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# AIP And AIC in a 73-Year-Old Man: A Rare Case Report

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**Abstract**

A 73-year-old man presented with epigastric pain and jaundice with a prior history of large vessel vasculitis. Elevated liver enzymes were seen on the standard blood sample. Ultrasound showed a dilated common bile duct, which required further imaging. CT scan and MRI a day later both showed an enlarged pancreatic body and tail without dilation of the pancreatic Wirsung duct and cholangitis, suggestive for auto-immune pancreatitis (AIP) with auto-immune cholangitis (AIC). The radiologist suggested the diagnosis and further steps were taken. The HISORt criteria aren't a textbook example, but AIP with AIC is likely. Steroid therapy was initiated.

**Keywords:** Auto-immune pancreatitis; Auto-immune cholangitis; Common bile duct; HISORt criteria; IgG4

## Introduction

Auto-immune pancreatitis is a rather rare disease with a prevalence of 4.6 per 100.000 and a yearly incidence of 1.4 per 100.000 [1]. The HISORt criteria are introduced by Suresh T. Chari from the Mayo Clinic and are essential to correctly diagnose AIP. Based on these new diagnostic criteria, the diagnosis AIP is made more often nowadays. The acronym HISORt resembles histology, imaging, serology, other organs and response to treatment (Table 1) [2,3]. Based on these criteria, auto-immune pancreatitis can be divided in two separate types with an approximately equal distribution in Europe and the USA. Type 1 is related to a systemic disease referred to as an IgG4-related disease while type 2 is an isolated type 2 pancreatic disorder [4]. Imaging cannot differentiate these two types; however, serology and histology can.

As mentioned, AIP is a rather rare condition and therefore AIP is most often not the first working diagnosis. A set of complaints can be jaundice, new onset diabetes, weight loss and or epigastric pain, though pain is less often a primary complaint. Normally the physician will take an orientating blood sample, which mostly shows an elevated liver set. Imaging is needed to explore mainly liver and gallbladder. In case of AIP a hypo-echoic, blurry area is seen on ultrasound and on CT scan there may be an enlarged pancreas with loss of pancreatic clefts, minimal peripancreatic fat stranding, a peripancreatic hypo-attenuation rim or "halo" and pancreatic/common bile duct dilation. Sometimes other organs may also be involved [5,6].

## Case Report

A 73-year-old man presents at the emergency department with epigastric pain and jaundice. The patient has an underlying large vessel vasculitis with optical neuropathy and has 9 pack years. Clinical examination is aspecific and a blood sample is taken which shows elevated liver enzymes (Table 2). An abdominal ultrasound is performed. The abdominal ultrasound shows dilated intrahepatic ducts as well as a dilated common bile duct (7 mm). The gallbladder is thickened; there is little sludge but absence of cholelithiasis. The pancreatic ducts appear normal, but the pancreatic body and tail are pronounced and hypo echogenic. Additional CT followed by MRI is performed.

CT abdomen shows an enlarged pancreatic body and tail without significant enhancement and a normal diameter of the pancreatic ducts. The common bile duct significantly enhances, is dilated and shows no obstructive pathology (no tumour nor cholelithiasis). These findings are most likely due to auto-immune pancreatitis (AIP) with associated auto-immune cholangitis (AIC) rather than primary sclerosing cholangitis (Figure 1). Further blood investigation and MRI are necessary to differentiate. MRI investigation confirms CT-graphic findings and is strongly suggestive for AIP with AIC rather than PSC (Figure 2).

To further confirm the proposed diagnosis of AIP with AIC, the HISORt criteria are used. Biochemical analysis confirms the previously elevated liver enzymes and shows slightly though not significantly elevated IgG4 level (Table 2). The ratio IgG4/IgG1 is larger than 0.24, which is more frequent in AIC than PSC. Blood results tend towards AIP with AIC but are inconclusive. An additional echo-endoscopic examination with associated biopsy is performed. Results show inflammation of pancreatic parenchyma but are inconclusive. There is little IgG4 positivity.

| | |
|---|---|
| Criterion H-Histology (at least one of the following) | Periductal lymphoplasmacytic infiltrate, obliterative phlebitis, storiform fibrosis |
| | Lymphoplasmacytic infiltrate, storiform fibrosis, abundant IgG4+cells (≥10 HPF) |
| Criterion I-Imaging of pancreas | Typical-diffusely enlarged gland with delayed (rim) enhancemnet; diffusely irregular, attenuated main pancreatic duct |
| | Others-Focal pancreatic mass/enlargement; focal pancreatic duct structure; pancreatic atrophy; pancreatic calcification; pancreatitis |
| Criterion S-Serology | Elevated serum IgG4 (normal: 8-140 mg/DL) |
| Criterion O-Other organ involvement (can be confirmed by biopsy or resolution/improvement with steroid therapy) | Hilar/intrahepatic biliary structures; persistent distal biliary structure; parotid/lacrimal gland involvemnet; mediastinal lumphadenopathy; retroperitoneal fibrosis |
| Criterion R-Response to steroid therapy | Resolution or marked improvement of pancreatic/extra pancreatic manifestation with steroid therapy |
| Diagnostic of autoimmune pancreatitis when any of the following is fulfilled | Criterion H |
| | Criterion I+S |
| | Strong clinical suspicion of autoimmune pancreatitis (idiopathic pancreatic disease+Criterion S and/or O)+Criterion R |

**Table 1:** The acronym HISORt resembles histology, imaging, serology, other organs and response to treatment.

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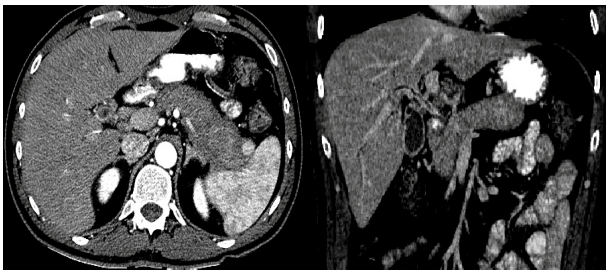

**Figure 1:** CT IV late arterial phase: Enlarged hypo-attenuating pancreas with a small capsule-like rim and a dilated common bile duct with rim enhancement and concentric thickening, No peripancreatic fluid.

