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Case Report

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IgG4 Pancreatitis: A case report of a rare entity

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ABSTRACT

IgG4-related disease (IgG4-RD) is a condition that was first described recently and is capable of affecting any organ of the body. Diagnosis is based on the correlation of clinical findings with histopathological findings and elevated serum IgG4 [1]. We report the case of a symptomatic patient diagnosed with IgG4 Pancreatitis. IgG4 Pancreatitis is a relatively rare chronic inflammatory disease of the pancreas. Due to the lack of specific clinical symptoms and imaging findings, it is often misdiagnosed as pancreatic cancer, causing a lot of unnecessary interventions.

Keywords: IgG4, pancreatitis,autoimmune

INTRODUCTION

Autoimmune Pancreatitis (AIP) is an idiopathic chronic pancreatitis associated with hypergammaglobulinaemia, considered an autoimmune disease. Diagnosis relies on clinical, serological, and morphological assessments. Symptoms include epigastric discomfort, weight loss, obstructive jaundice, and glycuresis, often seen in chronic pancreatitis (2). Serological tests show elevated γ-globulin, IgG, IgG4, trypsin, and autoantibodies. Morphological features include acinar atrophy, fibrosis, IgG4-positive plasmacytes, and vasculopathy (3) AIP was first described in 1892, later refined by Kawaguchi in 1995 and Yoshida in 1995, with further revisions in 2009 and 2013 by the Japanese Pancreatic Society. In South Asian countries, AIP has been less studied, and diagnostic challenges persist in India due to limited access to Endoscopic ultrasound (EUS)-guided biospy. A study in India by Rana et al. (2010-2017) diagnosed 50% of patients as probable type I AIP using the ICDC 2011(International Consensus Diagnostic criteria 2011) criteria. Deshpande (2011) reviewed AIP classification in the Indian population (4,5,6,7). This case report aims to raise awareness and encourage the creation of diagnostic SOPs in India to support large-scale studies and improve understanding of this rare disease. AIP shares features with chronic pancreatitis and pancreatic cancer, making diagnosis difficult without histopathological correlation. Increased awareness will aid in more accurate AIP diagnosis.

CASE REPORT

A 34-year-old male from Ghaziabad, Uttar Pradesh, India, presented in January 2024 with acute epigastric abdominal pain radiating to the back,

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relieved by forward bending. He had recurrent vomiting (3-4 episodes daily, non-projectile) and no passage of flatus or faeces for 3 days, and no fever. He reported a similar episode a year ago, but was not evaluated properly at that time. The patient had no history of alcohol, nicotine use, trauma, radiotherapy, Endoscopic retrograde cholangiopancreaticography (ERCP). On examination, the patient was conscious and oriented, with a temperature of 98.4°F, BP 136/90 mmHg, PR 108/min, SpO2 99% on room air, and a respiratory rate of 26/min. There was no pallor, jaundice, cyanosis, lymphadenopathy, or pedal edema. Abdominal examination showed tenderness in the epigastric region with no organomegaly, and absent bowel sounds. Chest auscultation revealed decreased air entry bilaterally with basal inspiratory crepitations. Cardiovascular and CNS exams were normal. The patient was admitted and managed conservatively. On day 5, he developed respiratory distress. Chest X-ray suggested an ARDS-like picture with mild bilateral effusion. Counts and serum procalcitonin levels were increased. IV methylprednisolone(MPS) was started, and IV methylprednisolone (1mg/kg) led to significant improvement (Table 1).

Table 1: Investigations on Day One are given below

Investigation	**values**
Random blood sugar	104 mg /dL
Amylase / lipase	780/2000
Hemoglobin	11.8mg/dL
TLC	14000/dL
DLC	88/8/2/2
Platelet count	2.33lacs/ dL
T. Bil/ D. Bil	0.9/0.4
ALT/AST	16/25 IU/ml
ALP	77mg/dL
TP/SA	5.4/2.6mg/dL
Urea / creatinine	125/4.2mg/dL
Na+/K+	143/3.9meq/dL
calcium/phosphorus	7.4/3.0 mg/dL
LDH/Uric acid	477/9.5mg/dL
Procalcitonin	192mg/dL
PT/INR	23.1/1.2
D-dimer	5000ng/ml
Total cholesterol	78 mg/dL
Triglycerides	128mg/dL
LDL	33mg/dL
Chest Xray PA	Normal study
USG- whole **abdomen**	Suggestive of ill-defined hypoechoic collection of 5.6*8.6*11.5 cm with internal echoes seen in epigastrium in relation to the body of the pancreas which appears bulky and heterogeneous head of 3.6 cm with surrounding fat shows increased echogenicity, suggestive of inflammation. Other organs liver, spleen and bowels, were unremarkable.
NCCT ABDOMEN	Bulky pancreas with few hypoattenuating areas within the pancreas parenchyma, multiple collections in Peripancreatic, Gastro splenic region, left para colic gutter and mesentery with inflammatory changes, suggestive of acute necrotising pancreatitis with Peripancreatic collection.

