Mass forming Autoimmune Pancreatitis (AIP) mimicking a pancreatic head lesion

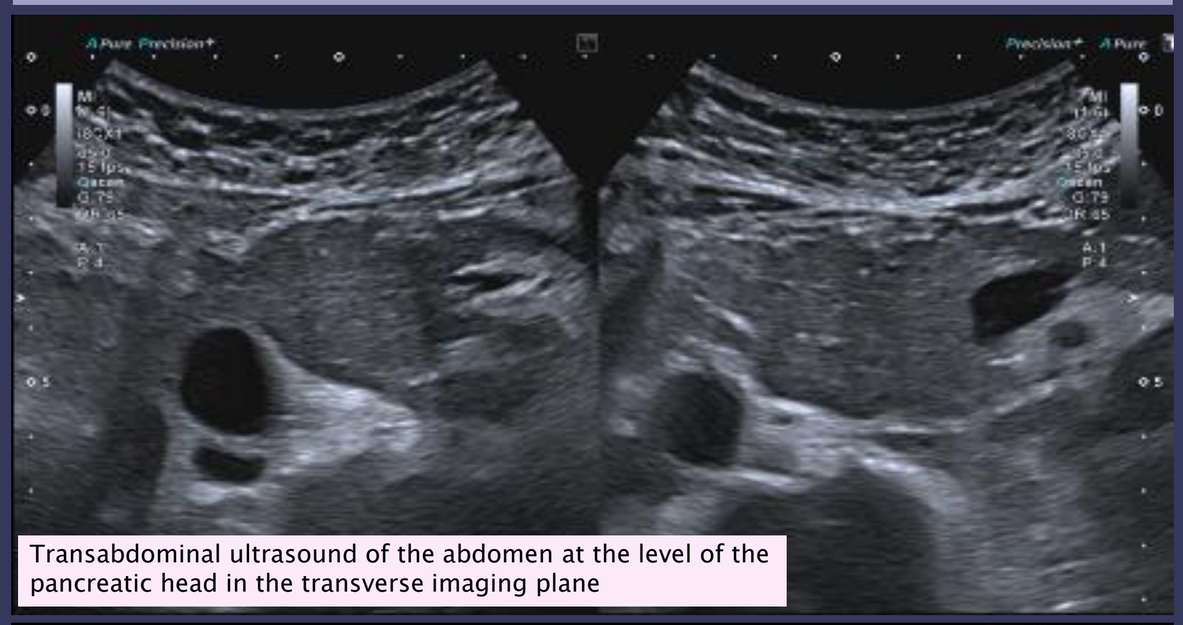
Siobhan Alsop
Ultrasound Department, Derriford Hospital, University Hospitals Plymouth NHS Trust

Background:

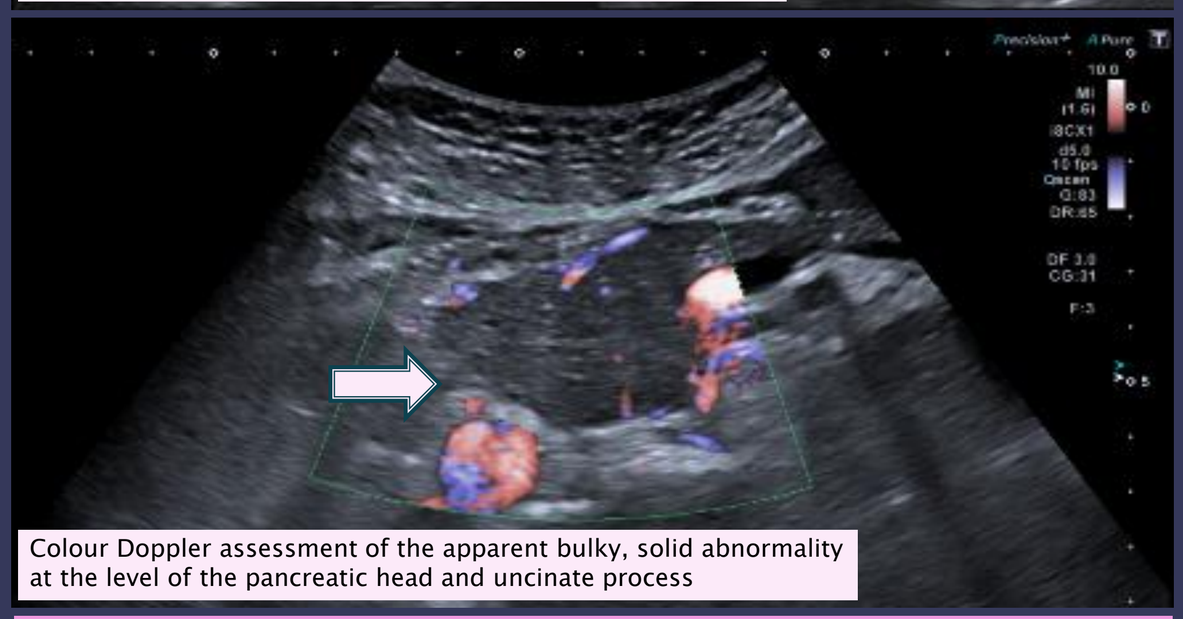
A 57-year-old female was referred for abdominal ultrasound with fatigue, jaundice on examination, deranged LFTs and a family history of haemochromatosis. The patient was not experiencing pain and there was no tenderness on assessment

