CHAPTER 1
Horny Layer

CHAPTER MENU

Reduced granular layer
Prominent granular layer

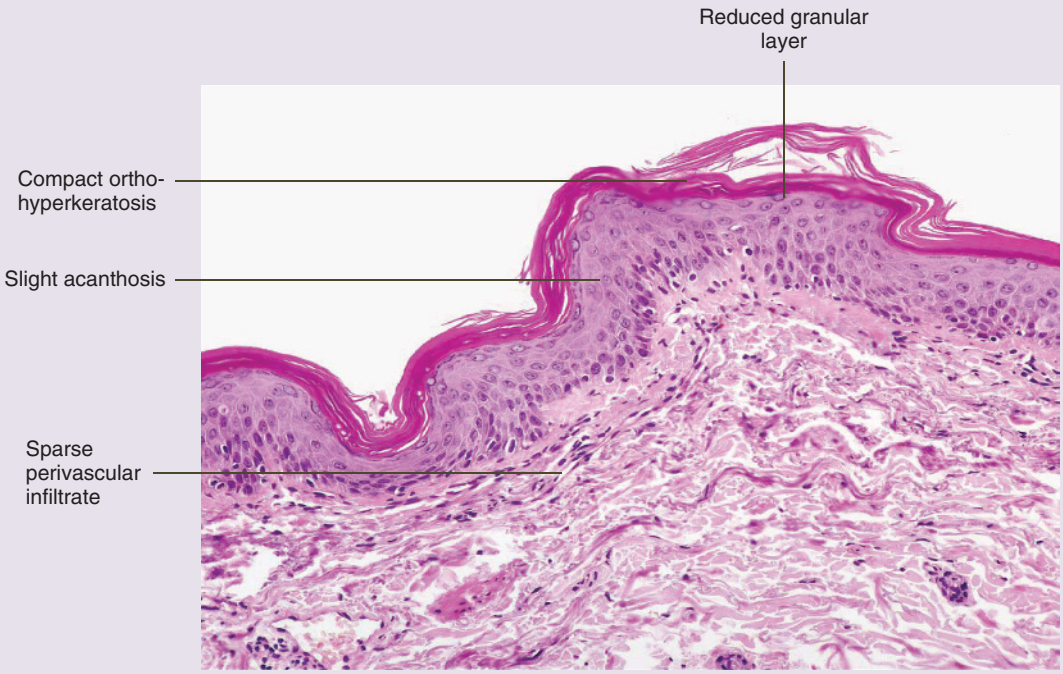
Atlas of Dermatopathology: Practical Differential Diagnosis by Clinicopathologic Pattern, First Edition.
Edited by Günter Burg MD, Werner Kempf MD, and Heinz Kutzner MD. Co-Editors: Josef Feit MD, and Laszlo Karai MD.

