

Erythrokeratoderma

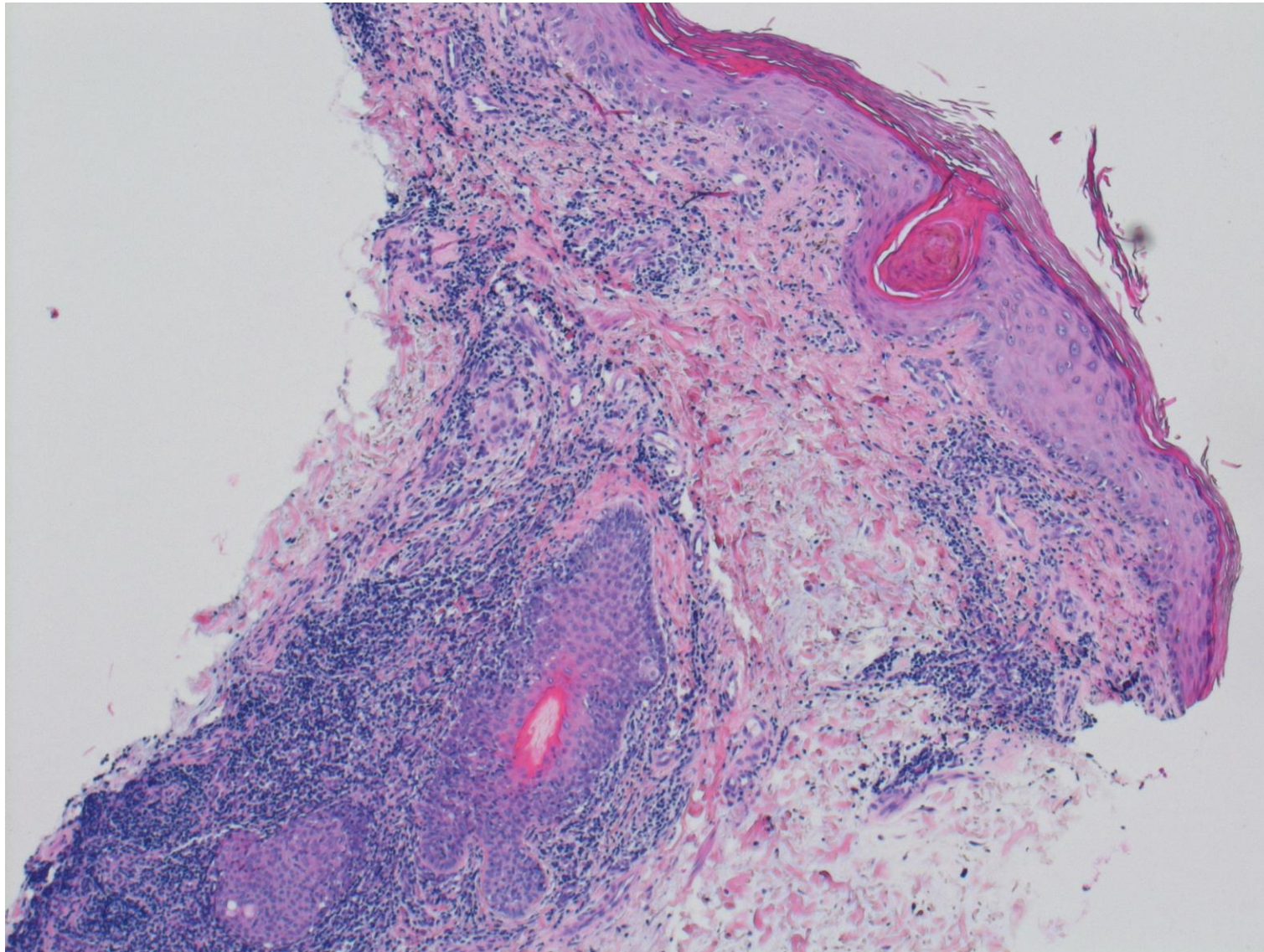
Erythrokeratoderma is the descriptive name given to a rare group of hereditary (autosomal dominant) disorders of hyperkeratosis with erythema. Well-demarcated (circumscribed) plaques are formed. The underlying defect is a mutation in one of the genes encoding connexin, a gap junction protein. Several types of erythrokeratoderma are known, including erythrokeratoderma variabilis, erythrokeratoderma progressiva symmetrica, progressive partially symmetrical erythrokeratoderma with peripheral neuropathy and deafness, erythrokeratoderma with ataxia, etc. It should be noted that the concept of erythrokeratoderma overlaps with congenital ichthyosis. Ichthyosis vulgaris shows generalized skin lesions but lacks erythema. In contrast, other types of ichthyosis commonly accompany erythematous changes. These include congenital ichthyosiform erythroderma, lamellar ichthyosis (Harlequin-type ichthyosis), Netherton syndrome, and X-linked ichthyosis.

