AUTOIMMUNE PANCREATITIS AND IGG-4 RELATED DISEASE

Prasad Kulkarni, MD FACG
Assoc Professor of Medicine USF
Director, GI Endoscopy
James A Haley VA Hospital, Tampa FL
Historical Background

• Initial description by Sarles in 1961
  • “A single case of pancreatitis associated with hyper-gammaglobulinemia in a series of 205 cases of acute, recurrent and chronic pancreatitis”

• Concept of AIP proposed by Yoshida in 1995

• More and more cases get reported leading to lack of unified approach to diagnosis and management

• International consensus diagnostic criteria agreed upon in 2010
Alternative Terminologies Used

- Sclerosis of the pancreas (Sarles 1961)
- Chronic sclerosing pancreatitis
- Lymphoplasmacytic sclerosing pancreatitis
- Non-alcoholic duct-destructive pancreatitis
- Sclerosing pancreatitis
- Sclerosing pancreatico-cholangitis
- Autoimmune chronic pancreatitis
- Autoimmune pancreatitis (Yoshida 1995)
AIP Consensus Definition

• AIP is a distinct form of pancreatitis characterized
  • Clinically by frequent presentation with obstructive jaundice with or without a pancreatic mass
  • Histologically by a lymphoplasmacytic infiltrate and fibrosis
  • Therapeutically by a dramatic response to steroids

• When thus defined, AIP has two distinct subtypes, Type 1 and Type 2

Pancreas 2011;40:352-358
Type 1 AIP

- Pancreatic manifestation of a multi-focal systemic disease, called IgG4-Related Disease (IgG4-RD)

- IgG4-RD is characterized by
  - Fibro-inflammatory process
  - Elevated serum IgG4 levels
  - Typical histopathology in pancreas as well as other organs
  - Tissue infiltration with IgG4 positive cells
Type 2 AIP

- Pancreas-specific disease that is not associated with elevated serum IgG4 or infiltration of tissues with IgG4 positive cells
- Presence of “granulocyte epithelial lesions (GEL)” in pancreatic ducts
- Approximately 20 to 30% patients also have IBD
# Clinical Profiles of AIP Type 1 and 2

<table>
<thead>
<tr>
<th></th>
<th>Type 1 AIP</th>
<th>Type 2 AIP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age (years), mean ± SD</td>
<td>61.8 ±14.2</td>
<td>47.7±18.8</td>
</tr>
<tr>
<td>Gender (M/F)</td>
<td>60/18</td>
<td>14/5</td>
</tr>
<tr>
<td>Imaging</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Diffuse swelling</td>
<td>40%</td>
<td>16%</td>
</tr>
<tr>
<td>Other features</td>
<td>60%</td>
<td>84%</td>
</tr>
<tr>
<td>Serum IgG4 elevation</td>
<td>80%</td>
<td>17%</td>
</tr>
<tr>
<td>Other organ involvement</td>
<td>60%</td>
<td>0</td>
</tr>
<tr>
<td>IBD</td>
<td>6%</td>
<td>16%</td>
</tr>
</tbody>
</table>

Chari et al, Gastroenterology 2010
How Common is AIP?

- Reported incidence between 2002 and 2004
  - Korea 5.4% (17/315)
  - Japan 4.6% (21/451)
  - Italy 6.0% (23/383)
- Estimated prevalence in Japan: 5 to 6% of all chronic pancreatitis
- US incidence and prevalence:
  - Unknown
  - Likely to be underdiagnosed due to lack of awareness
- In a Mayo Clinic study of 245 pathology specimens from patients that underwent resections for benign pancreatic disease, 27(11%) revealed AIP

*Yadav D. Clin Gastroenterol and Hepatol 2003; 1:129-35*
Pathogenesis of AIP

- Unclear
- Autoimmune
  - Elevated IgG4 level with lymphoplasmacytic infiltrates involving IgG4-positive plasma cells in affected organs
  - Autoantibodies against carbonic anhydrase, lactoferrin and other antigens
  - T helper Type 2 (Th2) cells and T regulatory (Tregs) cells predominate the immune reaction via Lymphotoxin (previously known as TNF-beta)
  - Strong association with other autoimmune conditions e.g Sjogren’s syndrome, PSC, IBD, SLE, retroperitoneal fibrosis, Hashimoto’s thyroiditis, etc
  - Dramatic response to steroid
IgG4-related Systemic Disease

