

Inflammatory linear verrucous epidermal nevus

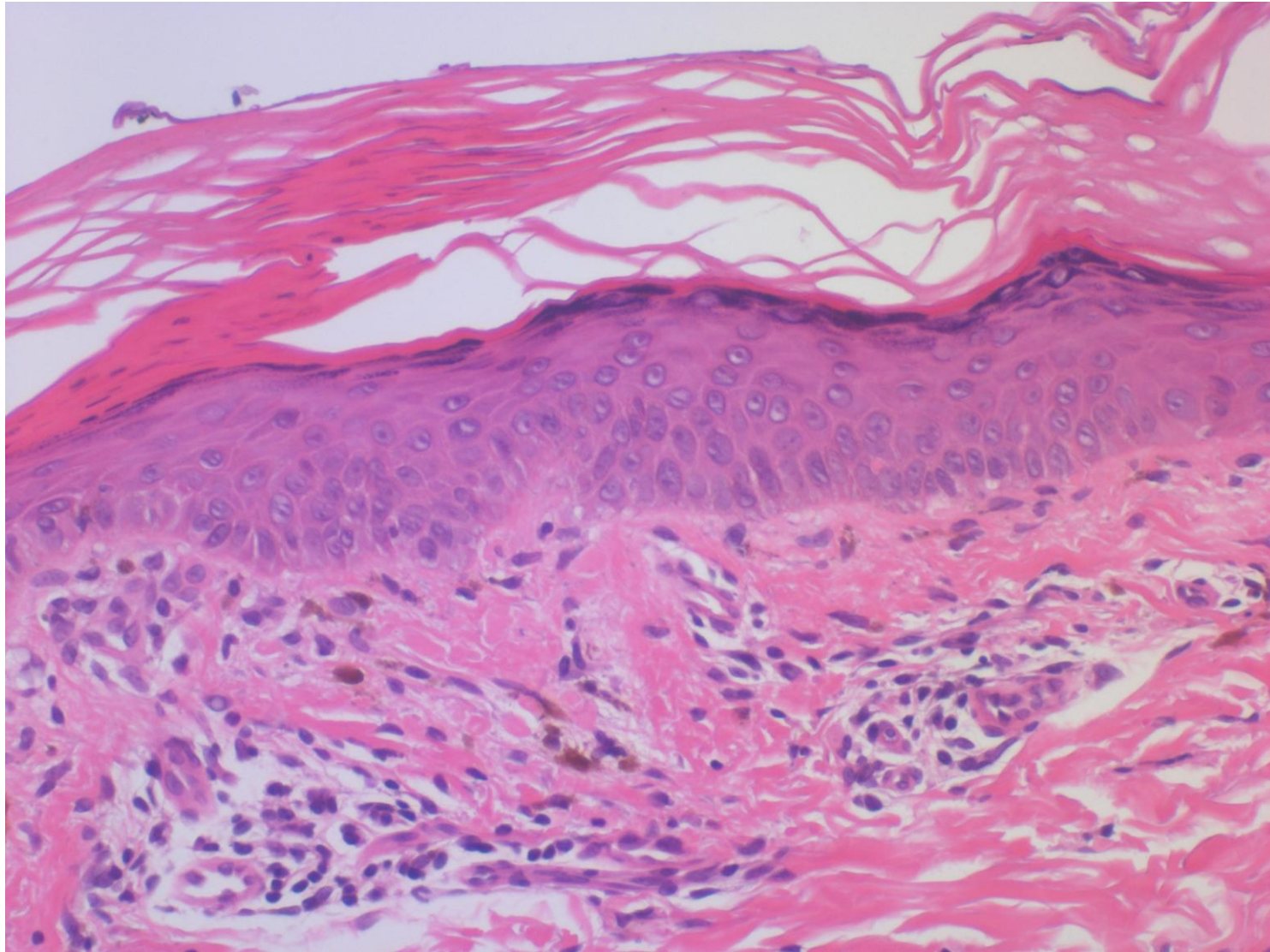
Inflammatory linear verrucous epidermal nevus (ILVEN) presents as multiple, discrete, red papules that tend to coalesce into linear plaques. The plaques can be slightly warty (psoriaform) or scaly (eczema-like). The linear lesions characteristically follow the lines of Blaschko. The lesions affect unilaterally, usually the left side of pediatric patients. It usually appears on an extremity in infancy or childhood. The features of ILVEN include 1) early age of onset, 2) predominance in females (4:1 female-male ratio), 3) frequent involvement of the left leg, 4) pruritus, 5) marked refractoriness to therapy, and 6) a distinctive psoriasiform and inflammatory histologic appearance. ILVEN is caused by somatic mutations resulting in genetic mosaicism. Most cases are sporadic, but familial cases with autosomal dominant trait have been recorded. As seen in psoriasis, interleukins 1 and 6, tumor necrosis factor α and intercellular adhesion molecule-1 are upregulated to provoke inflammatory reactions. Microscopically, the plaques are hyperkeratotic and acanthotic with elongation of the rete ridge, resembling linear psoriasis. Slightly raised parakeratotic areas with hypogranulosis and slightly depressed orthokeratotic areas with hypergranulosis are alternately distributed. The orthokeratotic area shows a basket-weave-pattern. The dermis shows scattered chronic inflammatory infiltrate.