Ref.-1: Abeyakirthi S. Erythrokeratoderma. 2007. DermNet.

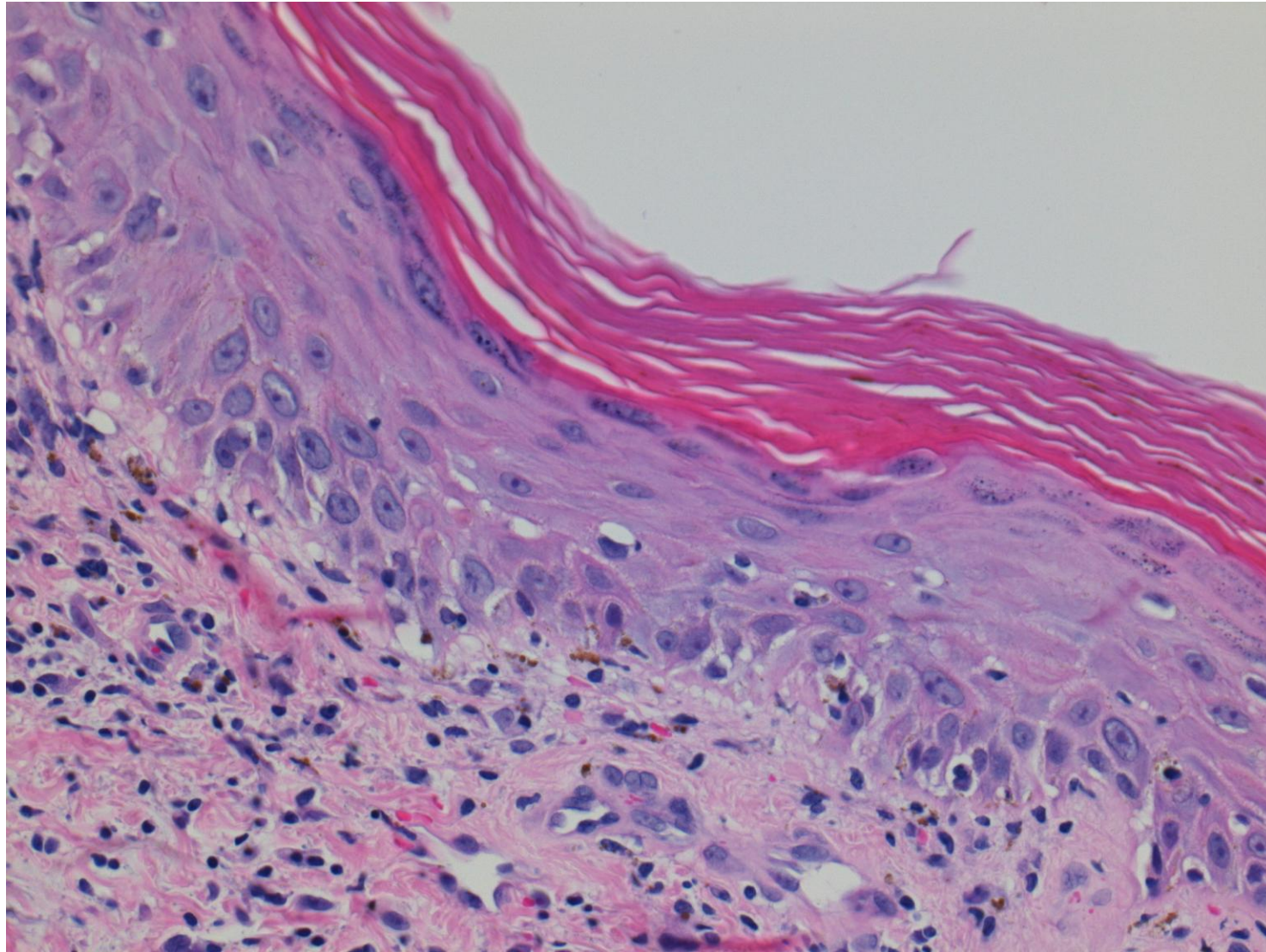
<https://dermnetnz.org/topics/erythrokeratoderma>

Ref.-2: Dr. Zero in Dermatology. 2019. Ichthyoses, erythrokeratodermas, and related disorders.