HORNY LAYER

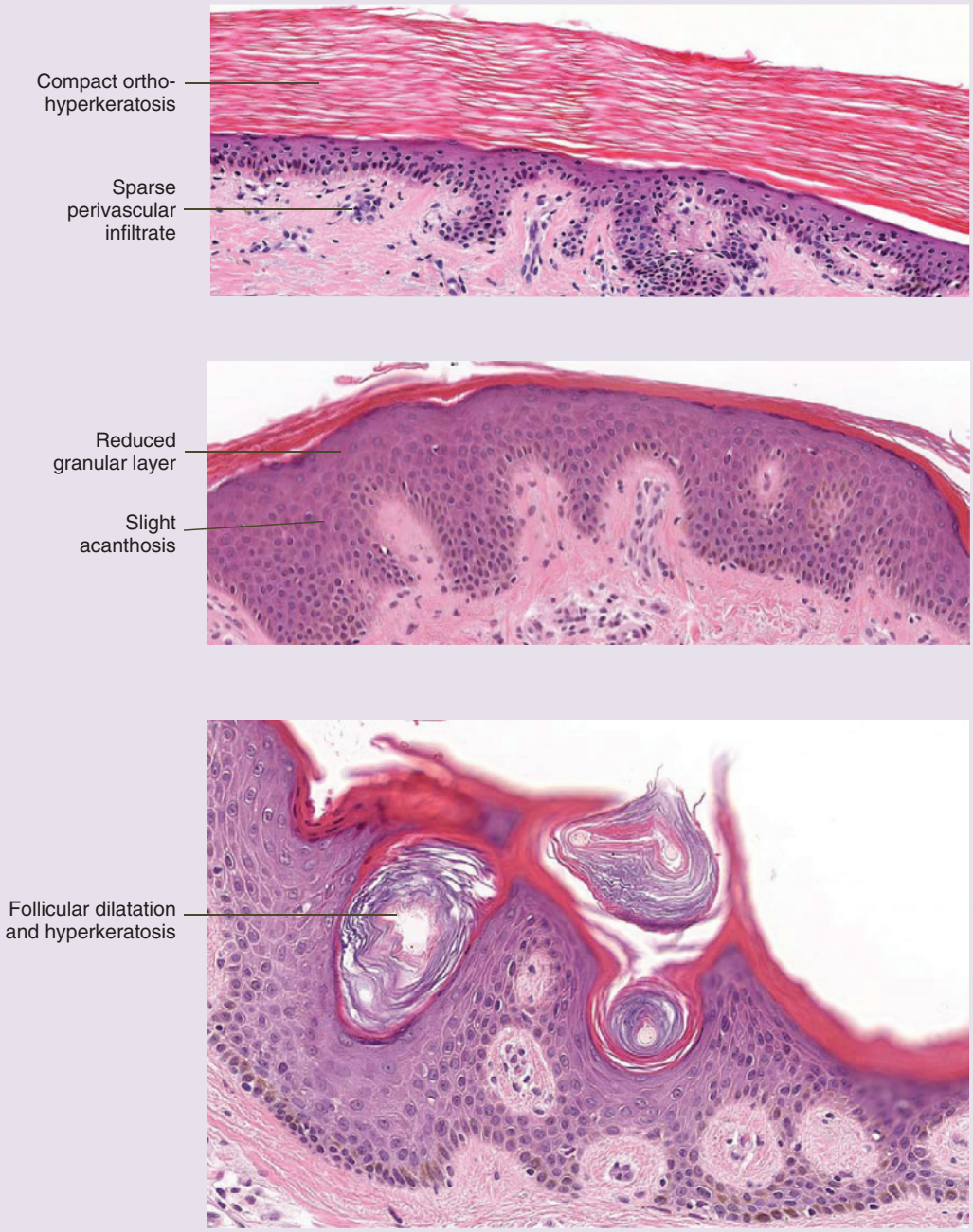
PROTOTYPE: Ichthyosis vulgaris

Gray-white scales

CI: Starts in first year of life, dry rough scaly skin, gray-white scales are shed, symmetrical sparing of flexural areas, hyperlinear palms and soles, often atopic dermatitis (50%).



Ichthyosis vulgaris



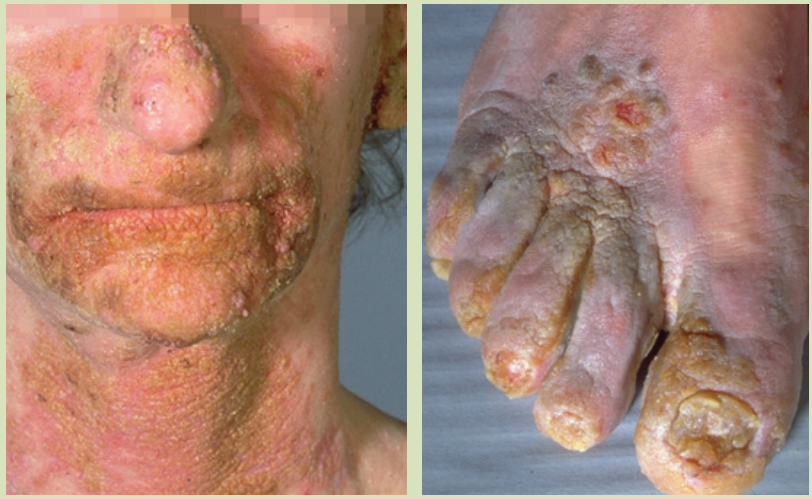
Hi: Compact orthohyperkeratosis, granular layer reduced or absent, lack of parakeratosis, follicular dilatation and hyperkeratosis. Epidermis usually normal, sometimes acanthotic or atrophic. No or sparse perivascular infiltrate in the papillary dermis.