The present case (a 7 y-o girl) presented with a linear pigmented plaque on the left lower leg along the Blaschko's line. The lesion was seen at birth. The inflammatory change became inactive along with ages.

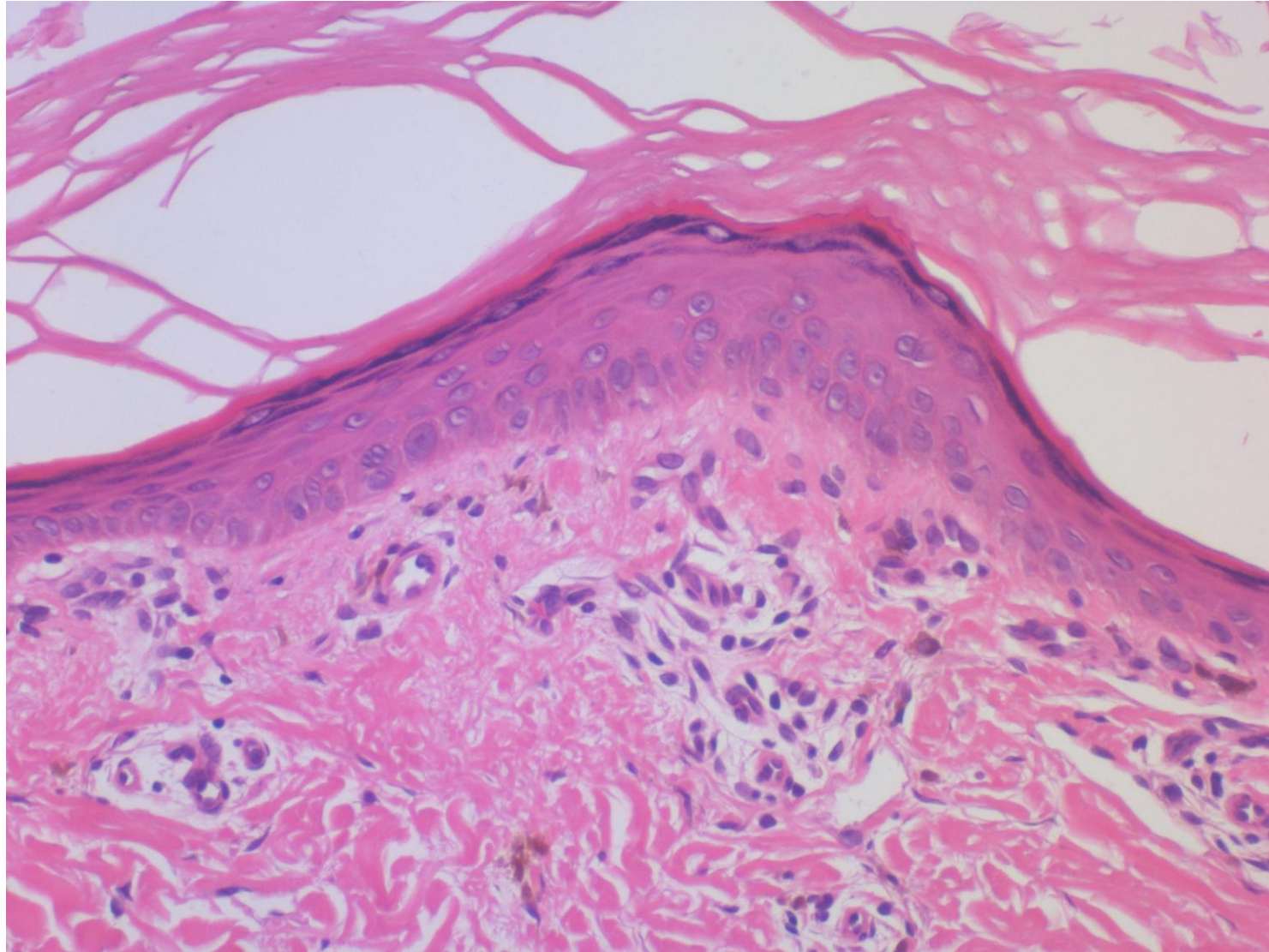
Ref.: Atzmony L, et al. Inflammatory linear