Imaging Manifestations of Autoimmune Pancreatitis

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BACKGROUND

- Autoimmune pancreatitis (AIP) was first described as a pancreatitis of autoimmune aetiology in 1995 by Yoshida et al.¹
- AIP may be clinically and radiologically difficult to distinguish from ductal pancreatic adenocarcinoma and lymphoma, and it is therefore important to consider it as possible differential in order to facilitate appropriate management.
- AIP It is a rare disorder with an estimated prevalence of 0.82 per 100,000 with a 2.85:1 male:female ratio.²
- Below is a brief overview of the clinical features and imaging findings in AIP. More detailed descriptions with further cases can be seen on the EPOS system.

CLINICAL FEATURES

More recently, AIP has been sub-divided into Type 1 and Type 2:

	Type 1 AIP	Type 2 AIP
Age	Older (6 th decade)	Young (4 th decade)
Gender	Male predominance	Male = Female
Clinical Presentation	Obstructive Jaundice	Obstructive Jaundice Abdominal Pain/Acute pancreatitis
Serum IgG4	Usually elevated	Normal
Histology	IgG-4 rich periductal lymphoplasmocytic infiltrates	Granulocyte epithelial lesions (GEL)
Extra-pancreatic Involvement	Proximal bile duct, salivary gland, kidney, retroperitoneum	None
Associated with Ulcerative Colitis	Occasional	Common
Steroid Response	Excellent	Excellent
Recurrence	Common	Rare

IMAGING FINDINGS

We present the main pancreatic and abdominal extra-pancreatic findings of AIP on cross-sectional imaging.

PANCREATIC

The morphological appearance of Type 1 and 2 AIP are radiologically indistinguishable.

AIP may either present as a diffuse form or a more focal form:

- Diffuse Form:** Smooth, sausage-like enlargement of the pancreas with loss of the normal pancreatic lobulations.
- Focal Form:** Focal enlargement, typically within the pancreatic head or uncinate process.

In both forms, there is an absence of inflammation, pseudocysts, peri-pancreatic collections and pancreatic calcification and the lack of these features can help to differentiate AIP from chronic pancreatitis.

CECT showing example of diffuse AIP with sausage shaped configuration to pancreas (arrows).

Only the extrapancreatic soft tissue shows high grade FDG uptake (arrows), atypical for lymphoma.

Patient with history of MALT lymphoma presenting with biopsy confirmed AIP. Axial T1FS postcontrast arterial phase initially shows reduced enhancement of the peripancreatic tissue (*) compared to the pancreas (arrows).

Axial T1FS postcontrast equilibrium phase showing delayed enhancement of the peripancreatic tissue. The delayed enhancement is a typical finding in AIP.

CONCLUSION

- AIP is an increasingly recognised entity, with appearances that may mimic other conditions such as pancreatic cancer and lymphoma.
- AIP readily responds to corticosteroid therapy, and therefore it is essential to correctly distinguish it from entities with similar appearances.

As stated in the table, extra-pancreatic findings of AIP are exclusive to Type 1 disease, although Type 2 is notably associated with IBD.

BILIARY

Biliary involvement is seen in the majority (up to 90%) of patients with AIP, presenting in the form of IgG-4 cholangitis. This is characterized by:

- Thickening and stricturing of bile ducts
- Strictures are usually long and smooth, with upstream dilatation.
- On imaging (CT & MRI), the affected bile duct walls demonstrate hyperenhancement post-contrast

Example of IgG4 biliary stricture: Coronal True FISP shows a tight stricture (arrow) in the distal CBD with upstream dilatation. The pancreas is also diffusely enlarged and sausage shaped (*).

Example of IgG4 related biliary stricture involving the common hepatic duct and distal CBD (arrows).

RENAL

Renal involvement is often seen with AIP, with parenchymal involvement seen in around 30% of patients. Common manifestations include:

- Generalised renal enlargement.
- Low density parenchymal lesions (either wedge-shaped or rounded).
- Development of a perirenal soft tissue rind.

CECT shows typical wedge shaped low density renal cortical lesions in the left kidney (arrows). Both kidneys are stented as IgG4 retroperitoneal fibrosis also.

Extrarenal IgG4 disease: Axial T2 HASTE shows perirenal soft tissue is of low T2 signal (*).

RETROPERITONEAL FIBROSIS

- AIP is also known to have an association with retroperitoneal fibrosis (estimated in 10% of patients).
- This presents as a retroperitoneal mass which encases the aorta and may cause hydronephrosis secondary to extrinsic compression of the ureters.

IgG4 retroperitoneal fibrosis: Axial CECT showing enhancing soft tissue partly encasing the IVC and aorta (arrows).

IgG4 retroperitoneal fibrosis: Axial FDG-PET/CT showing increased FDG uptake in the retroperitoneum (arrows).

References:
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2. Nishimori I, Tamakoshi A, Otsuki M. Prevalence of autoimmune pancreatitis in Japan from a nationwide survey in 2002. Research Committee on Intractable Diseases of the Pancreas, Ministry of Health, Labour, and Welfare of Japan. 2007 May;42 Suppl 18:6-8.