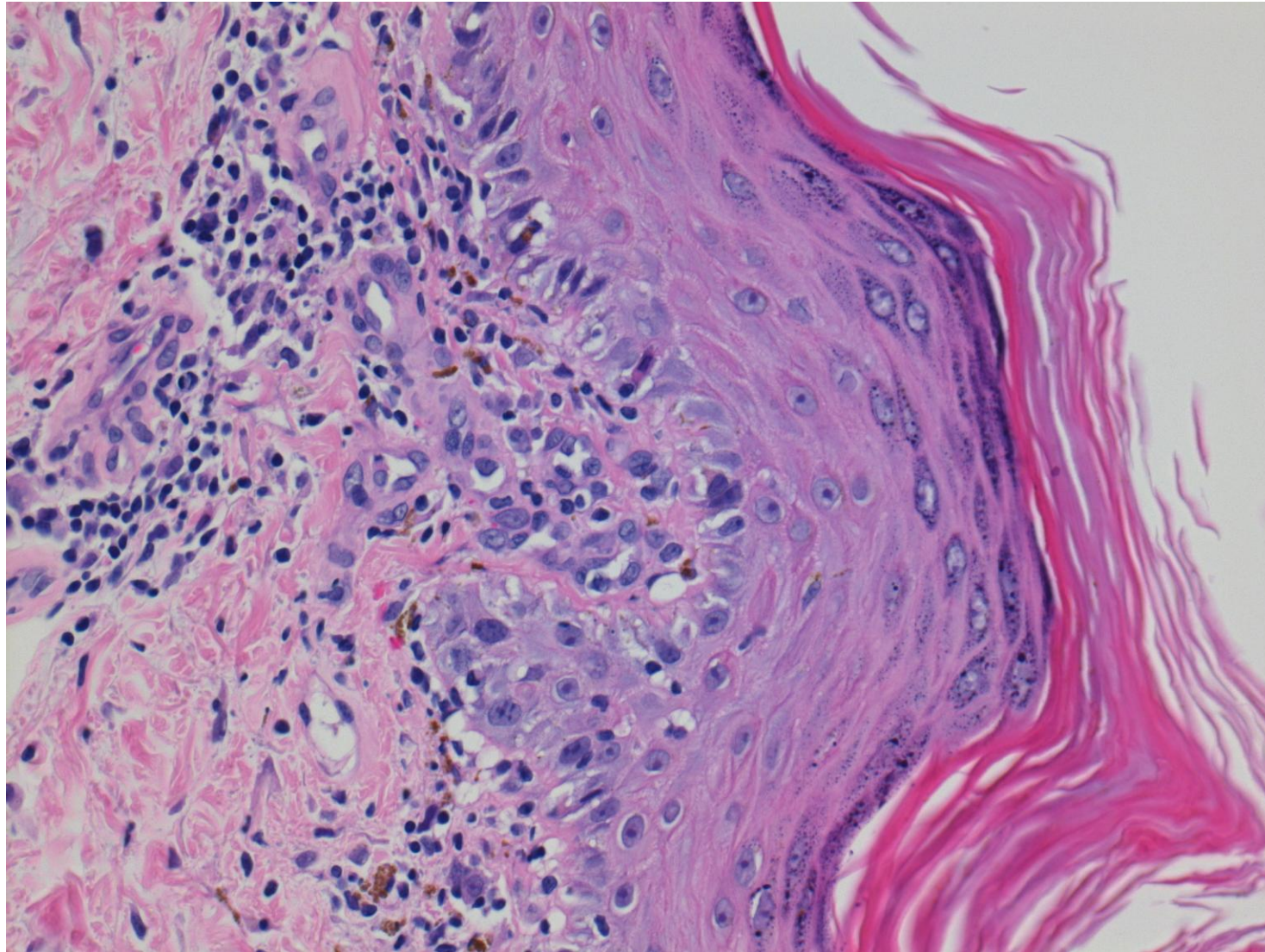
Plastic Surgery Key. <https://plasticsurgerykey.com/ichthyoses-erythrokeratodermas-and-related-disorders/>