verrucous epidermal nevus (ILVEN) encompasses a spectrum of inflammatory mosaic disorders. *Pediatr Dermatol* 2022; 39(6): 903-907. doi: 10.1111/pde.15094



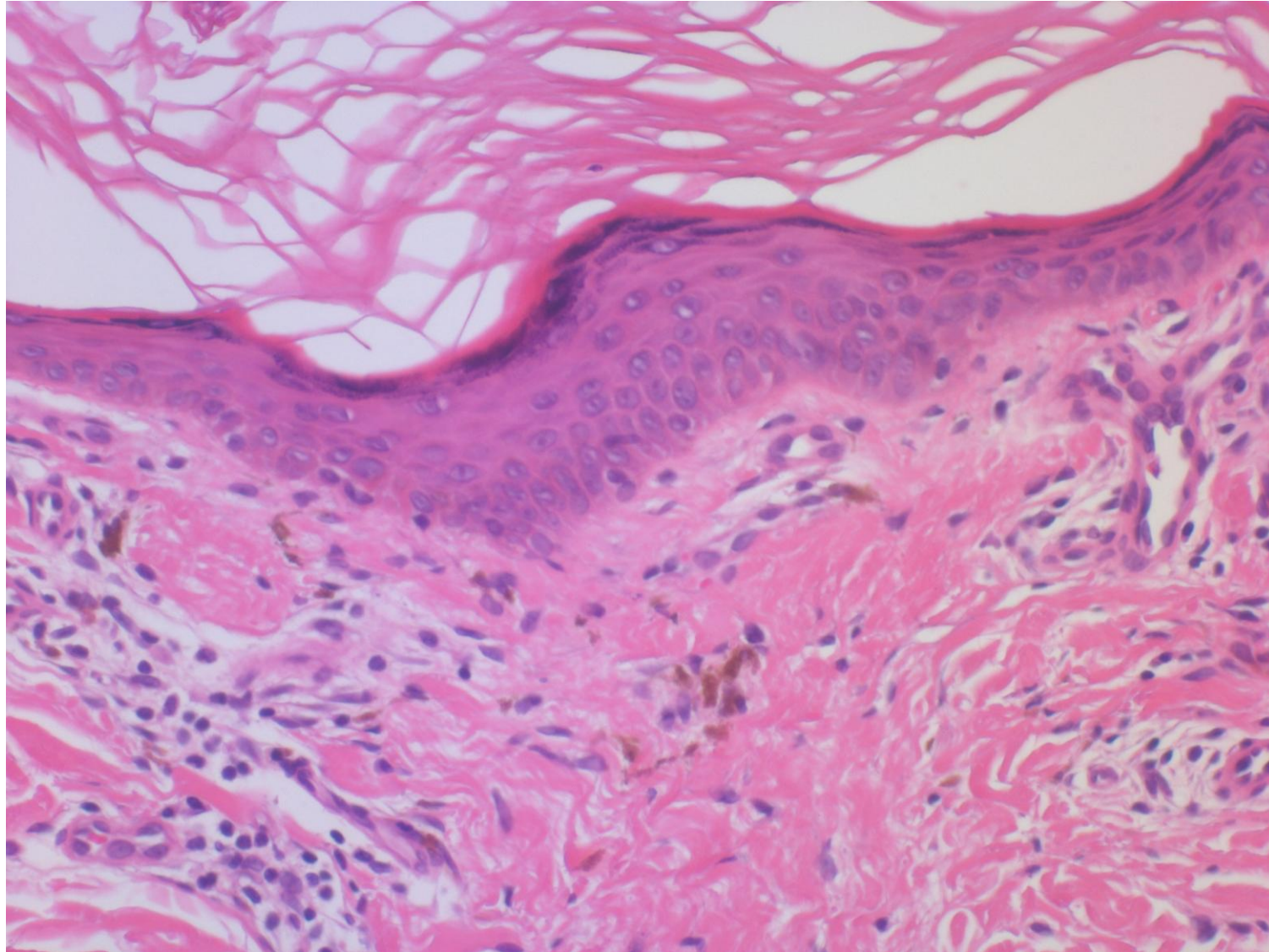
Inflammatory linear verrucous epidermal nevus on the left lower leg. Gross appearance: borrowed from ar.inspiredpencil.com



