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Update in deposition diseases

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Cutaneous deposition disorders

- Endogenous
- Exogenous
Cutaneous mucinosis

Hematoxylin-eosin

Alcian blue pH 2.5

Colloidal iron
The cutaneous mucinoses

- **Primary Mucinoses**
  - Mucin deposit is the main histologic finding resulting in clinically distinctive lesions

- **Secondary Mucinoses**
  - Mucin deposit is only an additional histologic finding
Secondary mucinoses

- Granuloma annulare
- Dermatomyositis
- Scleroderma
- Lupus erythematosus
- Degos’ disease
- ............
Primary Mucinoses

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

*Follicular*
- Pinkus’ follicular mucinosis
Primary Mucinoses

- The distribution of the mucin differs in the various dermal mucinoses and is generally not diagnostically specific

- The diagnosis requires clinicopathologic correlation

- Three histologic clues:
  1. The pattern of mucin distribution (focal or diffuse)
  2. The level of mucin deposit
  3. Some additional findings
Pattern of mucin distribution

Focal

Diffuse/interstitial
Level of mucin deposit

- Superficial
- Mid/lower
- Deep

Lichen myxedematosus

Pretibial myxedema
Primary Mucinoses

Dermal

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

- Generalized and sclerodermoid type (Scleromyxedema) with monoclonal gammopathy and systemic manifestations
- Localized type with only cutaneous involvement
Scleromyxedema

Generalized sclerodermoid eruption with stiff infiltration
Scleromyxedema

Closely spaced papules on sclerodermoid skin
Scleromyxedema

Papules in a linear fashion

Papules confluent into plaques

Papules in a linear fashion
Histopathology: microscopic triad of scleromyxedema

1. Mucin
2. Fibroblast proliferation
3. Fibrosis
Scleromyxedema is associated with a monoclonal gammopathy IgGλ (rarely progressing to myeloma)
Scleromyxedema is associated with systemic, even lethal, manifestations

- Muscular (dysphagia, myositis) 30%
- Pulmonary 17%
- Rheumatologic (carpal tunnel) 15%
- Neurologic (neuropathy, psychosis, coma) 10%
Lichen Myxedematosus (Papular mucinosis)

SCLEROMYXEDEMA

- Generalized papular and sclerodermoid form with monoclonal gammopathy and systemic manifestations
- Microscopic triad of mucin, fibrosis and increased fibroblasts

LOCALIZED LICHEN MYXEDEMATOSUS

- Papules confined to a few sites (trunk and limbs) with only cutaneous involvement. No monoclonal gammopathy or systemic involvement
- Mucin with variable fibroblast proliferation in the absence of fibrosis

Rongioletti & Rebora, JAAD, 2001
Localized lichen myxedematosus

- Discrete type
- Acral persistent papular mucinosis
- Cutaneous mucinosis of infancy
Localized lichen myxedematosus (discrete type)

Chronic eruption of papules from just a few to hundreds on trunk and limbs in the absence of sclerotic features.
Localized lichen myxedematosus (discrete type)

Diffuse, interstitial deposit of mucin with variable fibroblast proliferation in the absence of fibrosis. Slight infiltrate
Acral persistent papular mucinosis
Rongioletti et al, Arch Dermatol 1986

- > Females (3:1)
- Flesh-colored papules on the back of hands and forearms
- No resolution
Acral persistent papular mucinosis

- Focal mucin deposition
- Upper dermis
- Grenz zone, normal fibroblast number, no fibrosis

Rongioletti et al, Arch Dermatol 1986
Cutaneous mucinosis of infancy
(paediatric variant of localized lichen myxedematosus)
Podda M, Rongioletti F, Wolter M. Br J Dermatol 2001;144
Cutaneous mucinosis of infancy
(paediatric variant of localized lichen myxedematosus)

Mucin so superficial as to look as if it is ‘enclosed’ by epidermis
Lichen Myxedematosus (Papular mucinosis)  

**SCLEROMYXEDEMA**

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