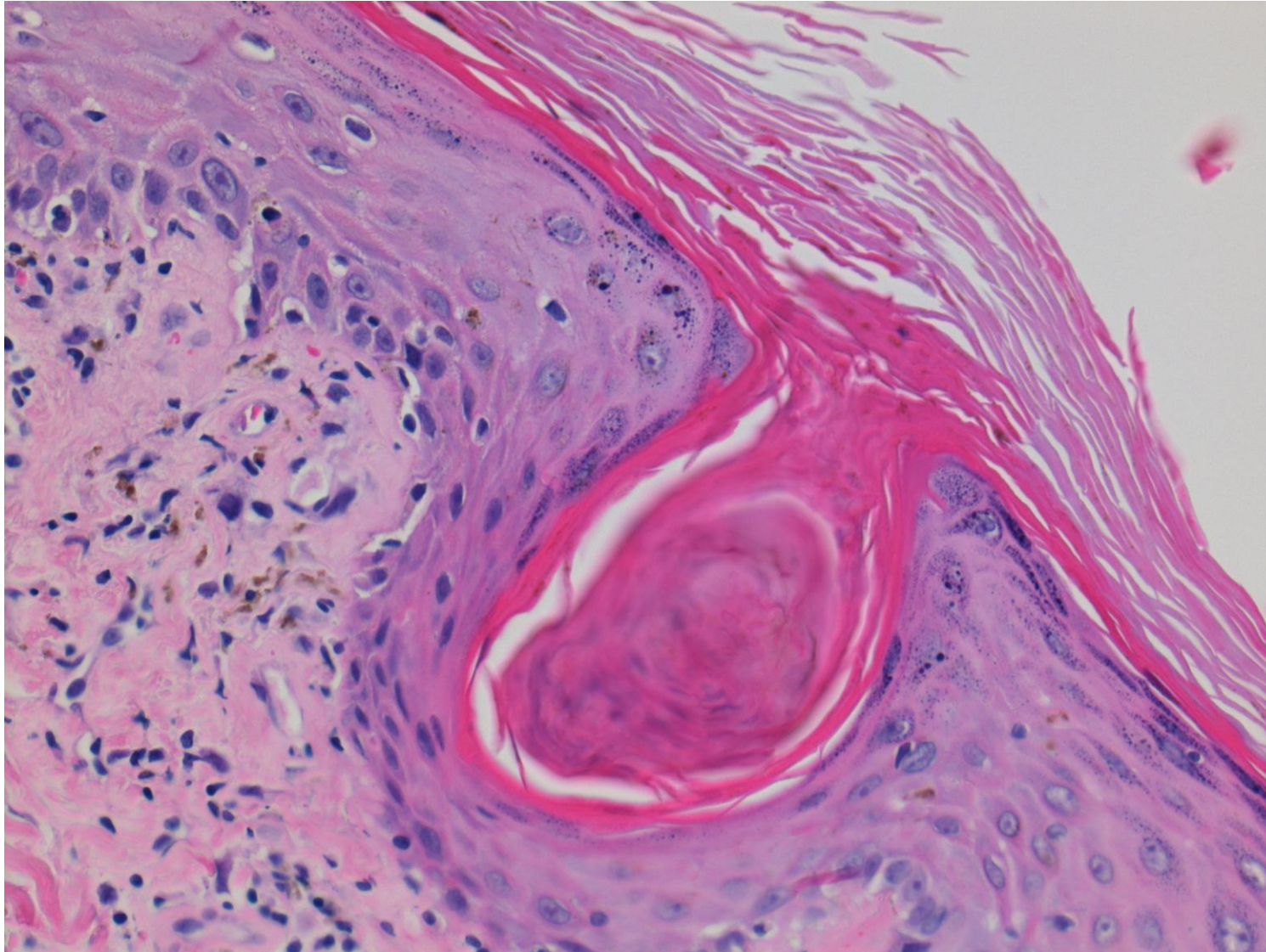
Erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Biopsy reveals hyperkeratosis and chronic dermal inflammation (H&E-1).