MRCP and serum IgG4 levels were done as no clear predisposing causes were found.Steroids were switched to oral prednisolone later on, tapering over 2 weeks and stopped on Day 18 (Figure1).
The patient was discharged asymptomatic, with no fever, breathlessness, or tachycardia. Pre-steroid serum IgG4 levels were elevated at 2084 mg/dL. Follow-up after 6 months showed the patient remained asymptomatic, with repeat IgG4 levels at 1598 mg/dL. He was advised to undergo an endoscopic ultrasound-guided biopsy, the patient declined due to socioeconomic factors. Based on clinical features, MRCP, and IgG4 levels, the patient was most likely diagnosed with IgG4 pancreatitis (Table 2).

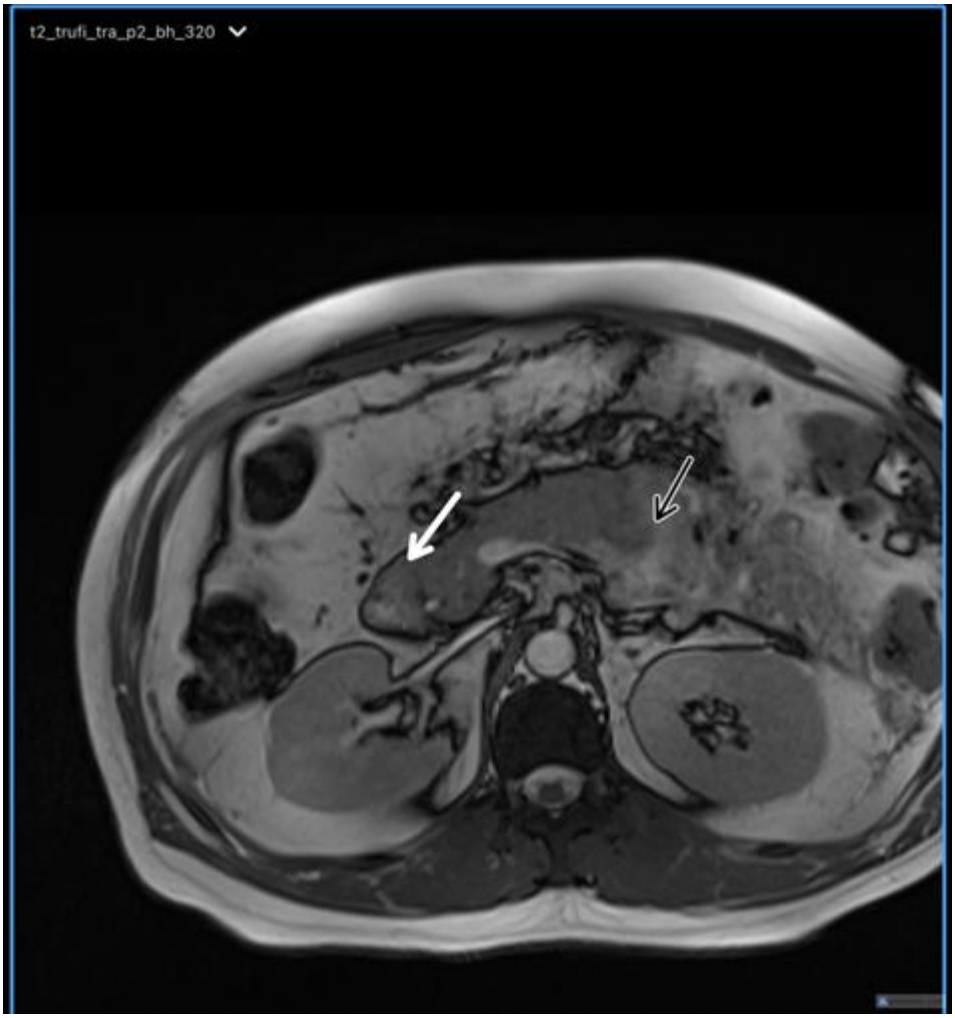

Figure 1: MRCP showing bulky head (white arrow), body and tail of pancreas with well-defined irregular walled peripheral enhancing fluid collection extending into lesser sac and retropancreatic region and peripancreatic edema (black arrow)

Table 2: Blood Autoimmune panel

Blood cultures	**No growth**
Antinuclear antibody	Negative
c ANCA, p ANCA	Negative
IgG4 (Ist value)	2084 mg/dL
IgG4 (follow-up value after steroids)	1598 mg/dL

