Autoimmune Pancreatitis in Children: diagnosis and treatment

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Autoimmune pancreatitis (AIP)

- Very rare condition (1-2% of pancreatitis?)

- Pediatrician relied on adult AIP guidelines (HISORt)
  
  Histology Imaging Serology Other organ involvement Response therapy

- Presentation and long-term outcome of AIP might be different in children than in adults

→ need for specific diagnostic recommendations

Hart, Gastroenterology 2015
Gathered epidemiologic and clinical data on AIP in children using 2 sources:

- Literature review (PubMed, EmBase, CENTRAL) – 30 patients

- INSPPIRE (largest intl. multicentric study on pancreatitis in children) / Cliniques Universitaires St Luc – 18 patients
AIP clinical features

• Symptoms
  – Abdominal pain (43/47, 91%)
  – Obstructive jaundice (20/47, 42%)
  – Weight loss (14/38, 37%)
  – Nausea, vomiting (10/38, 26%)
  – Fatigue (8/38, 21%)
  – Abnormal stool consistency (3/38, 8%)

• Biochemistry and serology
  – Increased amylase / lipase (14/26, 54%)
  – Increased IgG4 (9/40, 22%)
AIP features

- Gland enlargement mass/focal/global 39/47 (83%)
- Capsule-like rim (MRI) 5/32 (16%)
- Main pancreatic duct irregularity 30/47 (64%)
- Common bile duct stricture/tapering 26/47 (55%)
AIP features

• Histology
  – Pancreas fibrosis 19/26 (73%)
  – Lymphoplasmacytic infiltration 24/26 (92%)
  – IgG4 (+) plasmacytes (>10/HPF) 2/25 (8%)
  – Granulocytic epithelial lesion 18/26 (69%)
  – Acinar atrophy 7/26 (27%)

➢ In children oft mixed histology
➢ as per adult criteria-more AIP type 2
AIP features

• Other organ involvement (12/45, 27%)
  – Ulcerative colitis (7/12)
  – Crohn (2/12)
  – Glomerulonephritis (1/12)
  – Hemolytic Anemia (1/12)
  – Celiac Disease (1/12)
### AIP features

#### Treatment

<table>
<thead>
<tr>
<th></th>
<th># of patients/reported</th>
<th>Symptom resolution</th>
<th>(%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Prednisone</td>
<td>29/46</td>
<td>15/16</td>
<td>(63%) (94%)</td>
</tr>
<tr>
<td>No treatment</td>
<td>8/46</td>
<td>2/2</td>
<td>(8%) (100%)</td>
</tr>
</tbody>
</table>

#### (Long term) outcome
- Exocrine pancreatic insufficiency (4/25, 16%)
- Diabetes (3/27, 11%)
### AIP-Outcome – Sickkids experience

<table>
<thead>
<tr>
<th>Pts</th>
<th>DX</th>
<th>Treatment</th>
<th>Fecal elastase (µg/g) before</th>
<th>Fecal elastase (µg/g) after</th>
<th>Trypsinogen (ref 10-57 ng/ml) before</th>
<th>Trypsinogen (ref 10-57 ng/ml) after</th>
<th>Hb1AC (%) before</th>
<th>Hb1AC (%) after</th>
<th>MRCP</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>AIP/UC</td>
<td>Pred, IFX</td>
<td>&lt;15</td>
<td>263</td>
<td>&lt;2.5</td>
<td>6.2</td>
<td>7</td>
<td>5</td>
<td>atrophy</td>
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<tr>
<td>2</td>
<td>AIP/UC</td>
<td>Pred, 5-ASA</td>
<td>291</td>
<td>413</td>
<td>160/3.7</td>
<td>80.6/8.9</td>
<td>5</td>
<td>5</td>
<td>atrophy</td>
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<tr>
<td>3</td>
<td>AIP</td>
<td>Pred</td>
<td>102</td>
<td>430</td>
<td>6.2</td>
<td>6.9/10.5</td>
<td>5.2</td>
<td>5</td>
<td>atrophy</td>
</tr>
<tr>
<td>4</td>
<td>AIP</td>
<td>no tx</td>
<td>N/A</td>
<td>N/A</td>
<td>220/&lt;2.5</td>
<td>4.6</td>
<td>5.2</td>
<td></td>
<td>atrophy</td>
</tr>
<tr>
<td>5</td>
<td>CP, IgG4</td>
<td>no tx</td>
<td>460</td>
<td>350</td>
<td>9.6</td>
<td>46.7</td>
<td>5.2</td>
<td></td>
<td>normal</td>
</tr>
<tr>
<td>6</td>
<td>AIP</td>
<td>Pred</td>
<td>&lt;15</td>
<td>N/A</td>
<td>194</td>
<td>&lt;2.5</td>
<td>5.4</td>
<td>5.2</td>
<td>N/A</td>
</tr>
</tbody>
</table>
AIP lessons

• AIP is seen in children of all ethnical background

• Mean age at diagnosis 13y (2-17y)

• Symptoms were more consistently reported
  – More Abdominal pain, weight loss and fatigue compares to Adults

• IgG4 is rarely positive in children
AIP lessons

• Distinct pathologic imaging findings
  - more focal than diffuse pancreas enlargement compared to Adults

• Histology
  – Mixed features (GEL, plasmacyte infiltration, fibrosis)

• Responds to steroids
  – Higher dose
  – But also pts without steroids showed symptom resolution

• Follow up of exocrine/endocrine function needed
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