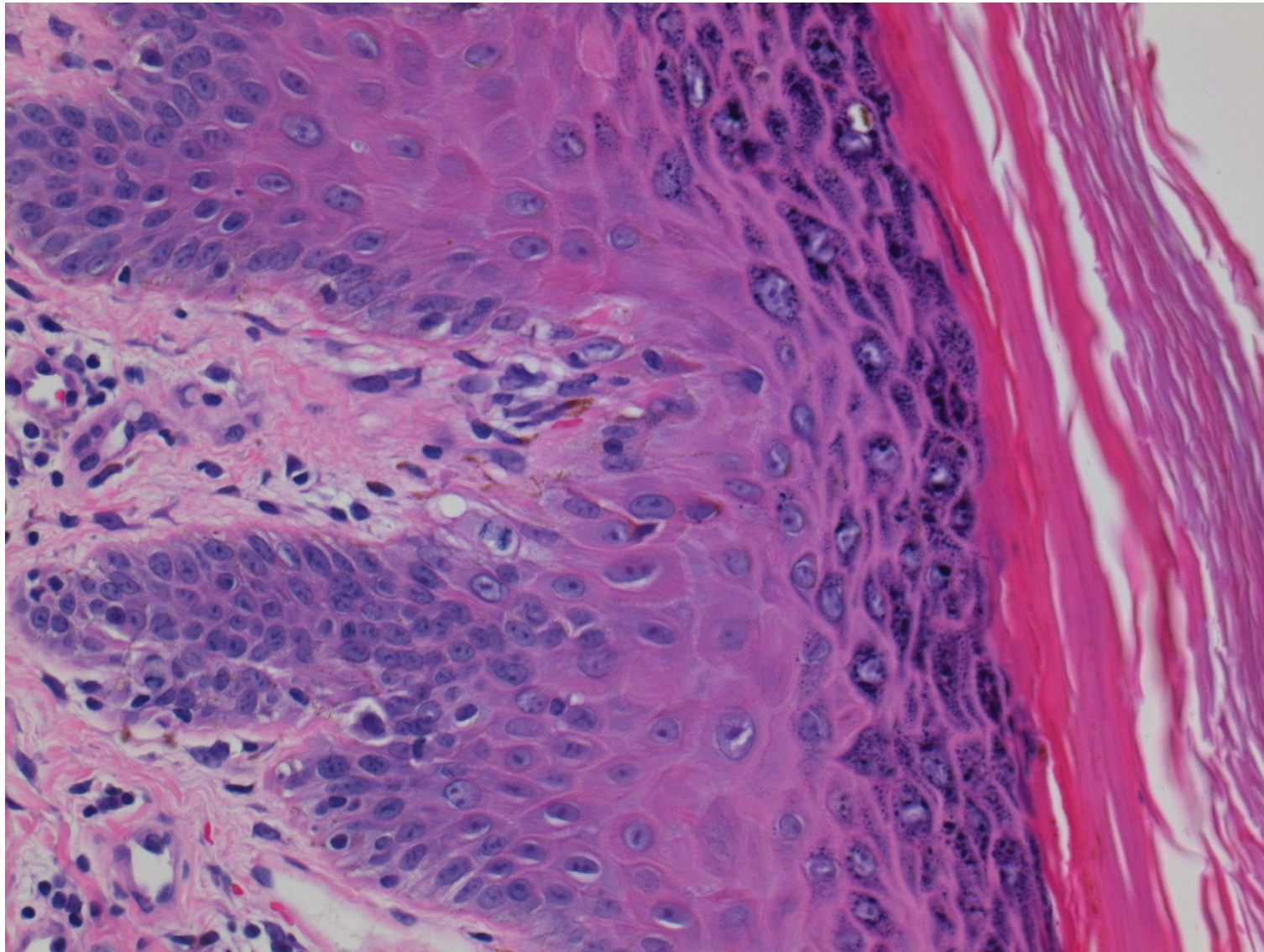
Erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Biopsy reveals hyperkeratosis, basal liquefaction and chronic dermal inflammation (H&E-2).



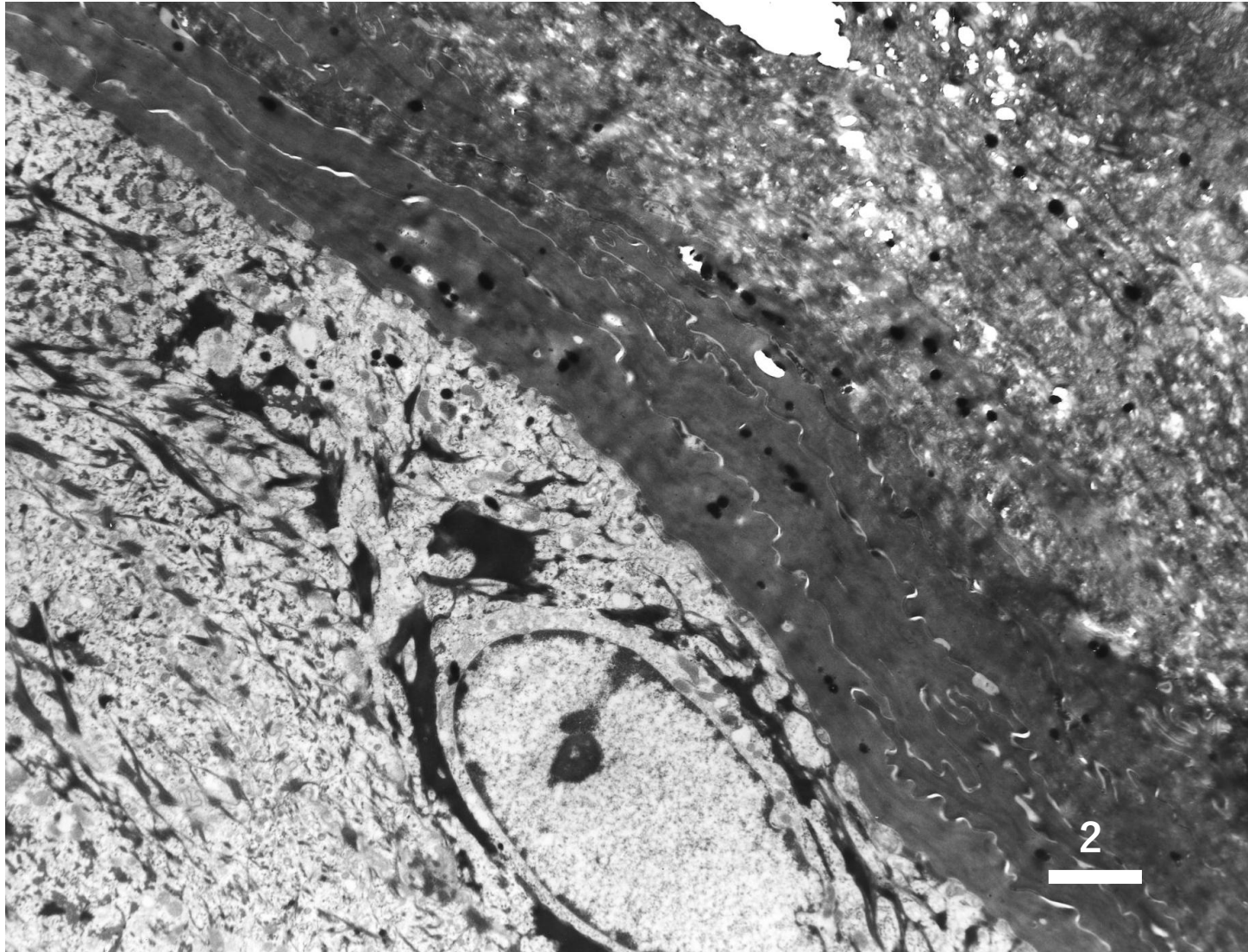
Erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Biopsy reveals hyperkeratosis, basal liquefaction with melanin incontinence and chronic dermal inflammation (H&E-3).