- Pancreas
- Biliary tree
- Salivary glands
- Retro-peritoneum
- Lymph nodes
- Kidneys
- Lungs
AIP Clinical Spectrum

Autoimmune Pancreatitis

Pancreatic

- Acute
  - Obstructive jaundice
  - Pancreatic mass
  - Pancreatitis
  - Steatorrhea
  - New-onset diabetes

- Post-acute/ Late
  - Pancreatic mass
  - Steatorrhea
  - Calcification/ Pseudocyst
  - Asymptomatic

Predominantly Extra-pancreatic

- Biliary strictures
- Renal failure
- Salivary “tumor”
- Lymphoma-like lymphadenopathy
AIP-associated Sclerosing Cholangitis

- Or inflammatory pseudotumour
- Or PSC mimicking chronic pancreatitis
- Or pancreatic pseudotumour with multifocal idiopathic fibrosclerosis
- Or lymphoplasmacytic sclerosing pancreatitis with cholangitis
- Or sclerosing pancreato-cholangitis
- Or atypical PSC associated with unusual pancreatitis
- Or lymphoplasmacytic sclerosing cholangitis without pancreatitis
- Or IgG4 related lymphoplasmacytic sclerosing cholangitis
Spectrum of IgG4-associated Cholangitis

Björnsson et al., *Hepatology* 2007; 45: 1547
<table>
<thead>
<tr>
<th>Year</th>
<th>Country</th>
<th>Prevalence</th>
<th>Count</th>
<th>Total PSC</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>2004</td>
<td>Japan</td>
<td></td>
<td>28</td>
<td>388</td>
<td>7%</td>
</tr>
<tr>
<td>2006</td>
<td>USA</td>
<td></td>
<td>12</td>
<td>127</td>
<td>9%</td>
</tr>
<tr>
<td>2009</td>
<td>Canada</td>
<td></td>
<td>4</td>
<td>91</td>
<td>15%</td>
</tr>
</tbody>
</table>
Gross Pathology of AIP

- Diffusely indurated and firm pancreas on gross exam
- Majority involvement is in pancreatic head. Minority in pancreatic tail or diffusely.
- Gray to yellowish-white induration of the affected tissue with loss of its normal lobular structure.
- Focal mass can be found in a subset of patients
Gross Pathology of AIP

- Diffusely indurated and firm pancreas on gross exam
- Majority involvement in the pancreatic head. Minority in pancreatic tail or diffusely.
- Gray to yellowish-white induration of the affected tissue with loss of its normal lobular structure.
- Focal mass can be found in a subset of patients
Histopathology of AIP

Characteristic peri-ductal cuffing of AIP (H&E)

Phlebitis is a characteristic feature of AIP. An elastic stain highlights obliterative phlebitis and arteritis
Histopathology of AIP

IgG4 Immunohistochemical stain showing a diffuse infiltrate of IgG4-positive plasma cells

Peri-ductal lympho-plasmacytic infiltrate in AIP
Laboratory Findings in AIP

- Increased Gamma Globulins (37 to 76%)
- Increased IgG4 (76%)
- Other Autoimmune antibodies
  - Anti-nuclear antibodies
  - AMA
  - Rheumatoid factor
  - Anti-lactoferrin antibodies
  - Anti-smooth muscle antibody
  - Anti-carbonic anhydrase antibody
Plasminogen Binding Protein in AIP

- An association between AIP and H. pylori infection has been proposed
- PBP, a protein found in H. pylori exhibits homology with UBR2 protein in pancreatic acinar cells
- H. pylori infection, in certain individuals leads to AIP through molecular mimicry
- In a study of 20 AIP patients, 19 (95%) were found to have anti-PBP antibody as opposed to 4 of 40 (10%) of patients with pancreatic carcinoma
- Another validating study found 14 of 15 (93%) AIP patients, and 1 of 70 (1.4%) of patients with pancreatic carcinoma to be positive for anti-ABP antibodies.
- Anti-PBP may be useful as a screening test for AIP