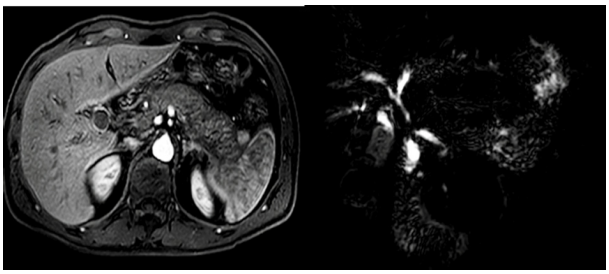

**Figure 2:** T1w late arterial phase and MRCP: Enlarged pancreas and stricture of the common bile duct.

| Blood Sample | Baseline | Day 1 | Day 6 | 2 Months | 6 Months | Reference |
|---|---|---|---|---|---|---|
| Total Bilirubin | 7 | 6.85 | 3.46 | 3.86 | 0.48 | 0.30-1.20 |
| AST | 96 | 93 | 112 | 199 | 17 | <40 |
| ALT | 208 | 196 | 202 | 337 | 13 | <41 |
| Alk Phos | 302 | 297 | 258 | 406 | 55 | 40-129 |
| GGT | 763 | 671 | 376 | 645 | 15 | 8-61 |
| Lipase | 32 | 25 | | 27 | -- | 13-60 |
| IgG4 | -- | -- | 2.54 | 3.56 | 0.69 | 0.03-2.00 |
| IgG1 | -- | -- | -- | -- | 4.52 | 4.9-11.4 |

**Table 2:** Blood results.

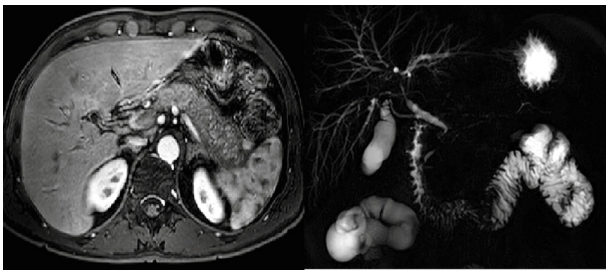

**Figure 3:** T1w dynamic late arterial phase and MRCP: Resolving pancreatic enlargement and not significantly changed common bile duct.

The HISORt criteria are used to confirm the diagnosis:

- **Histology:** Minimal IgG4 positivity, inconclusive inflammation.
- **Imaging:** Strongly suggestive for AIP with AIC. Enlarged pancreatic body and tail, no pancreatic duct dilation and dilation with enhancement of the common bile duct without obstructive pathology.

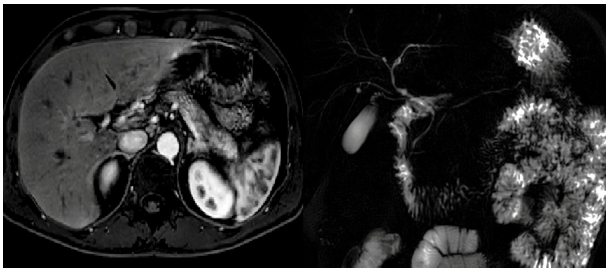

**Figure 4:** T1w dynamic late arterial phase and MRCP: Resolving pancreatic enlargement and not significantly changed common bile duct.

- **Serology:** Slightly elevated IgG4.
- **Other organs:** Associated cholangitis, presumably AIC rather than PSC.
- **Response to treatment:** Not started yet.

## Discussion

The HISORt criteria are only partially positive but are rather suggestive for AIP with AIC. AIP with AIC is withheld as a working diagnosis and treatment with Predinisolon 20 mg a day is initiated. Biochemical control is performed after 1 week and 8 weeks as well as follow up MRI investigation after 2 months. 6 months after initiation of therapy there is a normalisation of the IgG4 levels and liver parameters (Table 2).

MRI shows a significant decrease in gallbladder thickness, enlargement of the pancreatic body and tail, dilation and enhancement of the common bile duct. This is suggestive for AIP with AIC with good response to therapy (Figure 3). 7 months later another MRI is performed which shows further MR-graphic improvement (Figure 4).

## Conclusion

Auto-immune pancreatitis is a rare and rather unknown disease. Radiologists should be familiar with the diagnosis since they have a key role in the diagnostic process, as shown in this case. Often, they are the first to think about AIP, but they should also differentiate with a pancreatic ductal adenocarcinoma. On top of the diagnosis, they have to know which additional exams are required to confirm and differentiate and should be aware of the existence of the HISORt criteria. This may lead to a faster diagnosis and correct treatment. Furthermore, they may mention that further examination is required, e.g. additional MRCP and blood sample with titration of IgG4.

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