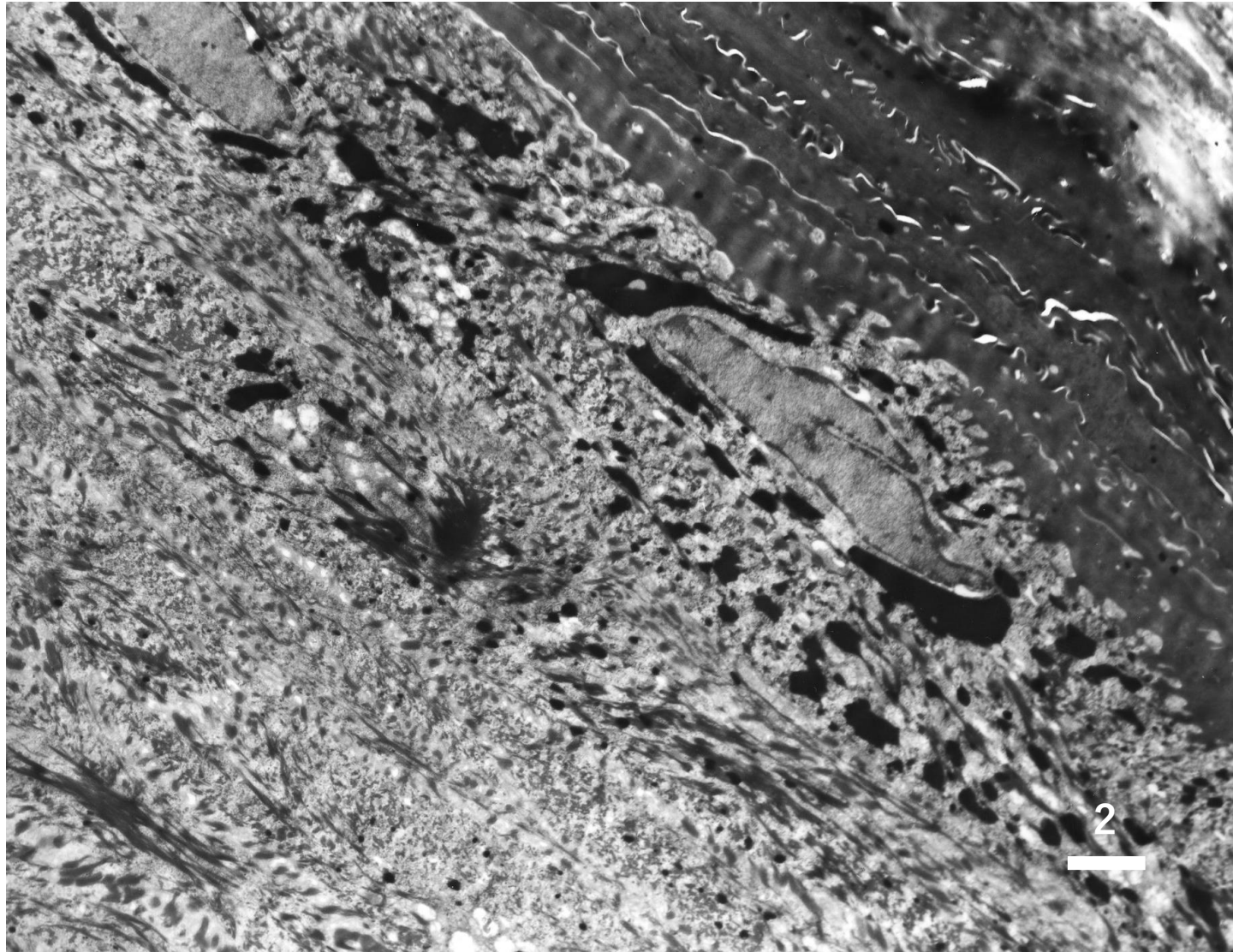
Erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Biopsy reveals hyperkeratosis, keratotic plugging, basal liquefaction and chronic dermal inflammation (H&E-4).