1. Abdominal Ultrasound:

- Solid pancreatic head mass, isoechoic to the pancreas
- Neoplasia suspected
- Normal liver and biliary tract, nil abnormality elsewhere



Transabdominal ultrasound of the abdomen at the level of the pancreatic head in the transverse imaging plane



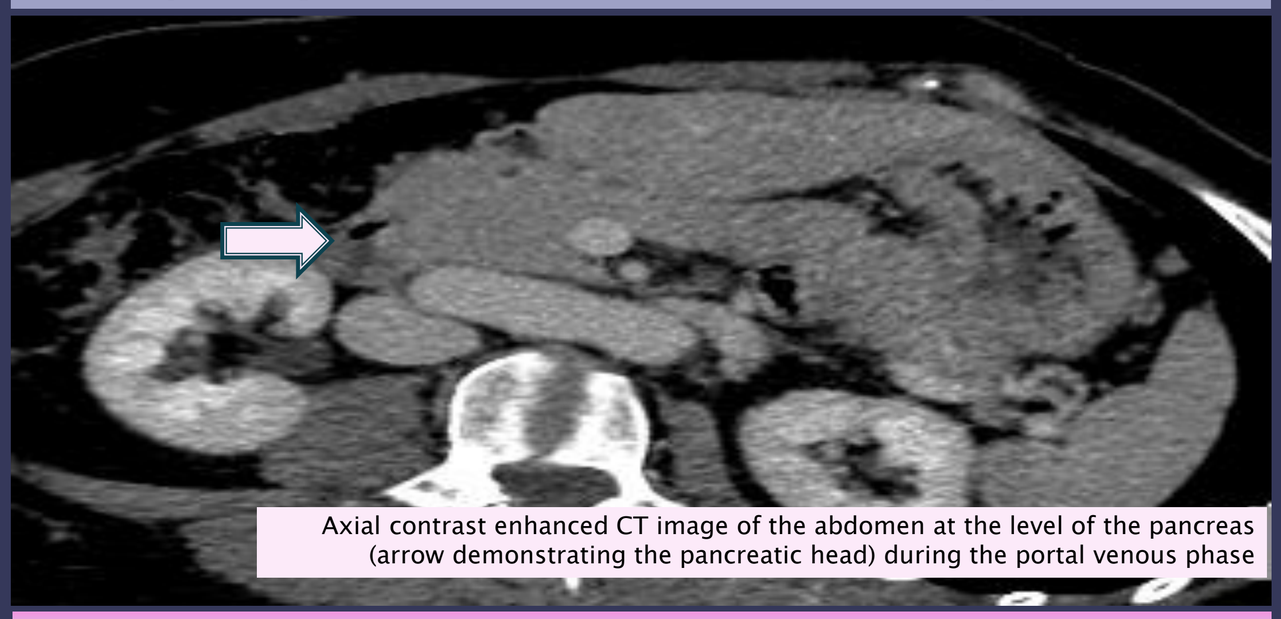
Colour Doppler assessment of the apparent bulky, solid abnormality at the level of the pancreatic head and uncinate process

2. 2WW CT Thorax, Abdomen & Pelvis:

The ultrasound findings prompted further assessment by CT staging, undertaken as a 2 week-wait referral:

- Diffusely enlarged pancreas with loss of definition of pancreatic clefts
- Subtle enhancement at biliary hilum
- Appearances suspicious of autoimmune pancreatitis/IgG4 disease

Subsequent IgG4 correlation confirmed the diagnosis of AIP



Axial contrast enhanced CT image of the abdomen at the level of the pancreas (arrow demonstrating the pancreatic head) during the portal venous phase

3. Magnetic Resonance Imaging:

MRI was performed 8 weeks after the diagnosis of AIP to review the patient's response to steroidal treatment