VARIANTS: Acquired ichthyosis vulgaris

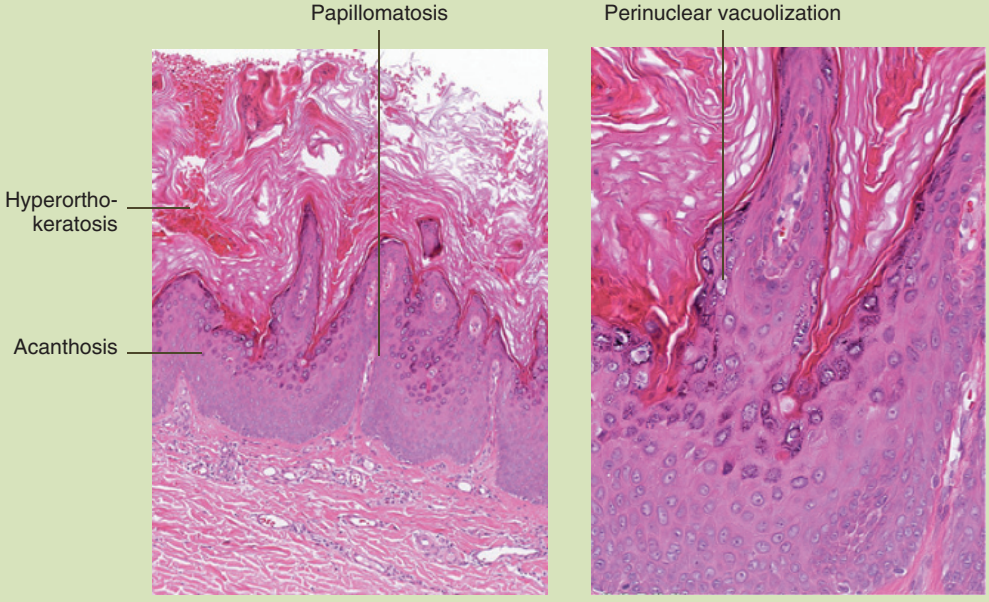
Histology is identical to ichthyosis vulgaris.

DIFFERENTIAL DIAGNOSIS: Ichthyosis hystrix

Massive hyperkeratosis



CI: Massive, dark, sometimes spiny hyperkeratosis. Various genetic forms exist. Flexures, palms and soles are involved.



Hi: Mild hyperorthokeratosis, acanthosis, papillomatosis, elongation of rete ridges. Perinuclear vacuolization of granular and spinous layer keratinocytes, presenting epidermolytic features.

Other Diagnosis

Refsum syndrome (heredopathia atactica polyneuritiformis): *Vacuolization of basal and suprabasal keratinocytes (accumulation of phytanic acid; Sudan red stain)*

X-linked dominant ichthyosis (Harlequin ichthyosis): *Clinical features similar to ichthyosis vulgaris, but flexures are involved, undescended testes in 30%. Vacuolization of basal and suprabasal keratinocytes (accumulation of phytanic acid; Sudan red stain)*

Lamellar ichthyosis: *Genetically heterogeneous disorder, usually present at birth presenting as collodion baby in case of generalized involvement. Erythrodermic and non-erythrodermic forms. Transglutaminase-deficiency in most forms. Histology shows mild to moderate hyperorthokeratosis, stratum granulosum normal or broadened, acanthosis, papillomatosis*