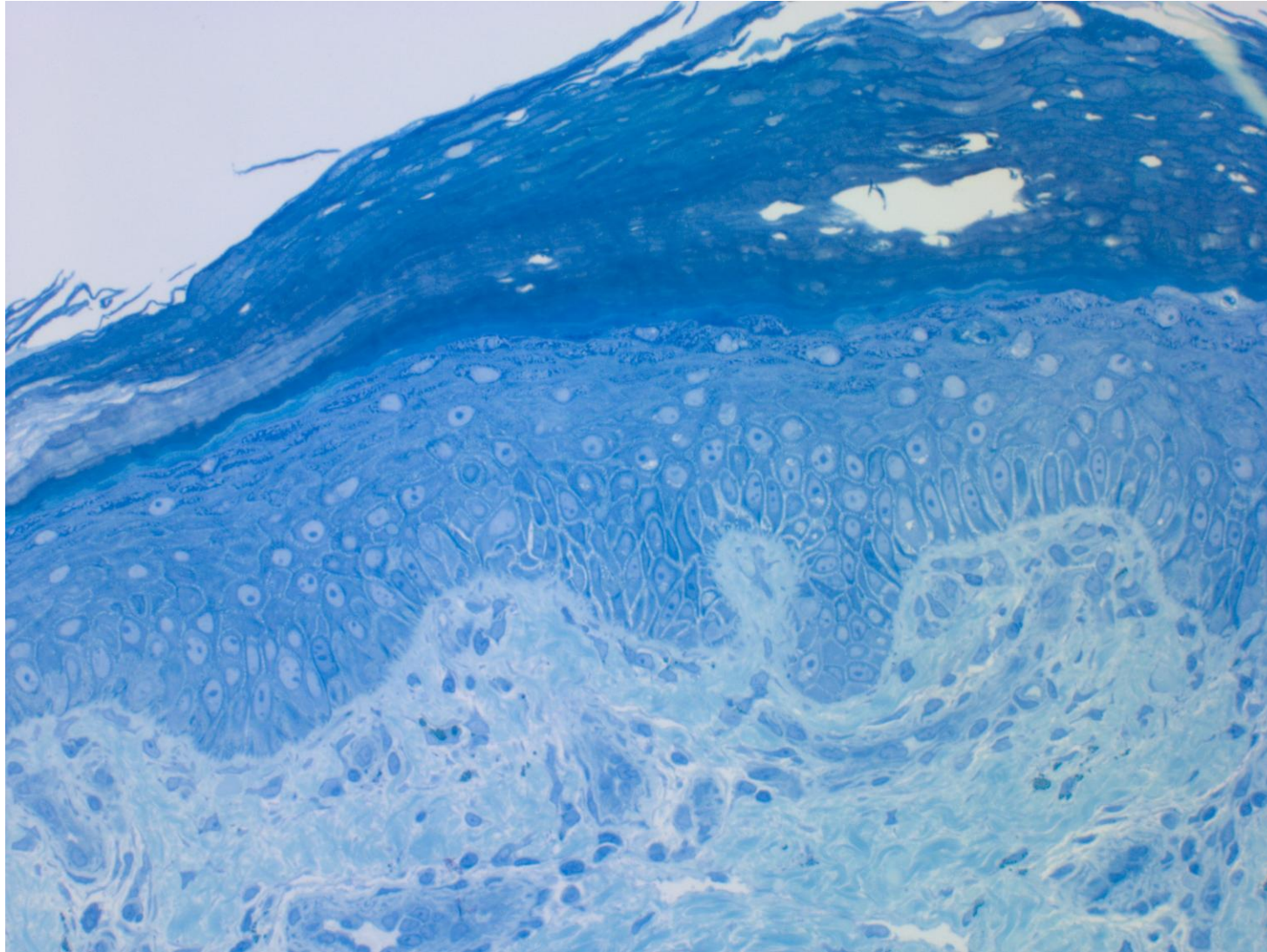
Inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. Mild acanthosis, hyperkeratosis with hypergranulosis, parakeratosis with hypogranulosis and mild dermal mononuclear infiltration with scattered melanophages are noted (H&E-1).



