GI Complications of Scleroderma

Mitchell D. Forman, D.O., FACR, FACP, FACOI
Dean & Professor
Touro University Nevada College of Osteopathic Medicine
Objectives

• Present a brief overview of the etiopathogenesis of Scleroderma
• Review the manner in which PSS affects the GI tract
• Summarize the management of specific GI complications of PSS
Systemic Sclerosis

• Connective tissue disease of unknown etiology

• Fibrosis & thickening of skin with internal organ involvement – GI, heart, lungs, kidneys

• Incidence ~ 20 per million persons per year
  Estimated 75 - 100,000 cases in USA

• Peak onset 30 - 50 yoa, 4 x more in W > M

• Heterogeneous group of disorders
The Cause of Systemic Sclerosis is Unknown

• Genetics: concordance in identical twins 5.9%, HLA linkage

• Silica, metal dusts, and PVC exposures linked to inc risk in some studies

• No conclusive evidence at this time for infectious agent
Fibroblasts, Endothelial Cells & Lymphocytes

Three important pathological themes:

• **Tissue fibrosis** – inc production of collagen & ECM by fibroblasts
• **Inflammation** – macrophage & T cell, espec at earlier stages
• **Micro-vascular disease** – intimal proliferation, vessel narrowing & thrombosis

It’s not clear what the initiating event is
Fibrosis in Scleroderma
Autoimmunity in Scleroderma
Subtypes of Systemic Sclerosis

- Diffuse cutaneous skin involved – proximal involvement, anti-Scl 70 antibodies
- Limited cutaneous aka CREST – dysmotility, skin involvement & anti-centromere antibodies
- Localized
  - Morphea
  - Linear
- Scleroderma sine scleroderma
- Pre – Scleroderma: Raynauds
CT Disease - Spectrum

Well Defined CT Disorder

Overlap CT Disorder

Undifferentiated CT Disorder
Raynaud’s Phenomenon

Characterized by:

- Episodic ischemia of the digits
- Tri-phasic color changes
  - blanching, cyanosis, erythema
- Often precipitated by cold, Rxs, emotional stress
- May be associated with pain or dysesthesias
- Primary vs Secondary
### Organ Systems Affected in Systemic Sclerosis

<table>
<thead>
<tr>
<th>Organ System</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pulmonary</td>
<td>Leading cause of mortality and major morbidity. Interstitial lung disease, pulmonary hypertension</td>
</tr>
<tr>
<td>GI</td>
<td>Esophageal disorders especially common. Stomach and bowel may be affected</td>
</tr>
<tr>
<td>Renal</td>
<td>“Scleroderma renal crisis” - sudden onset of malignant hypertension and renal insufficiency</td>
</tr>
<tr>
<td>Cardio</td>
<td>High prevalence of arrhythmias, EKG abnormalities</td>
</tr>
<tr>
<td>MS</td>
<td>Arthralgia, myopathy, acro-osteolysis, osteopenia, subcutaneous calcinosis</td>
</tr>
</tbody>
</table>
Oral Disease in Scleroderma
Oral Disease in Scleroderma

- 31 Female pts with PSS, 30+ yoa
- Xerostomia in 70%
  - Inc. dental caries
  - Diseased, missing & mobile teeth
  - Periodental disease & inc periodental memb width
  - Mandibular erosions in 9/31 pts
- Dec. oral aperture
- Inc mat – like telangiectasias


Esophageal Disease in Scleroderma

- Esophageal pathology is common in both diffuse & limited disease (90%) – freq. asymptomatic

- Smooth muscle atrophy & fibrosis leads to defective motility; upper esoph involve infreq

- Patients may experience dysphagia or odynophagia. Also, stasis predisposes to esophageal candidiasis

- Poor contraction of the LES makes GERD a significant problem for many pts. Reflux may lead to esophagitis &/or strictures & aspiration - (Acid suppression Tx)
Lower Esophageal Sphincter in PSS
This image from a double-contrast upper GI study done on a 49-year-old man with Limited Scleroderma shows:

- Dilatation of the distal esophagus
- A patent GE junction
- Numerous filling defects in the esophagus (arrow)

Further endoscopic evaluation lead to a diagnosis of esophageal candidiasis.
Barrett’s Esophagus In PSS
Gastric Involvement in PSS

- Less common than esophagus
- Easy satiety common
- Gastroparesis & vomiting
- Overgrowth of fragile vessels – “watermelon stomach”

- UGI bleeding due to telangiectasia
  Limited Scleroderma
- Gastric antral venous ectasia (GAVE)
- Multiple small meals & supplements
Small Bowel Disease in Scleroderma

- Smooth muscle atrophy & fibrosis impair motility (20 – 60% of pts)

- Symptoms may include bloating, pain, anorexia, N/V & diarrhea. Malabsorption (10 – 30%) may occur secondary to bacterial overgrowth; pseudo-obstruction - antibiotics

- The small bowel may be dilated & have an “accordion-like” appearance due to fibrosis - related shortening of the bowel
Small Bowel Involvement
This image was taken from the double contrast UGI series done on the 49 year old patient with Limited Scleroderma. Note the close approximation of the valvulae conniventes ("stack of coins" sign) in the proximal small bowel indicated by the arrow.
Colon & Anorectal Involvement in Scleroderma

- 10 – 50% espec anorectum
- Muscle weakness usually produces constipation & fecal incontinence
- Wide – mouth diverticuli, spont perforation
- High fiber diet or supplements that cause a bulkier and looser stool
<table>
<thead>
<tr>
<th>Part of GI tract</th>
<th>Manifestation</th>
<th>Therapeutic options</th>
</tr>
</thead>
<tbody>
<tr>
<td>Esophageal dysfunction</td>
<td>Gastroesophageal reflux</td>
<td>Diet modification</td>
</tr>
<tr>
<td></td>
<td>Barrett's esophagus</td>
<td>H2 blockers</td>
</tr>
<tr>
<td></td>
<td>Centrilobular interstitial lung disease</td>
<td>Proton pump inhibitors</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prokinetic agents</td>
</tr>
<tr>
<td></td>
<td></td>
<td>GABA(B) agonists (baclofen)</td>
</tr>
<tr>
<td>Gastric involvement</td>
<td>Gastroparesis</td>
<td>Prokinetic agents</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Botox injection into pyloric sphincter</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Laser therapy</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Immunomodulatory therapy</td>
</tr>
<tr>
<td>Small intestine</td>
<td>Bacterial overgrowth</td>
<td>Antibiotics</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prokinetic agents</td>
</tr>
<tr>
<td></td>
<td>Malabsorption</td>
<td>Antibiotics</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Enteral and parenteral nutrition</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prokinetic agent</td>
</tr>
<tr>
<td></td>
<td>Pseudoobstruction</td>
<td>Nil per os</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intravenous hydration</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Prokinetic agents</td>
</tr>
<tr>
<td>Large intestine</td>
<td>Large diverticuli</td>
<td>Diet modification</td>
</tr>
<tr>
<td></td>
<td>Anorectal sphincter dysfunction</td>
<td>Biofeedback</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sacral nerve stimulation</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Surgical repair</td>
</tr>
<tr>
<td>Biliary</td>
<td>Primary biliary cirrhosis</td>
<td>Ursodeoxycholic acid</td>
</tr>
</tbody>
</table>