Bullous, epidermolytic ichthyosis (bullous form of erythrodermia ichthyosiformis congenitalis): *Erythroderma at birth with diffuse blistering and erosions, like burned. Histologically the most striking feature is acanthokeratolysis with epidermal thickening leading to superficial blister formation. Tonofilaments can be seen as dark clumps in a shell-like arrangement around the nucleus*

Syndromes of ichthyosis and trichothiodystrophy (Tay syndrome): *Additional clinical symptoms and biochemical findings.*

References

de Berker, D., W. A. Branford, S. Soucek, and L. Michaels (1993). "Fatal keratitis ichthyosis and deafness syndrome (KIDS). Aural, ocular, and cutaneous histopathology." *Am J Dermatopathol* **15**(1): 64–9.

de Wolf, K., J. M. Gourdain, G. D. Dobbeleer, and M. Song (1995). "A particular subtype of ichthyosis congenita type III. Clinical, light, and electron microscopic features." *Am J Dermatopathol* **17**(6): 606–11.

Frenk, E. and B. Mevorah (1977). "The keratinization disorder in collodion babies evolving into lamellar ichthyosis. Its possible relevance for determining the primary defect in lamellar ichthyosis." *J Cutan Pathol* **4**(6): 329–37.

Hoang, M. P., K. R. Carder, A. G. Pandya, and M. J. Bennett (2004). "Ichthyosis and keratotic follicular plugs containing dystrophic calcification in newborns: distinctive histopathologic features of x-linked dominant chondrodysplasia punctata (Conradi-Hunermann-Happle syndrome)." *Am J Dermatopathol* **26**(1): 53–8.

Nabai, H. and A. H. Mehregan (1979). "Ichthyosis linearis circumflexa." *J Cutan Pathol* **6**(2): 148–9.

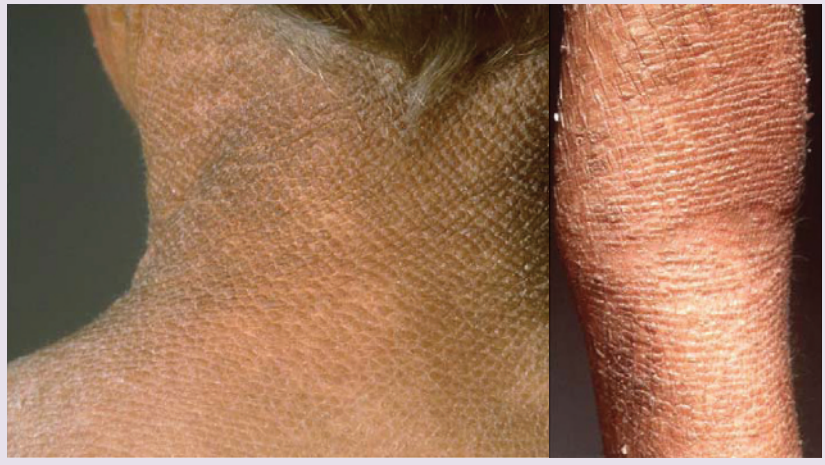
Niemi, K. M. and L. Kanerva (1989). "Ichthyosis with laminated membrane structures." *Am J Dermatopathol* **11**(2): 149–56.

Niemi, K. M., K. Kuokkanen, L. Kanerva, and J. Ignatius (1993). "Recessive ichthyosis congenita type IV." *Am J Dermatopathol* **15**(3): 224–8.

Okulicz, J. F. and R. A. Schwartz (2003). "Hereditary and acquired ichthyosis vulgaris." *Int J Dermatol* **42**(2): 95–8.

