Update in deposition diseases

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Primary Mucinoses

Dermal
- Lichen Myxedematosus
- Reticular erythematous mucinosis (REM)
- Scleredema
- Pretibial myxedema
- Cutaneous focal mucinosis
Reticular erythematous mucinosis

- Young to middle-aged people (> women)
- Erythematous reticular or plaque-like eruption in the middle of the chest or back
- Photoinduction, hormonal influence (pill)
Reticular erythematous mucinosis

Interstital mucin in the superficial dermis with perivascular T-lymphocytic infiltrate

No epidermal involvement

Alcian blue pH 2.5
Primary Mucinoses

**Dermal**
Lichen Myxedematosus
Reticular erythematous mucinosis (REM)
Scleredema
Pretibial myxedema
Cutaneous focal mucinosis
Scleredema

- Symmetrical diffuse non-pitting induration of the upper part of the body
- Hands and feet spared
- Association with
  - Streptococcal infection
  - Monoclonal gammopathy
  - Diabetes
Scleredema

- Diffuse (interstitial) mucin deposition in a thickened reticular dermis
- **Additional findings**
  - Fenestration of collagen bundles
  - Normal fibroblast number
Scleredema
Primary Mucinoses

**Dermal**

- Lichen Myxedematosus
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- Scleredema
- Pretibial myxedema
- Cutaneous focal mucinosis
Localized (pretibial) myxedema

- Indurated waxy plaques on the shins with peau d’orange appearance
- Rarely shoulders, arms, abdomen
- Associated with hyperthyroidism (mostly Graves’ disease) often following treatment of thyroid
- Frequent association with exophthalmos and thyroid acropachy
Localized (pretibial) myxedema

Mucin, hyperkeratosis, follicular plugging

Large amount of mucin throughout reticular dermis

Mucin in deep dermis
Localized (pretibial) myxedema

Hyperkeratosis, acanthosis

Large amount of mucin

mastcells
Pitfalls in pretibial myxedema
Obesity-associated lymphoedematous mucinosis (without thyroid disease)

Histological differences

- Pretibial (lymphedematous) mucinosis in euthyroid
  
  - No thyroid disease
  - Obesity
  - Superficial mucin
  - Stasis dermatitis
  - Lymphedema

- “Real” pretibial myxedema” in hyperthyroidism
  
  - Graves’ disease
  - Mucin throughout the reticular dermis
  - No features of angioplasia
Pretibial (lymphedematous) mucinosis in obese

Pretibial myxedema
Primary Mucinoses

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**Follicular**
- Pinkus’ follicular mucinosis
Cutaneous focal mucinosis

- Benign skin-colored papule < 1 cm
- Anywhere on the body, except over the joints of hands and feet
- Reactive lesion
- Diagnosis is a histologic one.
Cutaneous focal mucinosis

- Focal accumulation of mucin through much of the dermis resembling an early myxoid cyst
- Spindle-cells or stellate fibroblasts (Factor XIIIa+, CD34+)
Pitfalls
Superficial angiomyxoma

- Benign acquired neoplasm
- Solitary (without systemic abnormalities)
- Multiple (Carney’s complex)
- May recur after excision
- Head and neck
- Multilobular, diffuse mucin deposition involving subcutis
A poorly-circumscribed dermal lesion with multilobular areas of myxoid stroma. Thin-walled enlarged blood vessels.

Alcian blue stain pH2.5
## CLINICAL AND MICROSCOPIC CRITERIA FOR DISTINGUISHING MYXOMA FROM FOCAL CUTANEOUS MUCINOSIS

<table>
<thead>
<tr>
<th>CRITERIA</th>
<th>(ANGIO) MYXOMA</th>
<th>CUTANEOUS FOCAL MUCINOSIS</th>
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</thead>
<tbody>
<tr>
<td><strong>Size</strong></td>
<td>1-5 cm</td>
<td>&lt; 1 cm</td>
</tr>
<tr>
<td><strong>Mucin deposit</strong></td>
<td>Diffuse, multilobular</td>
<td>focal</td>
</tr>
<tr>
<td><strong>Level of deposit</strong></td>
<td>dermis, subcutis</td>
<td>dermis</td>
</tr>
<tr>
<td><strong>Vascularity</strong></td>
<td>increased</td>
<td>normal</td>
</tr>
<tr>
<td><strong>Epithelial component</strong></td>
<td>constant</td>
<td>rare</td>
</tr>
<tr>
<td><strong>Stromal cells</strong></td>
<td>$\alpha$-SMA +, FXIII -</td>
<td>$\alpha$-SMA -, FXIII +</td>
</tr>
<tr>
<td><strong>Additional findings</strong></td>
<td>reticulin network, microhemorrhages, inflammation</td>
<td></td>
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</tbody>
</table>
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*Follicular*
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Follicular mucinosis

Mucin within dilated follicles causing keratinocytes to disconnect
Follicular mucinosis


- Primary (Pinkus) (Idiopathic)
- Secondary associated with many disorders, mainly mycosis fungoides