Case Report

The Great Masquerader: Autoimmune Pancreatitis Presenting as an Obstructive Pancreatic Head Mass and the Avoidance of a Whipple Procedure

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Submission Date : 12 July 2025
Accepted Date : 06 August 2025
Published Date : 30 September 2025
DOI: https://doi.org/10.3329/jrpmc.v10i2.85693

Abstract

Autoimmune pancreatitis (AIP) is a unique, steroid-responsive focal form of chronic pancreatitis, particularly in the pancreatic head, often presents as a mass-forming lesion with obstructive jaundice, perfectly mimicking pancreatic adenocarcinoma. Distinguishing between these two entities is paramount, as their management is radically different. A 45-year-old male presented with a three-week history of painless jaundice, pruritus, and weight loss. Computed Tomography (CT) revealed a hypodense mass in the pancreatic head measuring 3.5 cm, with abrupt cutoff of the common bile duct and double-duct sign, accompanied by proximal biliary and pancreatic duct dilatation. Serum Carbohydrate Antigen 19-9 (CA 19-9) was elevated at 250 U/mL. The patient was scheduled for a pancreaticoduodenectomy (Whipple procedure). Prior to surgery, a review of the CT by a dedicated pancreatic radiologist noted a subtle "halo sign" around the pancreas. This prompted further testing, which revealed a serum IgG4 level of 650 mg/dL. A FNA cytology via ultrasonogram showed lymphoplasmacytic infiltration and >50 IgG4-positive plasma cells per high-power field, consistent with AIP Type 1. The Whipple procedure was canceled. The patient was started on oral prednisone (40 mg/day). His jaundice resolved within two weeks. A repeat CT scan after 4 weeks of therapy showed a dramatic reduction in the size of the pancreatic head mass and resolution of the biliary dilatation. CA 19-9 normalized. The steroid dose was tapered, and the patient was maintained on azathioprine. He remains asymptomatic at one-year follow-up. This case highlights that AIP must be a mandatory consideration in every patient with a suspected pancreatic head malignancy. The presence of a “halo sign” on imaging and elevated serum IgG4 is crucial diagnostic red flags. A definitive tissue diagnosis is often necessary to prevent unnecessary and morbid surgery. A high index of suspicion and a multidisciplinary approach are essential.

Keywords: Autoimmune pancreatitis, Pancreatic cancer, IgG4, Whipple procedure, Obstructive jaundice, Steroid therapy.

Citation: Hossain MS, Ali MS,Lucky JA. The Great Masquerader: Autoimmune Pancreatitis Presenting as an Obstructive Pancreatic Head Mass and the Avoidance of a Whipple Procedure. J Rang Med Col. 2025 Sep;10(2):166-169. doi: https://doi.org/10.3329/jrpmc.v10i2.85693

Introduction:

Pancreatic ductal adenocarcinoma (PDAC) is a lethal disease where timely surgical resection offers the only chance for cure. Consequently, a mass in the pancreatic head with obstructive jaundice is often presumed to be PDAC until proven otherwise. Autoimmune Pancreatitis (AIP), particularly its focal form, is the great benign imitator of PDAC.[1] Type 1 AIP, the pancreatic manifestation of the systemic IgG4-related disease (IgG4-RD), responds dramatically to corticosteroid therapy.[2] Failing to recognize AIP can lead to unnecessary pancreatic resection, with its associated significant morbidity and mortality, for

a condition that is medically treatable. We present a case that narrowly avoided this outcome.

Case Presentation:

A 45-year-old man, a former smoker, presented with progressive, painless jaundice, dark urine, and severe pruritus of three weeks duration. He reported an unintentional weight loss of 6 kg over the past two months. His past medical history was unremarkable. Physical examination revealed deep scleral icterus and excoriations but no palpable abdominal mass or lymphadenopathy. Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Laboratory Tests (Table-I): Total Bilirubin 12.8 mg/dL, Direct Bilirubin 10.5 mg/dL, Alkaline Phosphatase 780 U/L, Gamma-Glutamyl Transferase 850 U/L. Amylase and Lipase were within normal limits.

Tumor Marker: CA 19-9 was significantly elevated at 250 U/mL (normal <37).

Imaging and Procedural Findings:

Contrast-Enhanced CT Abdomen: Revealed a 3.5 cm hypodense, ill-defined mass in the pancreatic head. There was abrupt narrowing of the intrapancreatic common bile duct and main pancreatic duct ("double-duct sign"), with upstream dilatation of both systems. There were no distant metastases or vascular involvement. The initial radiology report stated "Findings highly suspicious for pancreatic head carcinoma" (Figure-1).