# IgG4 in AIP

<table>
<thead>
<tr>
<th>IgG subclass</th>
<th>Normals (n=20)</th>
<th>AIP (n=20)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>IgG1</td>
<td>664 mg/dl</td>
<td>868 mg/dl</td>
<td>0.25</td>
</tr>
<tr>
<td>IgG2</td>
<td>592 mg/dl</td>
<td>617 mg/dl</td>
<td>0.99</td>
</tr>
<tr>
<td>IgG3</td>
<td>34 mg/dl</td>
<td>53 mg/dl</td>
<td>0.12</td>
</tr>
<tr>
<td>IgG4</td>
<td>51 mg/dl</td>
<td>663 mg/dl</td>
<td>0.001</td>
</tr>
</tbody>
</table>
## IgG4 in Diagnosis of AIP

<table>
<thead>
<tr>
<th></th>
<th>IgG4 Upper Limit of Normal mg/dl</th>
</tr>
</thead>
<tbody>
<tr>
<td>Test Parameters</td>
<td>140</td>
</tr>
<tr>
<td>Sensitivity</td>
<td>76%</td>
</tr>
<tr>
<td>Specificity</td>
<td>93%</td>
</tr>
<tr>
<td>PPV</td>
<td>36%</td>
</tr>
</tbody>
</table>

## IgG4 in AIP: Response to Steroids

<table>
<thead>
<tr>
<th></th>
<th>Before Steroids</th>
<th>After 4 weeks of Steroids</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total IgG</td>
<td>2389 mg/dl</td>
<td>1138 mg/dl</td>
<td>0.002</td>
</tr>
<tr>
<td>IgG4</td>
<td>742 mg/dl</td>
<td>223 mg/dl</td>
<td>0.002</td>
</tr>
</tbody>
</table>

Non-pancreatic IgG4 as Surrogate

  - Biopsy of major duodenal papilla
  - 10 AIP, 10 pancreatic cancer and 10 papillitis patients
- Immuno-staining using anti-IgG4 antibodies
- IgG4-positive plasma cells per high-power field (HPF)

<table>
<thead>
<tr>
<th>IgG4 plasma cells</th>
<th>Significant (\geq 10/\text{HPF})</th>
<th>Moderate (4-9/\text{HPF})</th>
<th>Rare (\leq 3/\text{HPF})</th>
</tr>
</thead>
<tbody>
<tr>
<td>Autoimmune Pancreatitis</td>
<td>8</td>
<td></td>
<td>2 (body/tail)</td>
</tr>
<tr>
<td>Pancreatic Cancer</td>
<td>0</td>
<td>1</td>
<td>9</td>
</tr>
<tr>
<td>Papillitis</td>
<td>0</td>
<td>0</td>
<td>10</td>
</tr>
</tbody>
</table>
CT Features of AIP

- Diffuse or focal enlargement of pancreas (Up to 80% involves head of pancreas). “Sausage-like pancreas”
- Sharp outline, homogenous initial decreased enhancement with delayed increased enhancement
- Capsule like rim (fibro-inflammatory changes involving the peri-pancreatic adipose tissue)
- Minimal peri-pancreatic stranding, enlarged peri-pancreatic nodes
- Diffuse pancreatic ductal narrowing
PET and DWMR in AIP

- FDG-PET will enhance both AIP and pancreatic cancer

- Extra-pancreatic involvement is highly suggestive of AIP

- MRCP will show delayed enhancement in AIP and long/multiple PD strictures, but otherwise inferior to ERCP

- Diffusion-weighted MR (DWMR) will show high signal intensity areas both in AIP and pancreatic cancer
  - AIP: multiple and diffuse high signal intensity areas
  - Pancreatic cancer: solitary high signal intensity areas
## AIP vs Pancreatic CA: CT Scan

