Autoimmune Pancreatitis Presenting with Jaundice

Salil Garg, Harvard Medical School, Year III
Gillian Lieberman, M.D.
Our patient: A 73 yo male presenting with obstructive jaundice, history of autoimmune pancreatitis
Autoimmune Pancreatitis (AIP)

• Relatively rare, representing 5%-11% of chronic pancreatitis
• Twice as common in men as women
• Wide variance in age, most cases > 50 yo
• Most common presentation is jaundice or abdominal pain. Acute pancreatitis presentation is rare.
Criteria for AIP Diagnosis

- **No gold standard, HISORT used most frequently in United States**
- **Histology**
  - Periductal infiltrate, T lymphocytes and plasma cells (IgG4+)
- **Imaging**
  - Diffuse pancreatic enlargement, or focal mass
- **Serology**
  - Hypergammaglobulinemia, Highly elevated IgG4 is specific but not sensitive
- **Other organ involvement**
  - Gallbladder, bile ducts, kidney, lung, salivary glands
- **Response to Steroid Treatment**
  - Glucocorticoids (prednisolone)
Causes of Pancreatitis

- Acute
  - Alcohol
  - Gallstones
  - Metabolic
  - Drugs
  - Infection

- Chronic
  - Alcohol
  - Cigarette smoking
  - Hereditary/Congenital
  - Obstruction
  - Tropical pancreatitis
  - Idiopathic

- Autoimmune
Companion Patient I: Usual Signs of Pancreatitis on CT....

Diffusely enlarged pancreas often with irregular borders
Peripancreatic inflammation
Fat stranding
Heterogenously enhancing parenchyma
Necrosis, Abscess

In contrast, autoimmune pancreatitis is “dry” and often lacks fat stranding. Borders are regular.
Our patient’s 1st presentation: “Dry” Autoimmune Pancreatitis

Our patient shows diffuse pancreatic enlargement, most easily visualized here in the head of the pancreas. Pancreatic borders are regular and little to no fat stranding is apparent when compared to other forms of pancreatitis.
Biliary Tree Anatomy

http://www.hopkins-gi.org
Our patient, 1\textsuperscript{st} presentation: Endoscopic Retrograde Cholangiopancreatography (ERCP) reveals \textit{stricture} of the distal common bile duct with \textit{proximal dilatation}.
Our patient, 1\textsuperscript{st} presentation: Guidewires during ERCP allowed placement of a stent across the \textit{stricture}

Patient’s jaundice resolved, discharged on glucocorticoids
Our patient, 2\textsuperscript{nd} presentation: Two years later our patient again presents with obstructive jaundice, now with biliuria.

Pancreatic head is smaller but a dilated pancreatic duct is visible.
Our patient, 2\textsuperscript{nd} presentation: ERCP revealed new proximal biliary strictures

\textbf{Stricture of the left hepatic duct…}

…leading to \textbf{dilation} of the left hepatics
Our patient, 2nd presentation: The old stricture in the common bile duct which was previously stented has resolved
Our patient, follow up to second presentation: Stenting effectively removed stricture in left hepatic duct and cleared jaundice.

**Stricture** of left hepatic duct at second presentation

**Resolution** of stricture after stent removal two months later
Our patient, 3rd presentation: A year later, patient presented a third time with obstructive jaundice.

Stricture of common hepatic duct with marked dilation of proximal hepatics.

ERCP confirms stricture of the common hepatic duct with proximal dilation.
Our patient: Summary of clinical course

- Presented with obstructive jaundice due to common bile duct stricture.
- Stricture and jaundice resolved with ERCP placement of a stent.
- Strictures recurred at the left hepatic duct (2\textsuperscript{nd} presentation) and common hepatic duct (3\textsuperscript{rd} presentation).
- These were also resolved with ERCP stent placement.
What are other pathologies to worry about in this patient?

- Pancreatic Adenocarcinoma
- Primary Sclerosing Cholangitis
- Other autoimmune diseases: rheumatoid arthritis, Sjogren’s syndrome, inflammatory bowel disease
- Lymphocytic infiltrates in other organ systems (Lung, Kidney, salivary glands, soft tissues near Aorta)
- **Cholangiocarcinoma**
Testing for Malignancy:
Scrapings of all of the above biliary strictures were taken during ERCP and sent for cytology……

…luckily for our patient, cytological studies of bile duct cells were normal. What might malignant transformation look like?
Benign Bile Duct Cells

“Honeycomb” organization
Round, regular nuclei
Relatively high amount of cytoplasm
Reactive Bile Duct Cells

“Honeycomb” organization
Round, regular nuclei
Relatively low amount of cytoplasm
Malignant Ductal Cells

Single sheet organization lost, “3-D” like appearance

Irregular enlarged nuclei, with marked hyperchromatism

Very little cytoplasm
Recent progress in understanding the pathophysiology of Autoimmune Pancreatitis…

A monoclonal antibody which recognizes plasminogen binding protein from *Heliobacter pylori* is found specifically in patients with AIP.

A very similar peptide is extensively expressed on pancreatic acinar cells!!
Summary

• Autoimmune pancreatitis is a relatively rare but important cause of pancreatitis

• The most common presenting symptom is jaundice

• Radiological appearance is of a “dry” pancreatitis, often with pancreatic enlargement. Constriction or dilatation of the pancreatic duct is also possible.

• Treatment (Steroids, Interventional Radiology) is efficacious in most patients though not curative as symptoms (such as strictures) can reoccur

• Important to distinguish AIP from pancreatic cancer, cholangiocarcinoma, and primary sclerosing cholangitis and to monitor for these complications

• Progress is being made in understanding the etiology of this disease
References


Acknowledgements

• Dr. Gillian Lieberman, Course Director
• Dr. Jean-Marc Gauguet, Radiology
• Dr. Robert Najarian, GI-Pathology
• Larry Barbaras, webmaster
• Emily Hanson, coordinator