

# Probable Type 2 Autoimmune Pancreatitis: A Case Report and Clinical Management

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**Abstract** Autoimmune pancreatitis (AIP) is a rare type of chronic pancreatitis characterized by immune-mediated inflammation of the pancreas. It is often misdiagnosed due to non-specific symptoms such as abdominal pain, congestion, jaundice and radiographic findings that mimic pancreatic malignancy. Without appropriate treatment, AIP can lead to endocrine and exocrine pancreatic insufficiency, biliary complications, and irreversible fibrosis(1). In this article, we aimed to increase awareness of the clinical approach to IgG4 –unrelated autoimmune pancreatitis that mimics pancreatic malignancy

**Keywords:** Autoimmune pancreatitis, Steroid therapy, Type 2 AIP, Pancreatic inflammation

## 1. Introduction

Autoimmune pancreatitis (AIP) is a form of chronic pancreatitis that develops as a result of an autoimmune inflammatory process characterized by dense lymphocytic infiltration and fibrosis leading to organ dysfunction (2). Autoimmune pancreatic disease is the only pancreatic condition treated effectively with corticosteroids. Three types of AIP are clinically recognized:

Type 1: IgG4 –related pancreatitis associated with a serum IgG4 concentration greater than twice the normal reference range in most affected individuals. The pancreas may be the only organ affected, or there may also be other organs involved with corresponding clinical presentations, including(3):

- Bile ducts (secondary sclerosing cholangitis)
- Retroperitoneal fibrosis
- Kidneys



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- Lungs
- Submandibular and parotid glands (Sjogren disease)
- Sublingual glands (sclerosing sialadenitis)
- Orbits and lacrimal glands
- Joints (rheumatoid arthritis)

Type 2: Histologically this is idiopathic duct-centric pancreatitis with granulocytic epithelial lesions within the pancreatic duct, small numbers of IgG4 – positive plasma cells (fewer than 10/HPF) and no extra pancreatic involvement. Type 2 AIP is often associated with inflammatory bowel disease and is twice as common in patients with ulcerative colitis or proctitis as in those with Crohn disease (4). Unlike patients with type 1 AIP, about half of those with type 2AIP present with abdominal pain or acute pancreatitis.

Type 3: Immune checkpoint inhibitor – induced pancreatic injury is a type of progressive immune –mediated pancreatitis, an adverse effect of cancer treatment with immune checkpoint inhibitor drugs. The risk of this immune response is heightened when multiple immune checkpoint inhibitors are used (5). Most patients with type 3 AIP are asymptomatic.

Our clinical case involved a middle –aged patient in whom type 2 autoimmune pancreatitis with isolated pancreatic involvement was identified and a dramatic clinical response was achieved with steroid therapy.

## 2. Case Report

A 42 –year – old male patient presented to us with complaints of jaundice, band-like pain in the epigastric region and nausea. The patient had experienced epigastric pain lasting 2-3 days and diarrhea lasting one day approximately 1.5 months earlier. There is no history of significant comorbid disease or prior surgery. The family history is negative for autoimmune diseases. He does not smoke or consume alcohol.

On physical examination, the patient's general condition was assessed as moderately, severe. The skin and visible mucous membranes were icteric. On abdominal palpation there was marked tenderness in the epigastric region.

### 2.1. Laboratory findings

Test name	Result	Unit	Reference range
ALP	510	IU/L	41- 137
ALT	211	U/L	5-41
AST	60	U/L	5-40
Total bilirubin	10.68	mg/dL	<1.2
Direct bilirubin	9.41	mg/dL	<0.2
Serum lipase	41.42	U/L	13-60

Pancreatic amylase	13.45	U/L	0-53
CRP	3.57	mg/L	0-5
CA 19-9	661.7	U/mL	<37
IgG4	1.20	g/L	0-1.25
WBC	6.22	k\mm <sup>3</sup>	4.5-11
HGB	15.0	g\dl	13.5-17.5
MCV	87.8	pg	76-100
PLT	160	k\mm <sup>3</sup>	140-400

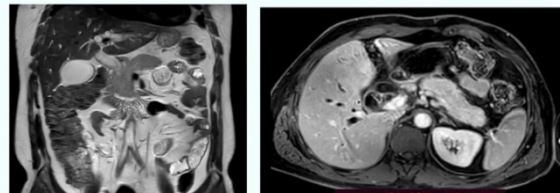
EUS examination: A mass measuring 35 x 27 mm was identified in the pancreas; intrahepatic and extrahepatic bile ducts are dilated.

During gastroscopy, erosive gastritis was observed, duodenum appeared deformed and scarred.

On abdominal MRI, the pancreas is enlarged. On T2- weighted sequences, the pancreatic parenchyma is slightly hyperintense. Peripancreatic edema is present. Intrahepatic bile ducts are dilated. The common bile duct measures 16 mm; no stones are seen. The spleen is normal.