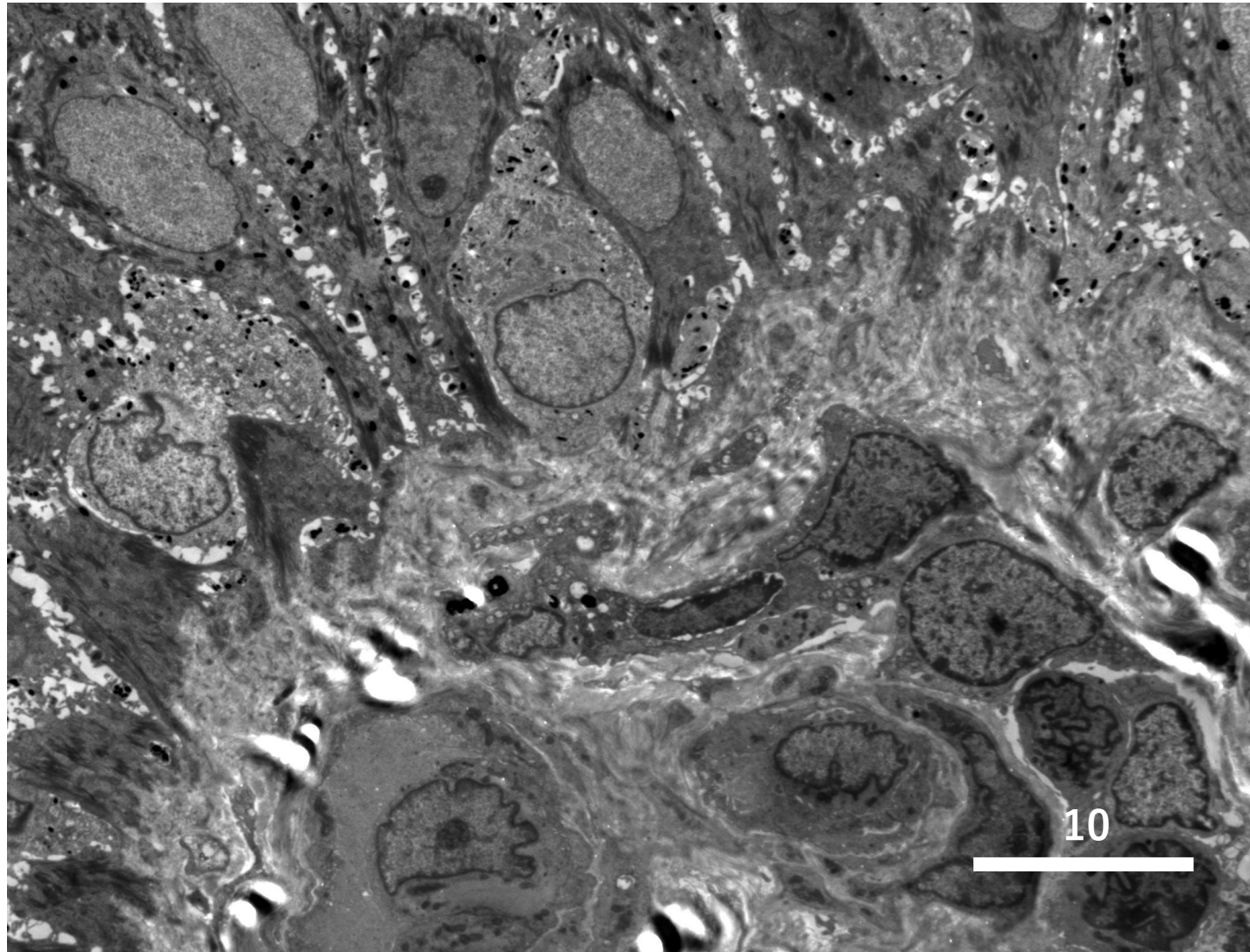
Inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. Mild acanthosis, hyperkeratosis with hypergranulosis and mild dermal mononuclear infiltration with mild angiectasia are noted (H&E-2).