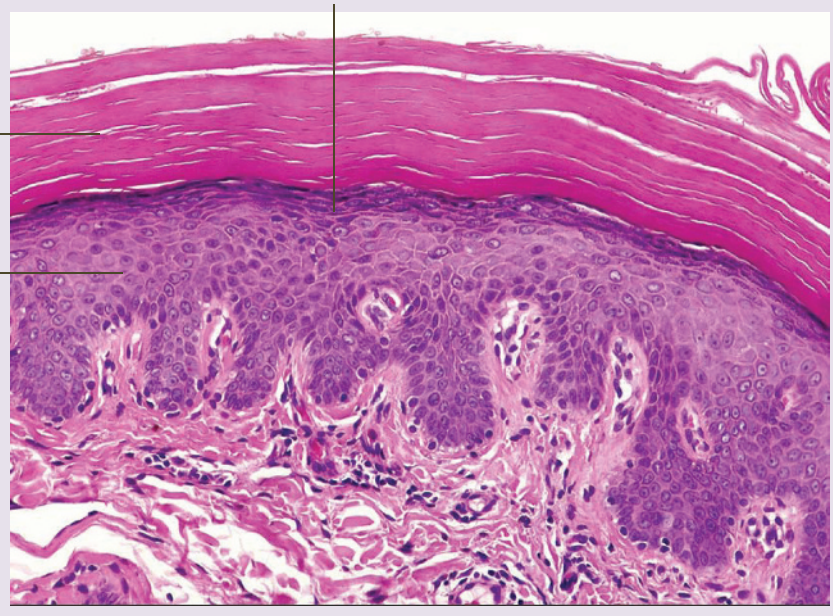
Sandler, B. and K. Hashimoto (1998). "Collodion baby and lamellar ichthyosis." *J Cutan Pathol* **25**(2): 116–21.

PROTOTYPE: Lamellar ichthyosis



Lamellar ichthyosis. Neck and cubital area

CI: Genetically heterogeneous disorder, usually manifest at birth presenting as collodion baby in case of generalized involvement. Erythrodermic and non-erythrodermic forms. Transglutaminase deficiency in most forms.



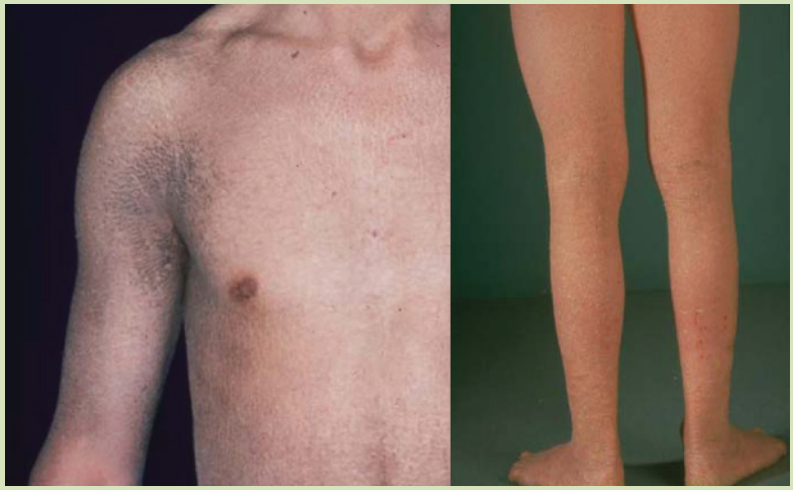
Hi: Mild to moderate hyperorthokeratosis, stratum granulosum normal or broadened, acanthosis, papillomatosis.

