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Update in deposition diseases

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Pink-gray, amorphous, hyaline deposits

Hyaline

Hyalinosis cutis et mucosae

Amyloid

Amyloidoses
Hyalinosis cutis et mucosae (Lipoid proteinosis)

- Hyaline material in superficial dermis around capillaries and adnexa
- Extensive deposits later (vertically oriented)
- PAS++, Congo-red weakly or neg
- Alcian blue+, Colloidal iron +
- type IV collagen, laminin, neutral and acid GAG

Hyperkeratosis, achantosis
Pink-gray, amorphous, hyaline deposits

Hyaline
  ➔ Hyalinosis cutis et mucosae

Amyloid
  ➔ Amyloidoidoses
Amyloidoses with skin manifestations

- Primary systemic amyloidosis, multiple myeloma-related
  - AL (immunoglobulin light chain)

- Skin-limited amyloidoses
  - AK (amyloid keratin protein)
    - Lichen, macular, biphasic
    - Nodular

- AL (immunoglobulin light chain)
Skin lesions in AL Amyloidosis (myeloma-related)

- Pinch purpura
- Periorbital ecchymoses precipitated by coughing, Valsalva maneuver, proctoscopy

« Raccoon sign »
Skin lesions in AL Amyloidosis

- Waxy translucent or purpuric papules, nodules, plaques
- Macroglossia
AL amyloidosis (myeloma-related)

The hyaline deposits in the entire dermis with some artifactual separation. No inflammatory infiltrate.
Skin-limited amyloidoses (cutaneous localized myloidoses)

Lichen (papular) amyloidosus on the shin

Macular amyloidosis on the back
Lichen (Papular) amyloidosus

The hyaline deposits expand dermal papillae with hyperkeratosis and acanthosis
Lichen (Papular) amyloidosus

The hyaline deposits expand dermal papillae with hyperkeratosis and acanthosis

Red Congo stain
Endogenous

Mucin

Hyaline deposit

Calcium
Calcium

- Stains purple/blue with H-E
- Owing to the presence of phosphate and carbonate, calcium stains black with the Von Kossa stain
Calcinosis cutis
(Cribier B, Grosshans E, Ann Dermatol Venereol)

- **Idiopathic**
- **Metastatic** associated with elevated levels of calcium or/and phosphate
- **Dystrophic** with calcium deposition in degenerate tissue
- Von Kossa, Alizarin red stains
Dystrophic calcification

- Most common variant with calcium deposition in degenerate tissue after inflammation or trauma
- Connective tissue disease
- Iatrogenic
- Acne scars
- Pseudoxanthoma elasticum

Calcinosi in scleroderma

Von Kossa

Pseudoxanthoma elasticum
Subepidermal calcified nodule (idiopathic calcinosis)

- Solitary nodule on the head (ear) or extremities in young people
- Basophilic calcium deposits are multiple, small, globular in the uppermost dermis
- Idopathic calcinosis (preexisting nevus?)
Calciphylaxis in patients with serious renal failure (metastatic calcinosis)

- Skin necrosis and ulceration in chronic renal failure
- Increased calcium-phosphorus product and hyperparathyroidism
- Calcification of medium-sized arterioles affecting both media and intima
Take-home message

- **Cutaneous deposition disorders** are a heterogeneous group of relatively uncommon diseases
- Histopathology is the most specific diagnostic tool sparing more invasive methods
- Many of these disorders cause only subtle or non-specific changes in biopsy specimens
- Special stains in addition to a clinico-pathological correlation are essential
- Special processing (immunohistochemistry, electron microscopy) may help