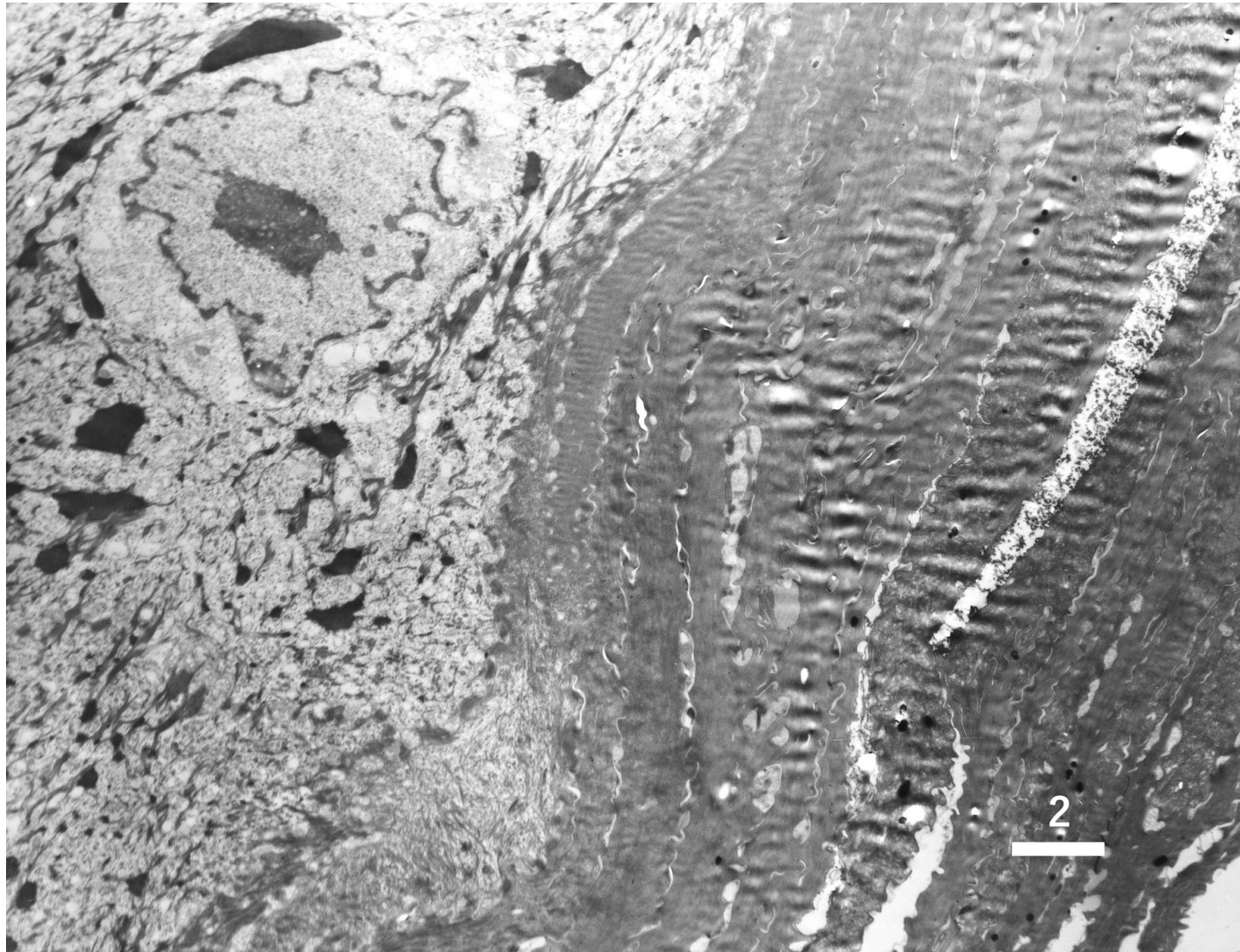
Erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Biopsy reveals hyperkeratosis, hypergranulosis, acanthosis and chronic dermal inflammation (H&E-5).