DIFFERENTIAL DIAGNOSIS: Congenital ichthyosis group X-linked dominant ichthyosis (Harlequin ichthyosis)

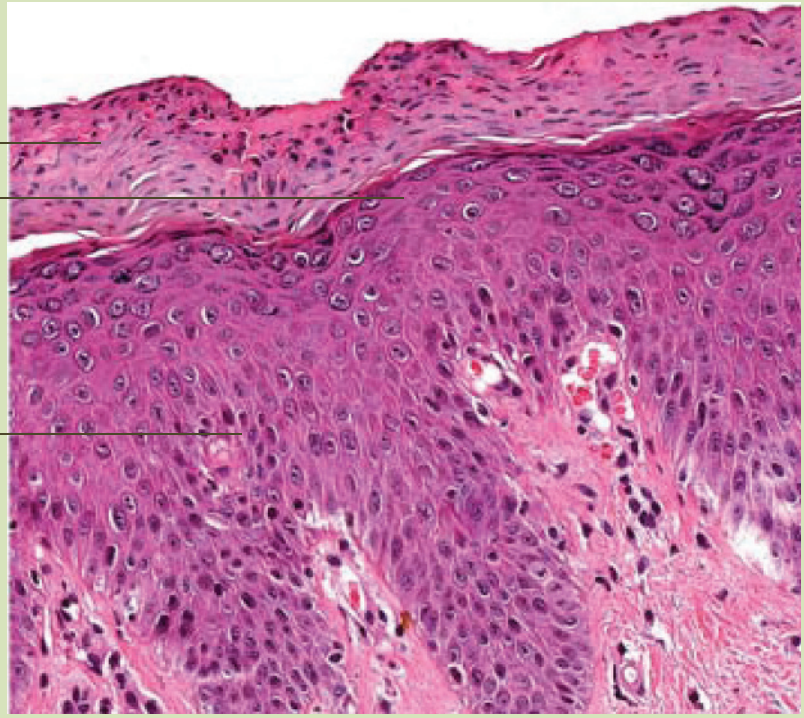
Cl: Similar to ichthyosis vulgaris, but flexures are involved, undescended testes in 30%.

Hi: Vacuolization of basal and suprabasal keratinocytes (accumulation of phytanic acid; Sudan red stain).

DIFFERENTIAL DIAGNOSIS: X-linked recessive ichthyosis



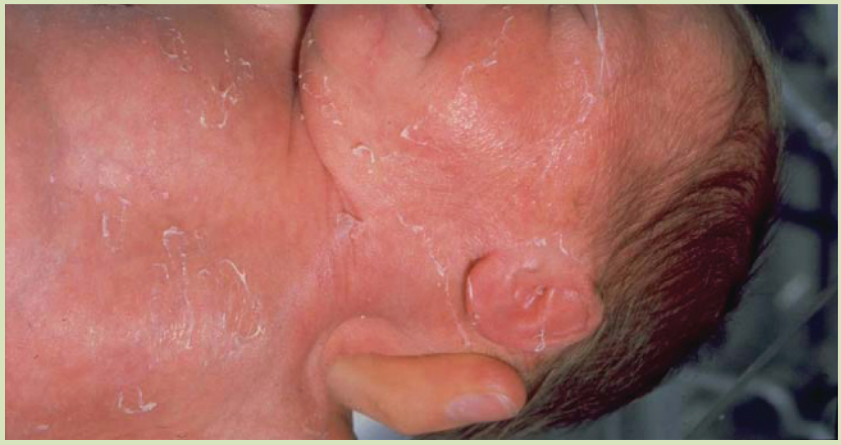
CI: Starts in the first week of life with fine scales and mild erythema, aggravating after a few months. Brown scales giving a dirty appearance cover the whole integument, without sparing of flexural areas.