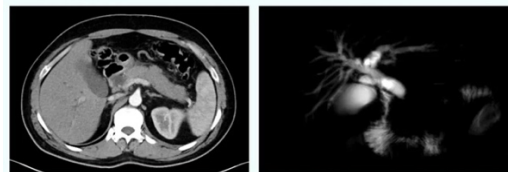
MRCP shows dilatation of the intrahepatic and extrahepatic bile ducts. The common bile duct is dilated with smooth distal narrowing. No stones are seen.

### MRT



*Image 1. Abdominal MRI*

### KT vø MRCP



*Image 2. Abdominal CT and MRCP*

## 2.2. Treatment

Steroid therapy was initiated in the patient with suspected autoimmune pancreatitis.

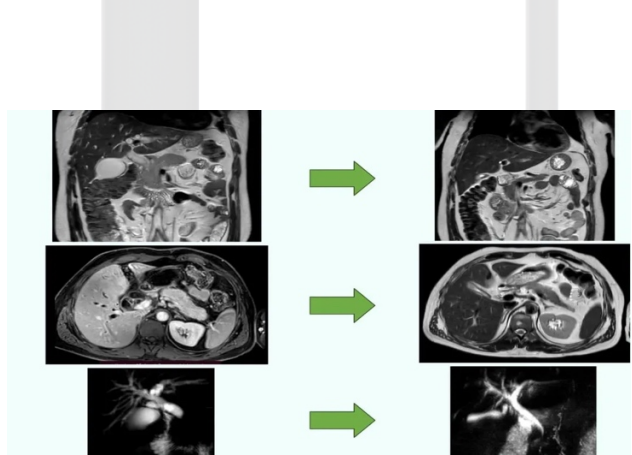
1. Prednol 16 mg 1x2
2. Calcium 500 mg 1x2
3. PPI 40 mg 1x1
4. UDCA 500mg 1x2

The patient was scheduled for regular follow-up examinations. The steroid dose was gradually reduced after 1.5 months.

At 1.5 month after treatment, when the patient presented for follow-up, significant improvement was observed in clinical symptoms as well as in laboratory and instrumental findings. The results of the

follow-up examinations performed two months later are summarized below.

Test name	Result	Unit	Reference range
ALP	95	IU/L	41-137
ALT	48.2	U/L	5-41
AST	17.2	U/L	5-40
Bilirubin total	0.51	mg/dL	<1.2
Direct bilirubin	0.22	mg/dL	<0.2
GGT	29	U/L	3-60
CA19-9	9.82	U/ml	<37
WBC	7.4	k/mm <sup>3</sup>	4.5-11
HGB	15.7	g/dL	13.5-17.5
PLT	250	Lk/mm <sup>3</sup>	140-400
ESR	8	mm/h	0-15



**Image 3.** The biliary ducts are within normal limits and the previously noted diffuse pancreatic enlargement has significantly regressed.

### 3. Discussion

Type 2 autoimmune pancreatitis (AIP-2) is a rare pancreatic disease characterized by normal IgG4 levels and usually without systemic organ involvement. Therefore, the diagnostic and therapeutic management of AIP -2 differs from type 1 AIP. Steroid therapy is considered the main treatment for AIP -2 and allows patients to achieve rapid remission.

In this case, the clinical presentation and imaging findings were consistent with autoimmune pancreatitis. However, due to the lack of histopathological confirmation, the diagnosis was considered probable. The rapid response to steroid therapy further supported this diagnosis.

Given the elevated CA19-9 levels and mass-line imaging findings, alternative diagnoses such as pancreatic malignancy and cholangiocarcinoma were considered. Careful evaluation and the patient's rapid response to steroid therapy supported the diagnosis of probable type 2 autoimmune pancreatitis.

In a study titled "Clinical features and long-term outcomes of patients with type 2 autoimmune pancreatitis", 88 patients were observed. Patients receiving steroid therapy had a 5-year relapse rate of approximately 13%, whereas those who underwent surgical intervention had a relapse rate of 33%. These

results indicate that steroid therapy is effective not only in controlling the disease but also in reducing long-term relapse risk (6).

Another study, "Type 2 Autoimmune Pancreatitis (Idiopathic Duct-Centric Pancreatitis): A Single-Center Experience," analyzed 27 patients. Of these, 96.3% received steroid therapy and almost all achieved remission. During a median follow-up of 32 months, only two patients experienced relapse, both of which responded effectively to retreatment with steroids (7).

In our case, a 42-year-old male patient was observed. He started steroid therapy at 32 mg and achieved remission within 1.5 months, with the dose gradually tapered after 1.5 months. No relapse was observed.

Rapid achievement of remission, infrequent relapses and gradual tapering of steroids represent an effective clinical management strategy. The absence of comorbidities in our patient and his overall health support the favorable response to therapy.

#### 4. Conclusion

This clinical case highlights the diagnostic challenges of autoimmune pancreatitis mimicking pancreatic malignancy. Isolated pancreatic involvement and the dramatic response to steroid therapy support the likelihood of type 2 autoimmune pancreatitis. Assessing clinical presentation, radiological findings and treatment response together plays a crucial role in establishing an accurate

diagnosis. Early recognition of autoimmune pancreatitis can prevent unnecessary invasive procedures and allow for effective management.

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