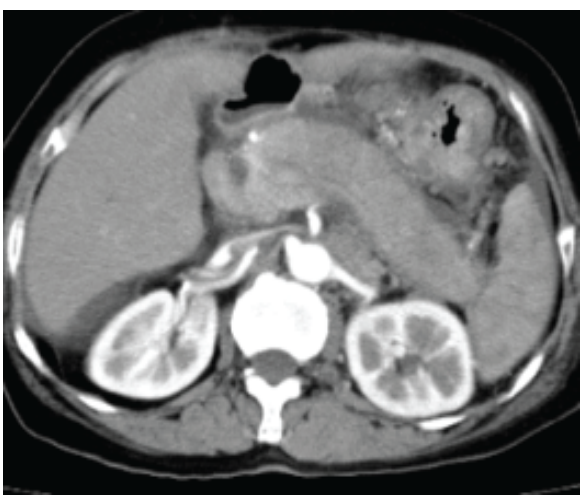

Figure-1: CT Abdomen (Axial View). Pre-treatment CT showing a hypodense mass in the pancreatic head causing biliary dilatation

ERCP with Stenting:A biliary stricture was visualized and a plastic biliary stent was placed to relieve the obstruction (Figure-2).

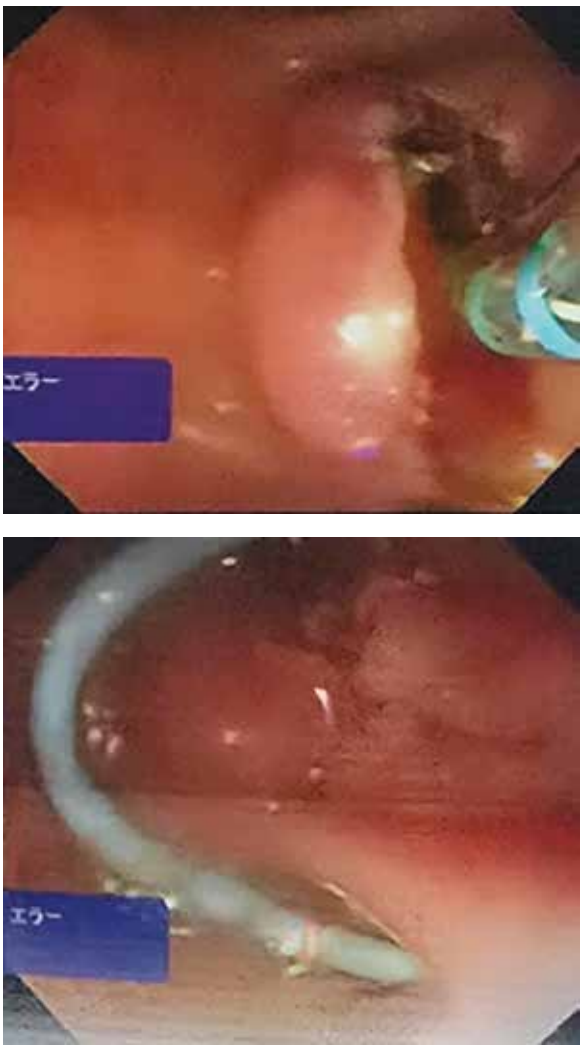

Figure-2: ERCP with biliary Plastic stenting

The patient was presented at a multidisciplinary tumor board. Given the classic imaging and cytology findings, a consensus was reached to proceed with a pancreaticoduodenectomy.

Diagnostic Turning Point:

During the pre-operative anaesthetic workup, the CT scan was reviewed again by another radiologist with a specialty in pancreatic diseases. The radiologist noted a subtle, low-attenuation rim surrounding the pancreatic mass and the rest of the gland—the "halo sign" (Figure-3). This finding is highly suggestive of AIP. This prompted a pause and further investigation.

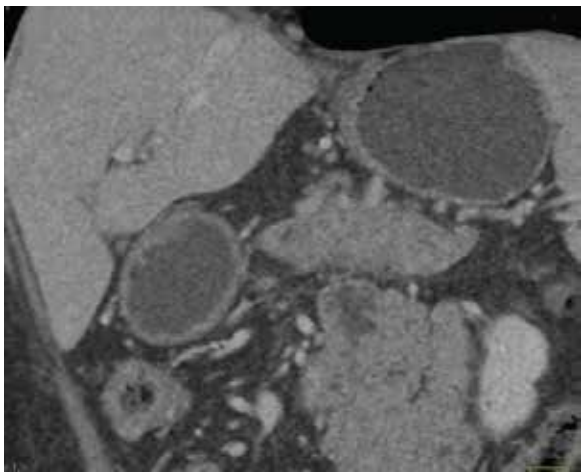

Figure-3: CT Abdomen (Coronal View). Pre-treatment image highlighting the "halo sign"

Serum IgG4: Markedly elevated at 650 mg/dL (normal <135 mg/dL).