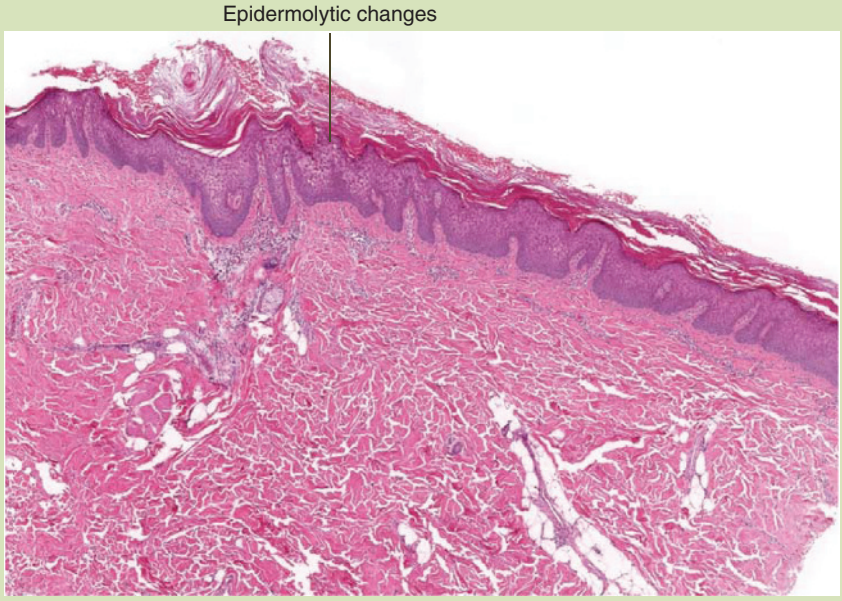
Hi: Marked hyperkeratosis, thickened or normal and sometimes thinned granular layer, spinous layer variably acanthotic and papillomatous, mild to marked perivascular infiltrate in the papillary dermis.

DIFFERENTIAL DIAGNOSIS: Bullous epidermolytic ichthyosis (bullous form of congenital ichthyosiform erythroderma)