DISCUSSION

In our case, MRCP, CT abdomen, clinical profile, and IgG4 suggested Autoimmune Pancreatitis (AIP), but the features did not align with typical AIP types 1 or 2. Many groups, including the Valona Korean and Japanese societies, have proposed diagnostic criteria for AIP, with the 2011 ICC criteria being the most widely accepted. The ICDC 2011 was developed after reviewing criteria from JPS (2002, 2006), HISORT (2006, 2009), Korean (2007), Asian (2008), Manheim (2009), and Italian (2003, 2009). Using ICDC 2011, we observed steroid response, increased IgG4, and focal pancreatic enlargement with delayed enhancement, suggestive of AIP but not confirmatory. Many Indian authors highlight the diagnostic challenges of AIP, requiring specialised tests like EUS-guided biopsy and histopathology stains (8).

Japanese diagnostic criteria for AIP (2006) include:

1. Imaging findings of diffuse or segmental pancreatic duct narrowing and enlargement.

2. Elevated serum γ-globulin, IgG, IgG4, or autoantibodies.

3. Marked fibrosis and lymphocyte/plasma cell infiltration in the pancreas.

AIP diagnosis requires criterion 1 along with criteria 2 and/or 3, while excluding malignancies like pancreatic cancer.

The International Consensus Diagnostic Criteria 2011 aimed to create a global diagnostic standard to minimise misdiagnosis, incorporating features like imaging findings, serological markers, organ involvement, histopathology, and steroid response(5). Because pancreatic histology often is not available, the terms type 1 and type 2 AIP have been introduced to describe the clinical profiles associated with lymphoplasmacytic sclerosing pancreatitis (LPSP) and idiopathic duct centric pancreatitis (IDCP), respectively, while recognising the similarities between the 2 entities. Lymphoplasmacytic sclerosing pancreatitis (LPSP) or AIP without granulocyte epithelial lesions (GELs) or AIP -1, the pancreatic histopathology shows 4 characteristic features: (1) dense infiltration of plasma cells and lymphocytes, particularly periductal; (2) peculiar storiform fibrosis; (3) venulitis with lymphocytes and plasma cells often leading to obliteration of the affected veins; and (4) abundant (9-10 cells per high-power field [HPF]) immunoglobulin (IgG4 positive plasma cells. Clinically, this form of AIP seems to be the pancreatic manifestation of an IgG4-related systemic disease characterised by elevated serum IgG4 levels and extra-pancreatic lesions (eg, sclerosing cholangitis, sclerosing sialadenitis, and retroperitoneal fibrosis) associated with infiltration with abundant IgG4-positive plasma cells. This form of AIP presents predominantly with obstructive jaundice in elderly male subjects, and the pancreatic and extra-pancreatic manifestations respond to steroid therapy. Idiopathic duct-centric pancreatitis or AIP with GELs or AIP-2 seems not to be a systemic disease; rather, it seems to be a pancreas-specific disorder. It is not associated with either serum IgG4 elevation or with other organ involvement (OOI) typically seen in LPSP. Approximately 30% of reported cases of IDCP have associated inflammatory bowel disease, such as ulcerative colitis. Patients with IDCP are, on average, a decade younger than LPSP patients and do not show a sex predilection. Currently, IDCP lacks a serological biomarker. Because IDCP patients are seronegative and lack other organ involvement, definitive diagnosis requires pancreatic histology. The consensus opinion was that the terms type 1 and type 2 should be used to describe the clinical profiles associated with LPSP & IDCP, respectively (10).

A recent global survey of AIP showed that most Asian cases fit the LPSP profile (AIP-1), while European and American cases had both LPSP and IDCP (AIP-2). IDCP diagnosis requires adequate pancreatic biopsy specimens, which are not always available, explaining fewer diagnoses worldwide. AIP is rare, with limited data from countries like India. However, Indian cases show similar clinical features, imaging, and treatment outcomes as reported globally. Our patient meets all diagnostic criteria, fitting the probable type I AIP according to ICDC 2011, with parenchymal imaging and serology evidence.[8]. Patients with AIP should be educated about treatment options, medication adherence, and the risks of inadequate treatment. Steroids are the primary treatment, with initial higher doses for 2-4 weeks, followed by tapering. Relapses are treated with the same regimen. Patients should be informed about potential side effects of long-term steroid use, including osteoporosis, weight gain, and hyperglycemia (9).

CONCLUSION

AIP can be challenging to diagnose due to nonspecific symptoms. After ruling out pancreatic cancer, patients need ongoing follow-up to assess their response to treatment. Coordinating care in both inpatient and outpatient settings is essential to improve outcomes and quality of life.

Author declaration

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The data of the patient is available with the first author.

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