<table>
<thead>
<tr>
<th>FEATURE</th>
<th>AIP (%)</th>
<th>CANCER (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diffuse decreased enhancement</td>
<td>28</td>
<td>3</td>
</tr>
<tr>
<td>Capsule-like rim</td>
<td>40</td>
<td>9</td>
</tr>
<tr>
<td>Peri-pancreatic stranding</td>
<td>60</td>
<td>27</td>
</tr>
<tr>
<td>Pancreatic calcifications</td>
<td>32</td>
<td>9</td>
</tr>
<tr>
<td>Bile duct wall enhancement</td>
<td>52</td>
<td>6</td>
</tr>
<tr>
<td>Renal involvement</td>
<td>28</td>
<td>0</td>
</tr>
<tr>
<td>Pancreatic duct dilation</td>
<td>24</td>
<td>67</td>
</tr>
<tr>
<td>Abrupt PD cut-off</td>
<td>16</td>
<td>55</td>
</tr>
</tbody>
</table>

AJR 2008; 190: 280-6
ERCP in AIP

(i) long (>1/3 the length of the pancreatic duct) stricture
(ii) lack of upstream dilatation from the stricture (<5 mm)
(iii) multiple strictures
(iv) side branches arising from a strictured segment
Effect of Steroid Therapy on AIC
ERCP in AIP: Response to Steroids

Steroids for 3 months
Mayo HISORt Criteria

- **H**  Diagnostic Histology
- **I**  Imaging (CT/ MRI or ERCP)
- **S**  Serology (elevated IgG4)
- **O**  Other organ involvement
- **Rt** Response to steroid therapy — usually a two-week steroid trial
## Comparison of Diagnostic Criteria: AIP

<table>
<thead>
<tr>
<th></th>
<th>JPS</th>
<th>Kim</th>
<th>HISORt</th>
<th>Italian</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Imaging</strong></td>
<td>* Essential</td>
<td>* Essential</td>
<td>Not essential</td>
<td>Not included</td>
</tr>
<tr>
<td><strong>Laboratory</strong></td>
<td>Elevated IgG4</td>
<td>Elevated IgG4/IgG or autoantibodies</td>
<td>Elevated IgG4</td>
<td>Not included</td>
</tr>
<tr>
<td><strong>Histopathology</strong></td>
<td>Marked lympho-plasmacytic infiltration and fibrosis</td>
<td>Lympho-Plasmacytic infiltration and fibrosis</td>
<td>*LPSP IgG4 plasma cells</td>
<td>* Histology or cytology</td>
</tr>
<tr>
<td><strong>Response to Steroid</strong></td>
<td>Not included</td>
<td>Included</td>
<td>Included</td>
<td>Included</td>
</tr>
<tr>
<td><strong>Other organ involvement</strong></td>
<td>Not included</td>
<td>Not included</td>
<td>Included</td>
<td>Included</td>
</tr>
</tbody>
</table>
EUS in AIP

Study of 14 patients, 10 of whom had subsequent surgery with resection or biopsy.

- Focal or diffusely hypoechoic pancreatic enlargement.
- Lymphadenopathy up to 3 cm common.
- Vascular involvement noted in 21% of patients. Increased ‘stiffness’ on elastography.
- EUS-FNA showed chronic inflammatory cells and no evidence of malignancy in the pancreas or lymph nodes.
- EUS and EUS guided biopsy may support a diagnosis of AIP, and allow steroid therapy and avoidance of surgery in right clinical setting.

Farrell J. Gastrointest Endosc 2004; 60: 927.
EUS-Guided Trucut Biopsy

- Levy et al, *Gastrointestinal Endoscopy* 2005
- 3 patients with suspected AIP
- Results:
  - 2 patients -- AIP
  - 1 patient -- non-specific changes of chronic pancreatitis

- Managed conservatively with close monitoring
- Avoidance of surgery
EUS-guided Trucut Biopsy vs. FNA

- Mizuno et al; Journal of Gastroenterology; 2009
- 14 patients with AIP suspected on imaging
- All underwent EUS-FNA and EUS-Trucut bxs. No complications with either
- 8/14 were diagnosed with AIP using Japanese criteria
- EUS-FNA:
  - 3/8 were reported as p-LPSP (probable LPSP)
  - 1/8 was reported as normal,
  - 4/8 were inconclusive
- EUS-TCB
  - 4/8 were reported as p-LPSP (probable LPSP)
  - 4/8 were reported as d-LPSP (definite LPSP)
EUS-Guided Trucut Biopsy