US-guided FNA cytology: A US guided FNA cytology needle revealed dense lymphoplasmacytic infiltrate and storiform fibrosis. Immunohistochemical staining showed a prominent IgG4-positive plasma cell infiltrate, with a count of over 80 cells per high-power field. These findings are diagnostic for Type 1 AIP.

Management and Outcome

The planned Whipple procedure was immediately canceled. The patient was started on oral prednisone at 40 mg per day (0.6 mg/kg). His pruritus resolved within days, and his jaundice cleared within two weeks. Liver function tests normalized progressively. A follow-up CT scan performed after 4 weeks of steroid therapy showed

Table-I: Summary of Diagnostic Investigations and Response to Therapy

Investigation Parameter	Pre-Treatment (At diagnosis)	Post-Treatment (4-6 weeks)	Reference Range
Liver Function Tests			
Total Bilirubin	12.8 mg/dL	1.2 mg/dL	0.1 - 1.2 mg/dL
Direct Bilirubin	10.5 mg/dL	0.4 mg/dL	0.0 - 0.3 mg/dL
Alkaline Phosphatase (ALP)	780 U/L	110 U/L	40 - 120 U/L
Tumor Marker			
Carbohydrate Antigen 19-9 (CA 19-9)	250 U/mL	18 U/mL	< 37 U/mL
Serology			
Serum IgG4	650 mg/dL	Not repeated	10 - 135 mg/dL
Imaging (CT scan)			
Pancreatic Head Mass Size	3.5cm (hypodense)	1.2cm (near resolution)	Normal pancreatic anatomy
Common Bile Duct Diameter	Dilated (10 mm)	Normal (5mm)	< 6 mm
"Halo Sign"	Present	Absent	Not present
FNA Cytology			
H&E Stain	Dense lymphoplasmacytic infiltrate, storiform fibrosis, obliterative phlebitis	Not repeated	Normal pancreatic tissue
IgG4 Immunostain	>80 IgG4-positive plasma cells/HPF	Not repeated	< 10 cells/HPF

HPF: High-Power Field.

a dramatic reduction in the size of the pancreatic head mass (from 3.5 cm to 1.2 cm) and complete resolution of the biliary and pancreatic duct dilatation and the "halo sign" . The biliary stent was subsequently removed endoscopically. His CA 19-9 level normalized to 18 U/mL. Prednisone was tapered over 3 months, and the patient was started on azathioprine (2mg/kg/day) for maintenance therapy. At 6-months follow-up, he remains in clinical and radiological remission.

Discussion:

This case exemplifies one of the most critical differential diagnoses in pancreatology. The presentation with painless jaundice, a pancreatic head mass, double-duct sign, and elevated CA 19-9 has a >90% predictive value for PDAC.[3] However, several features, often overlooked initially, can point toward AIP: (1) The "Halo Sign": This is a key radiological discriminator. It represents a rim of edematous, inflamed pancreatic tissue and is highly specific for AIP, though not universally present.[4] (2) Serum IgG4: While a level >2x the upper limit of normal is highly suggestive of Type 1 AIP, it is not perfectly specific. However, in the context of supportive imaging, it is a powerful diagnostic tool.[2,5] (3) Rapid Steroid Response: The dramatic resolution of both symptoms and the mass lesion on imaging with steroid therapy is both therapeutic and serves as a confirmatory diagnostic test. The normalization of CA 19-9 is another crucial teaching point, as its elevation is common in biliary obstruction from any cause and should not be used in isolation to confirm malignancy.[6]

Conclusion:

This near-miss case underscores a vital lesson: AIP should be considered in every patient with a presumed pancreatic cancer. A systematic diagnostic approach, including a careful review of cross-sectional imaging for classic signs (e.g., halo, diffuse enlargement), measurement of serum IgG4, and procurement of an adequate core biopsy for histology, is essential. A multidisciplinary team that includes gastroenterologists, radiologists, pathologists, and surgeons is the best safeguard against performing a Whipple procedure for a steroid-responsive disease.

Patient Perspective

The patient provided written consent for this report. He expressed immense gratitude for the team's thoroughness, which spared him from a major operation. He stated he felt "like he had been given a second chance."

Author Contributions

All authors contributed to the management of the patient and the writing of the manuscript. All authors reviewed and approved the final version.

Conflict of interest

The authors declare that they have no competing interests.

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