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TYPE 2 AUTOIMMUNE PANCREATITIS

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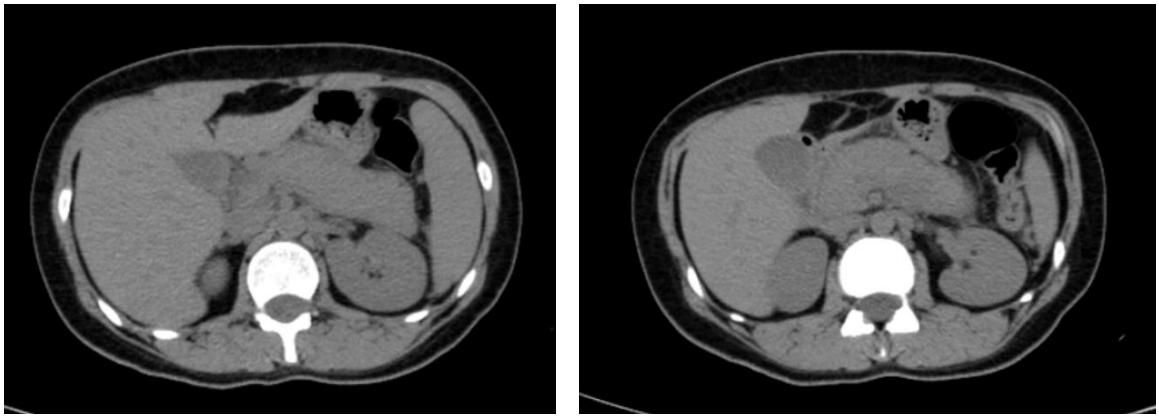
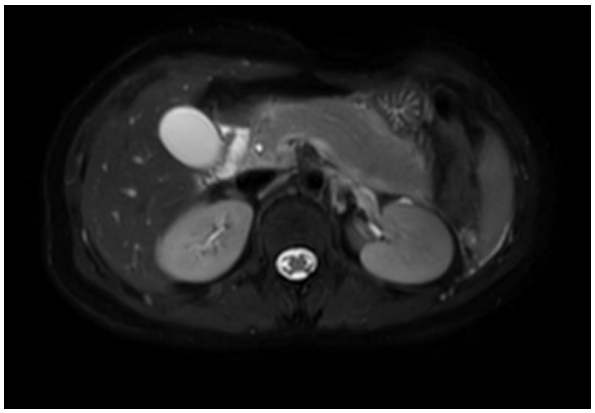
A. CLINICAL HISTORY: A 23-year-old female patient with no previous co-morbidities came to our hospital with a history of back pain for 4 days. No history of nausea/ vomiting/ loose stools/ fever/ melena/ hematemesis/ jaundice/ethanol abuse. The patient was not on any alternative medication. Physical examination was within normal limits. Serum amylase and lipase were elevated , measured 152U/L and 780 U/L respectively. AST/ ALT/ ALP were elevated , measured 88/139/308 respectively. Rest of the liver parameters and blood investigations were within normal limits.

B. IMAGING:

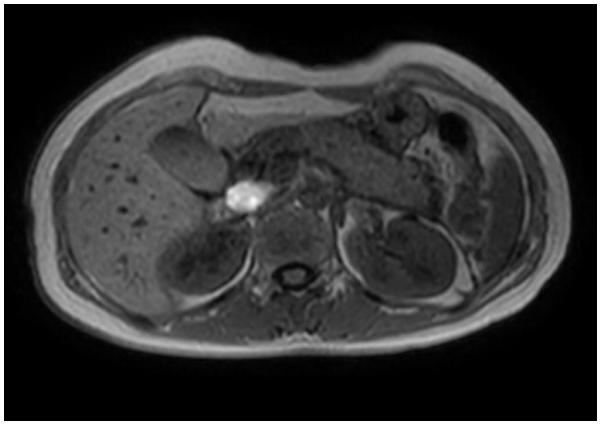
PLAIN CT ABDOMEN (Fig 1 and 2): Pancreatic parenchyma appears diffusely bulky in size and sausage-shaped with minimal peri-pancreatic fat stranding.



MR ABDOMEN T2W-SPAIR (Fig 3): Diffuse parenchymal enlargement of the pancreas with loss of feathery outline resulting in 'sausage-shaped pancreas'. Minimal peri-pancreatic fat stranding noted. The pancreatic duct was normal in calibre.



MR ABDOMEN T1W (Fig 4): Bulky pancreas with hypointense rim completely encasing the pancreas resulting in 'peripancreatic halo'. It is thought to represent inflammatory cell infiltration.



C. **DIAGNOSIS:** Diffusely bulky and sausage-shaped pancreas with minimal peripancreatic inflammatory changes and peripancreatic halo - s/o Acute edematous pancreatitis - Features are in keeping with autoimmune pancreatitis. *Needs HPE/ IG4 level correlation.