• Advantages
  • Larger biopsy specimen
  • EUS has superior resolution that can improve accuracy of lesion targeting
  • Risk of seeding (if pancreatic CA is a concern) is lower than trans-abdominal biopsy

• Drawbacks
  • Technically difficult (especially when lesion at pancreatic head) due to angulation
  • Patchy distributions of AIP may lead to false negative
<table>
<thead>
<tr>
<th>Detection Rate</th>
<th>Core Biopsy</th>
<th>Resection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bang <em>et al.</em></td>
<td>26%</td>
<td>100%</td>
</tr>
<tr>
<td><em>Pancreas</em> 2008</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zamboni <em>et al.</em></td>
<td>22%</td>
<td>90%</td>
</tr>
<tr>
<td><em>Virchows Arch.</em> 2004</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chari <em>et al.</em></td>
<td>44%</td>
<td>92%</td>
</tr>
<tr>
<td><em>Clin Gastroenterol Hepatol</em> 2006</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
AIP: Differential Diagnosis

Pancreatic Carcinoma
Alcohol Induced Pancreatitis
Primary Sclerosing Cholangitis
Primary Biliary Cirrhosis
Autoimmune Hepatitis
## AIP Vs Pancreatic Carcinoma

<table>
<thead>
<tr>
<th>Feature</th>
<th>AIP</th>
<th>Pancreatic CA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ductal strictures</td>
<td>Multiple</td>
<td>Single</td>
</tr>
<tr>
<td>Complete obstruction of main PD</td>
<td>Uncommon</td>
<td>Common</td>
</tr>
<tr>
<td>Upstream ductal dilation</td>
<td>Mild</td>
<td>Marked</td>
</tr>
<tr>
<td>Duct in mass</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Diffuse pancreatic swelling</td>
<td>Almost always</td>
<td>Rare</td>
</tr>
<tr>
<td>Double duct sign</td>
<td>Common</td>
<td>Common</td>
</tr>
</tbody>
</table>
## AIP Vs Alcoholic Chronic Pancreatitis

<table>
<thead>
<tr>
<th>Feature</th>
<th>AIP</th>
<th>ACP</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pancreatogram</td>
<td>Irregular narrowing</td>
<td>Irregular dilation</td>
</tr>
<tr>
<td>Calcifications</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Pseudocyst/ phlegmon</td>
<td>Rare</td>
<td>Common</td>
</tr>
<tr>
<td>Pancreatic parenchyma</td>
<td>Enlargement</td>
<td>Atrophic</td>
</tr>
</tbody>
</table>
Approach to Patient with Suspected AIP

Pancreatic Imaging

- Diffuse pancreatic swelling on CT
- Sclerosing pattern on ERCP

Laboratory Data

Histopathology

Trial of Steroids

Autoimmune Pancreatitis
IgG4-Associated Cholangitis

**Biliary Stricture(s):** intra-hepatic, proximal extra-hepatic duct, intra-pancreatic

- **Group A**
  - Previous pancreatic/biliary resection or core biopsy of pancreas showing diagnostic features of AIP/IAC

- **Group B**
  - Classical imaging findings of AIP + Elevated serum IgG4

- **Group C**
  - Two or more of the following:
    - Elevated serum IgG4
    - Suggestive pancreatic imaging findings (see Table 4)
    - Other organ involvement
    - Bile duct biopsy with > 10 IgG4 + ve cells/hpf

---

**Definite IAC**

- Complete 11 week course of steroids for distal bile duct strictures
- Consider adding azathioprine 2 mg/kg for maintenance of remission in proximal/intra-hepatic strictures

**Probable IAC**

- Steroid treatment for 4 weeks*

---

Repeat evaluation shows:
- Markedly improved biliary strictures allowing stent removal
- Liver enzymes < 2 ULN
- Decreasing serum IgG4 and Ca 19-9

---

*Every effort should be made to exclude biliary/pancreatic malignancy before initiating steroid therapy which should not be used as a substitute for a thorough search for malignancy

- The following features are not suggestive of IAC and should suggest alternate diagnosis:

  a) Before steroid therapy:
    - Atypical biliary cytology, positive DIA/FISH
    - Markedly elevated serum CA 19-9
    - Pancreatic duct dilation with pancreatic atrophy

  b) On steroid therapy
    - Rising serum CA 19-9
    - No improvement in liver enzymes or biliary stricture despite 4-6 weeks of therapy