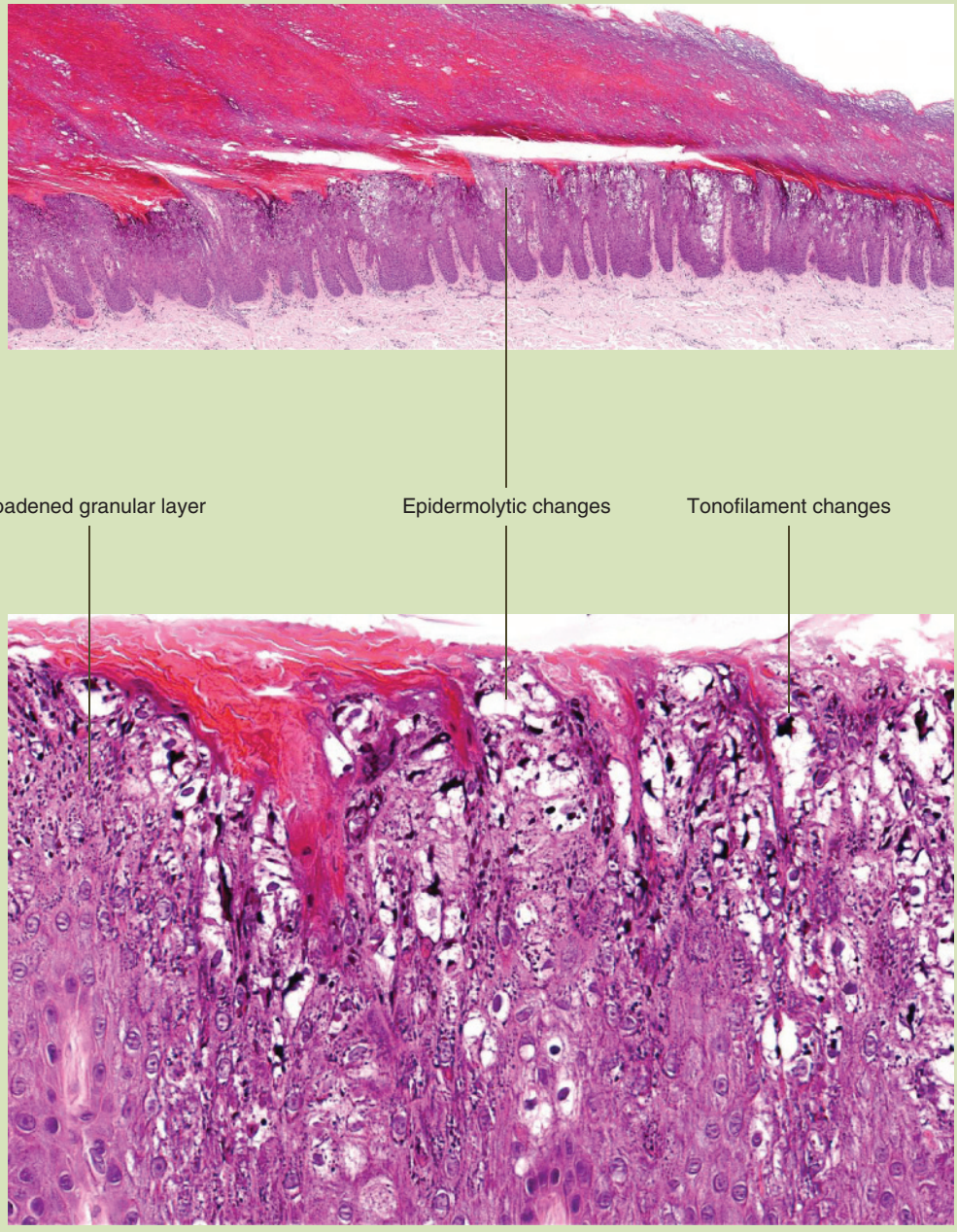
Congenital ichthyosiform erythroderma



CI: Erythroderma at birth with diffuse blistering and erosions, as if burned.



Bullous epidermolytic ichthyosis



Hi: Epidermolytic changes in the upper part of the spinous and the broadened granular layer, which may lead to superficial blister formation. Tonofilaments can be seen as dark clumps in a shell-like arrangement around the nucleus.

Ichthyosis and deafness syndromes: *Additional clinical symptoms and biochemical findings*

- *Ichthyosis and deafness syndromes*
  - *Hystrix-like ichthyosis with deafness (HID)*
  - *Keratitis, ichthyosis-like hyperkeratosis and deafness (KID)*
- *Ichthyosis hystrix Curth-Macklin: epidermolytic changes without bullae*
- *Erythrodermia congenitalis ichthyosiformis*
- *Neutral lipid storage disease with ichthyosiform erythroderma (Dorfman syndrome): foamy cytoplasm of keratinocytes in the basal and the granular layer*

Erythrokeratoderma variabilis, various forms: *Migratory erythema and/or persistent hyperkeratotic plaques. Orthohyperkeratosis over a normal granular layer, acanthosis and papillomatosis. Perivascular lymphocytic infiltrate of variable intensity in the upper dermis*

DIFFERENTIAL DIAGNOSIS: Other Skin Diseases

- ***Acanthosis nigricans****: confined to flexural areas. Hyperpigmentation of epidermal basal layer*
- ***Epidermal nevus*** (*see* Chapter 2, Pruriginous, page 47) *circumscribed lesion with acanthosis and hyperkeratosis*
- ***Palmoplantar keratodermas****: confined to palmoplantar areas*
- ***Chronic eczema*** *(lichen simplex chronicus)* (*see* Chapter 2, Chronic, page 36) *foci of parakeratosis, perivascular lymphocytic infiltrate in the upper dermis*
- ***Pityriasis rubra pilaris*** (*see* Chapter 2, psoriasiform, page 56): *Horizontally and vertically alternating ortho- and hyperparakeratosis (checkerboard sign). Subtle perivascular infiltrate, clinically nappes claires*
- ***Clavus*** (*see* Chapter 2, Pruriginous, page 46): *Circumscribed lesion with acanthosis and hyperkeratosis. No inflammation*

References

Hoang, M. P., K. R. Carder, *et al.* (2004). "Ichthyosis and keratotic follicular plugs containing dystrophic calcification in newborns: distinctive histopathologic features of x-linked dominant chondrodysplasia punctata (Conradi-Hunermann-Happle syndrome)." *Am J Dermatopathol* **26**(1): 53–8.