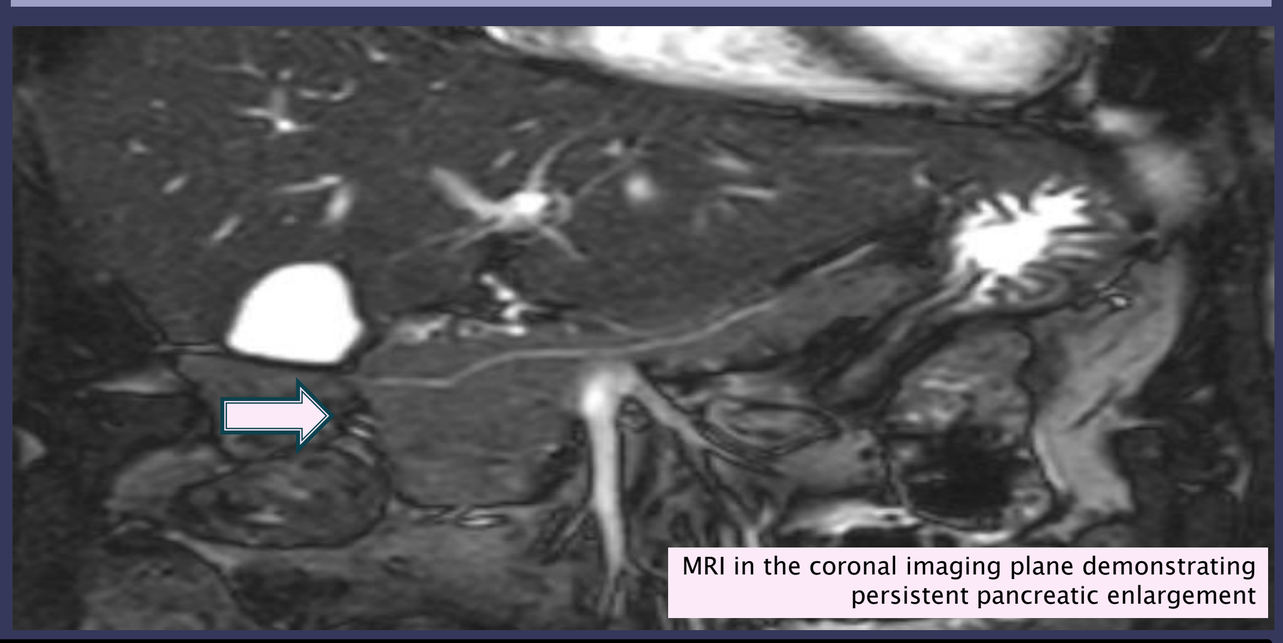
- Persistent pancreatic enlargement with loss of normal clefts
- No restricted diffusion, no inflammatory change
- Normal liver & bile ducts

4. Further Care & Management:

- LFT derangement (GGT, ALT and Bilirubin) and jaundice resolved
- Persistent IgG 4 elevation: 1.52 (normal range: 0.01-1.3) & abnormal MRI following steroidal treatment
- Ongoing patient fatigue but otherwise well
- For immunosuppressant control (Azathioprine)

****Immunosuppressant control risks suppression of bone marrow and requires full haematological monitoring in early treatment****

Further CT imaging demonstrated reduced pancreatic volume three months post Azathioprine treatment. The patient had persistent IgG4 elevation and will therefore undergo annual imaging surveillance



MRI in the coronal imaging plane demonstrating persistent pancreatic enlargement

Pathophysiology & Learning Points

- **AIP is an organ-specific manifestation of systemic immunoglobulin G4 (IgG4) related disease, categorised by marked serum IgG4 antibody elevation[1]**
- **There can be multiple organ involvement: affecting the pancreas, bile ducts, lungs, salivary glands and kidneys[1]. Like pancreatic or biliary cancer, AIP often presents with painless jaundice. AIP is a great mimicker of many neoplastic, inflammatory and infectious conditions[2]**
- **IgG4 related disease is highly treatable and responds promptly to glucocorticoids, but can lead to end stage organ failure, and even death, if unrecognised[2] and so it is important that all diagnosticians are aware of this complex disease**
- **Pancreatitis does not always present with epigastric pain or tenderness on scanning. The patient may present with symptoms of cancer, such as fatigue or jaundice**
- **Sonographic appearances of mass forming AIP may mimic that of a malignant pancreatic head mass[3] . If the mass is non-obstructive and isoechoic to the pancreas, it is more likely to represent an inflammatory mass[3] however cross-sectional imaging and biochemical correlation is always required for characterisation**
- **Understanding AIP and mass forming manifestations of pancreatitis will aid in structuring ultrasound reports to formulate appropriate differential diagnoses, and guide clinicians in further management**

References:

1. Hara A, Watanabe T, Minaga K, Yoshikawa T, Kamata K, Kudo M. Biomarkers in autoimmune pancreatitis and immunoglobulin G4-related disease. *World Journal of Gastroenterology*. 2021;27(19):2257-2269. doi:10.3748/wjg.v27.i19.2257
2. *Lanzillotta M, Mancuso G, Della-Torre E.* Advances in the diagnosis and management of IgG4 related disease. *British Medical Journal. 2020; 369; m1067. doi: 10.1136/bmj.m1067*
3. Masood, M. Autoimmune pancreatitis: what we know so far. *Journal of Gastroenterology and Hepatology Open*. 2021 Dec 10;6(1):3-10. doi: 10.1002/jgh3.12688. PMID: 35071782; PMCID: PMC8762623.