---

Ghazale et al, Gastroenterology 2008
Differentiating between PSC and IgG4-Associated Cholangitis

<table>
<thead>
<tr>
<th>Feature</th>
<th>PSC</th>
<th>IgG4-associated Cholangitis</th>
<th>$p$ value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Average age (years)</td>
<td>39.2</td>
<td>63.8</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Male/Female</td>
<td>1/3</td>
<td>29/5</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Obstructive jaundice +/-</td>
<td>0/4</td>
<td>30/4</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Elevated IgG4 +/-</td>
<td>0/2</td>
<td>26/36</td>
<td>&lt;0.01</td>
</tr>
<tr>
<td>Associated sclerosing disease +/-</td>
<td>0/4</td>
<td>20/14</td>
<td>&lt;0.05</td>
</tr>
<tr>
<td>Associated UC</td>
<td>2/2</td>
<td>0/34</td>
<td>&lt;0.01</td>
</tr>
</tbody>
</table>
**PSC Vs IgG4-associated Cholangitis**

<table>
<thead>
<tr>
<th>Feature</th>
<th>PSC</th>
<th>IgG4-associated Cholangitis</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Age (years)</strong></td>
<td>25-45</td>
<td>65</td>
</tr>
<tr>
<td><strong>Gender (male)</strong></td>
<td>65%</td>
<td>80%</td>
</tr>
<tr>
<td><strong>Response to steroids</strong></td>
<td>-</td>
<td>+++</td>
</tr>
<tr>
<td><strong>Association with IBD</strong></td>
<td>+++</td>
<td>-</td>
</tr>
<tr>
<td><strong>Association with cholangio-CA</strong></td>
<td>+++</td>
<td>?</td>
</tr>
<tr>
<td><strong>Other organ involvement</strong></td>
<td>?</td>
<td>+++</td>
</tr>
<tr>
<td><strong>Histology</strong></td>
<td>Obliterative cholangitis and cirrhosis</td>
<td>Abundant IgG4 positive plasma cells</td>
</tr>
<tr>
<td><strong>Cholangiogram</strong></td>
<td>Beaded strictures</td>
<td>Diffuse, segmental and distal CBD strictures</td>
</tr>
<tr>
<td><strong>Elevated serum IgG4</strong></td>
<td>7-9%</td>
<td>&gt;70%</td>
</tr>
</tbody>
</table>
Medical Management of AIP

- Treatment for acute pancreatitis not required
- Diabetes mellitus may respond to steroid
- ERCP with stent placement for jaundice, especially if cholangitis is present
- Steroid therapy is usually effective for biliary strictures, pancreatic duct strictures and chronic pain.
- Surgery is only necessary for unresponsive common bile duct strictures and if malignancy cannot be ruled out.
Medical Management of AIP

- Steroids
  - Oral prednisolone 30-40mg/day for 3-4 weeks → tapering of various duration +/- maintenance therapy
  - No consensus of dosage and duration
  - Steroids may be tapered and sometimes discontinued after a maximal response has been achieved
- Alternate immunomodulatory medications
  - Azathioprine
  - Mycophenolate mofetil
- Insufficient long term follow-up is available to determine the percentage of patients who can eventually eliminate all immunosuppressive therapy.
Summary

- Autoimmune pancreatitis is a recently described disorder presenting with mild epigastric discomfort and jaundice.
- Patients may have associated autoimmune disorders and autoantibodies.
- Imaging shows diffuse or focal pancreatic enlargement, and pancreatic duct and bile duct stenoses.
- There is a low level of awareness of AIP. A high index of suspicion in the correct clinical setting is necessary to make a diagnosis.
- Steroid therapy is almost always effective.
- AIP is clinically important since it is treatable, and may be mistaken for pancreatic carcinoma. It has been aptly called “The Great Masquerader”.
IgG4-related Sclerosing Disease

IgG4-related sclerosing disease

Autoimmune pancreatitis

IgG4-related sclerosing sialadenitis

IgG4-related pseudotumors

IgG4-related sclerosing cholangitis

IgG4-related retroperitoneal fibrosis