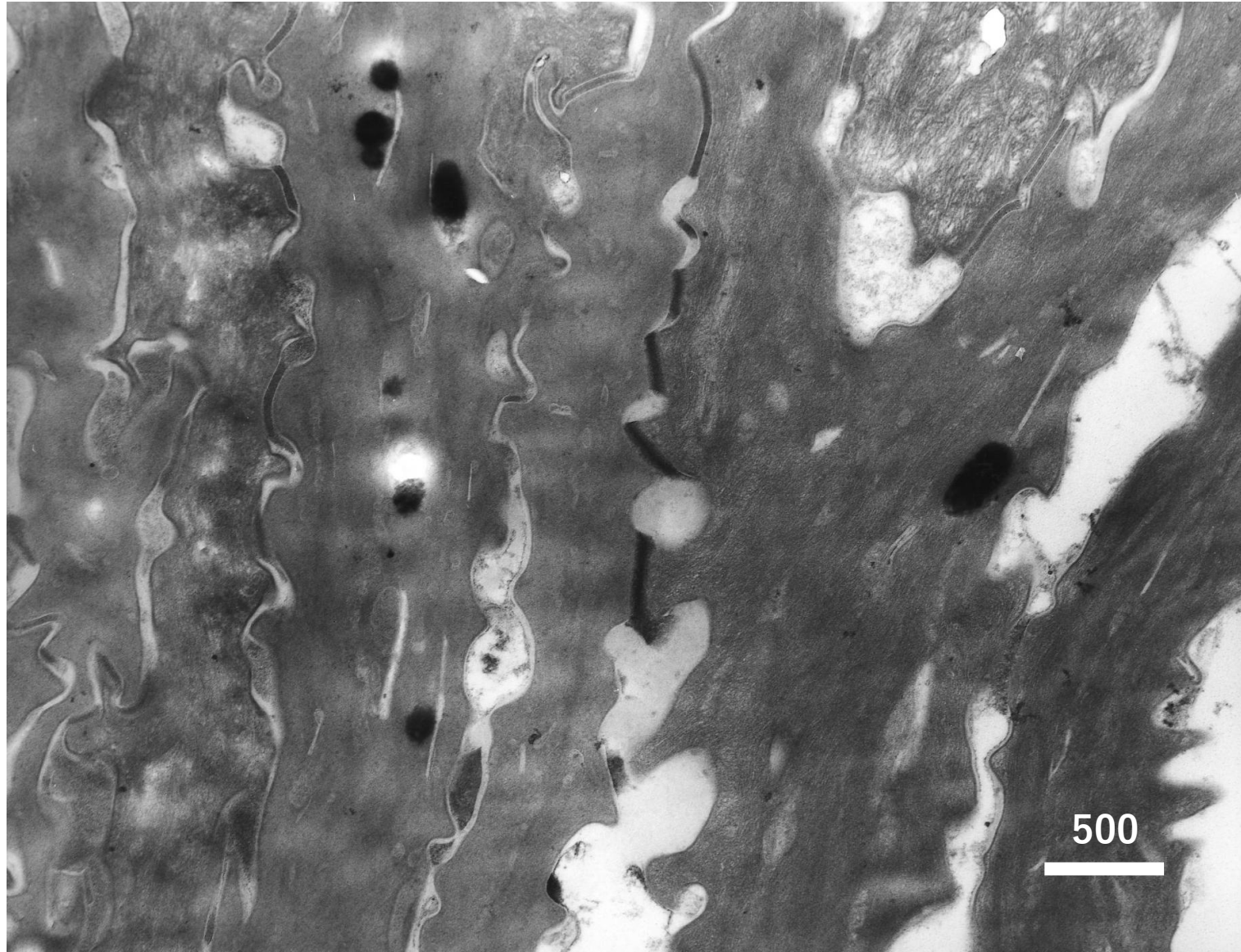
Ultrastructure of erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Hyperkeratosis and hypergranulosis with keratohyaline granules are observed (TEM-1).



Ultrastructure of erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Hyperkeratosis and hypergranulosis with keratohyaline granules are observed (TEM-2).



Ultrastructure of erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. Hyperkeratosis and hypergranulosis with keratohyaline granules are observed (TEM-3).



Ultrastructure of erythrokeratoderma seen on the thigh of a 12 y-o boy. Demarcated scaling plaques with erythema and pigmentation appeared on the right thigh one year ago. The hyperkeratotic layer shows well-developed desmosomal junctions (TEM-4).