D. **DISCUSSION:**

Introduction and Epidemiology:

- Autoimmune pancreatitis is an autoimmune fibro-inflammatory systemic disorder, a rare form of chronic pancreatitis with involvement of several other organ systems, including the bile ducts, the kidneys, the retroperitoneum and the salivary glands.
- IgG4-related autoimmune pancreatitis is seen in an estimated 2%–8% of patients with chronic pancreatitis.
- Middle-aged and elderly men are commonly affected, with 95% of patients older than 45 years.
- Male-to-female ratio of 3–7:1.

Clinical features and Pathogenesis:

- Patients presents with little or no abdominal pain, obstructive jaundice, weight loss, new onset of diabetes, pancreatic enlargement, or accompanying extra pancreatic lesions.
- The predominant histologic feature of autoimmune pancreatitis is infiltration of IgG4-positive lymphocytes into pancreatic or extra-pancreatic tissue.

Diagnostic criteria:

HISORt criteria for diagnosis of AIP-SC

H Bile duct	Lymphoplasmacytic sclerosing cholangitis on resection: LP infiltrate, > **10 IgG4 + cells/hpf**, storiform fibrosis, phlebitis
I Bile duct	One or more strictures involving IH, EH, or intrapancreatic BD Fleeting/migrating biliary strictures
S	IgG4 **> 2 ULN** value
O	Pancreas: Classic features of AIP on imaging or histology Suggestive imaging findings: mass, stricture, atrophy Retroperitoneal fibrosis Renal: single/multiple parenchymal low-attenuation lesions Salivary/lacrimal gland enlargement
Rt	Normalization of liver enzyme or resolution of BD stricture
Definitive dg	**Group A**: diagnostic histology on resection or TCB **Group B**: typical imaging of AIP + serology
Probable dg	**Group C**: ≥ 2 of suggestive pancreatic imaging, S, OOI & Rt

Ghazale A et al. Gastroenterology 2008 ;134 :706 – 715.

Subtypes:

- Two separate subtypes have been identified:

  - Type 1 AIP (lymphoplasmacytic sclerosing pancreatitis) - a pancreatic manifestation of a multiorgan disease, named immunoglobulin G4 (IgG4)-related disease.

  - Type 2 AIP (idiopathic duct-centric chronic pancreatitis) - pancreas-specific disorder not associated with IgG4.

General features

- There are two main recognized patterns of autoimmune pancreatitis: **Diffuse and focal**.

- **Diffuse disease** is the more common pattern and is characterized by a uniformly enlarged pancreas with absence of pancreatic clefts. The pancreas has a sharp margin and loss of lobular contours, resulting in a featureless sausage-like appearance.

- **Focal disease** is characterized by enlargement of the pancreatic head or, less frequently, the pancreatic body or tail, resulting in a mass-like appearance - its radiologic appearance may be difficult to differentiate from that of pancreatic carcinoma.

Imaging

Ultrasonography is the primary modality of choice for evaluation of the pancreas and shows diffuse enlarged of pancreas with decreased echogenicity.

Contrast enhanced CT:

- Diffusely enlarged pancreas with delayed 'rim' enhancement – a highly specific finding presumed to represent a fluid collection, a phlegmon, or fibrosis.
- Diffusely irregular and attenuated pancreatic duct can be visualised. Other findings include focal pancreatic mass/enlargement; focal pancreatic duct stricture; pancreatic atrophy; pancreatic calcification.

MRI with MRCP:

- Pancreas appears mildly hyperintense at T2W images and hypointense at T1W images with 'hypointense rim' completely encasing the pancreas.
- The apparent diffusion coefficient value of autoimmune pancreatitis is significantly lower than that of pancreatic cancer because of the greater cellularity of the former condition.

Other extra pancreatic disorders associated with IG4:

- CNS:
  - Hypertrophic pachymeningitis
  - Autoimmune hypophysitis
  - Orbital pseudotumor
- Salivary / lacrimal glands:
  - Mikulicz's disease
  - Kuttner's tumor
- Thyroid
  - Riedel's thyroiditis
  - Hashimoto's thyroiditis
- Lungs
  - Pulmonary pseudotumor
  - Interstitial pneumoniaLymphadenopathy
- Hepatobiliary:
  - Autoimmune pancreatitis
  - Sclerosing cholangitis
  - Liver pseudotumor
- Retroperitoneal fibrosis
- Tubulointerstitial nephritis
- Inflammatory aortic aneurysm
- Prostatitis, testicular involvement

Conclusion and follow up.

- IgG4 related disease is a rare recently established systemic disorder where pancreas is most commonly involved organ among others.
- In this present case, the imaging findings suggested the possibility of an autoimmune pancreatitis. IgG4 titres was within normal limits in this patient and type 1 AIP was ruled out. Patient was treated with steroids and responded well on follow up.
- Radiologists must be familiar with this disorder because patients usually demonstrate a marked response to corticosteroid therapy, and timely recognition may help avoid delays in diagnosis and unnecessary invasive procedures.

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3. Grainger & Allison's Diagnostic Radiology - 7th Edition.