he is likely to have type 1 AIP as his IgG level was markedly elevated. In UKMMMC, the facility to perform IgG<sub>4</sub> measurement is unavailable and thus total serum IgG was measured. It is also possible that this patient has sclerosing cholangitis as ERCP revealed the presence of a proximal common bile duct stricture. The finding of an extrapancreatic manifestation lends further support to type 1 AIP. He responded well to a trial of oral prednisolone as evidenced by a progressive decline in IgG titres and a reduction in the size of his pancreas.

Diabetes mellitus is a common complication of AIP, and is seen in about half of the patients (Zen et al., 2011). Although our patient is also diabetic, his diagnosis of AIP was made a few years after he was diagnosed with diabetes mellitus.

### CONCLUSION

AIP is suspected in patients showing anomaly or reversal of the albumin/globulin ratio with initial investigation. A pancreatic biopsy might be useful; however, this invasive investigation is not mandatory if there are other features: radiological findings and a raised IgG or antinuclear antibody titer. In AIP management, a trial of steroids is warranted and long-term follow up is desirable as this disease is prone to relapse or recur.

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