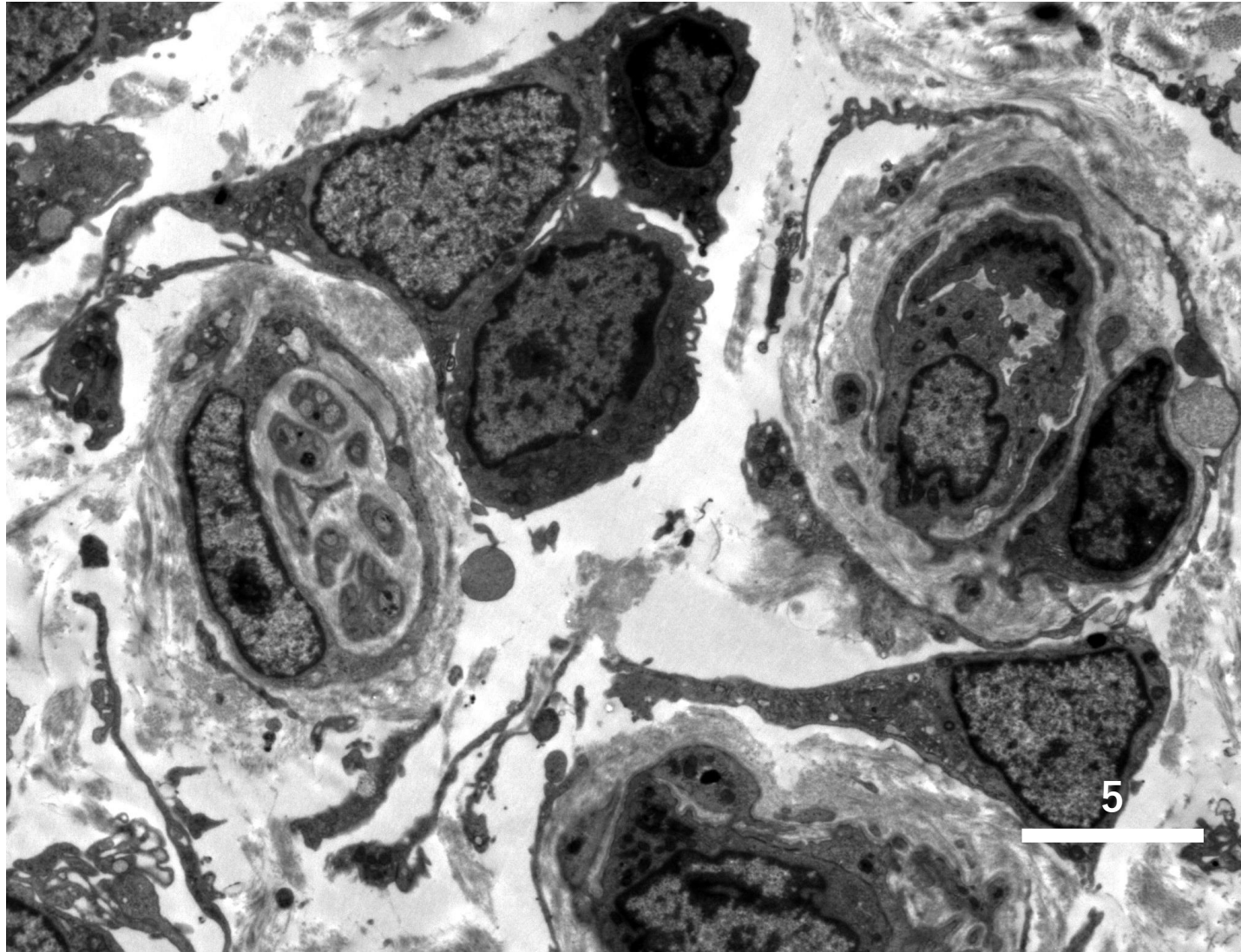
Inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. Mild acanthosis, hyperkeratosis with hypergranulosis, and mild dermal mononuclear infiltration with scattered melanophages and angiectasia are noted (H&E-3).



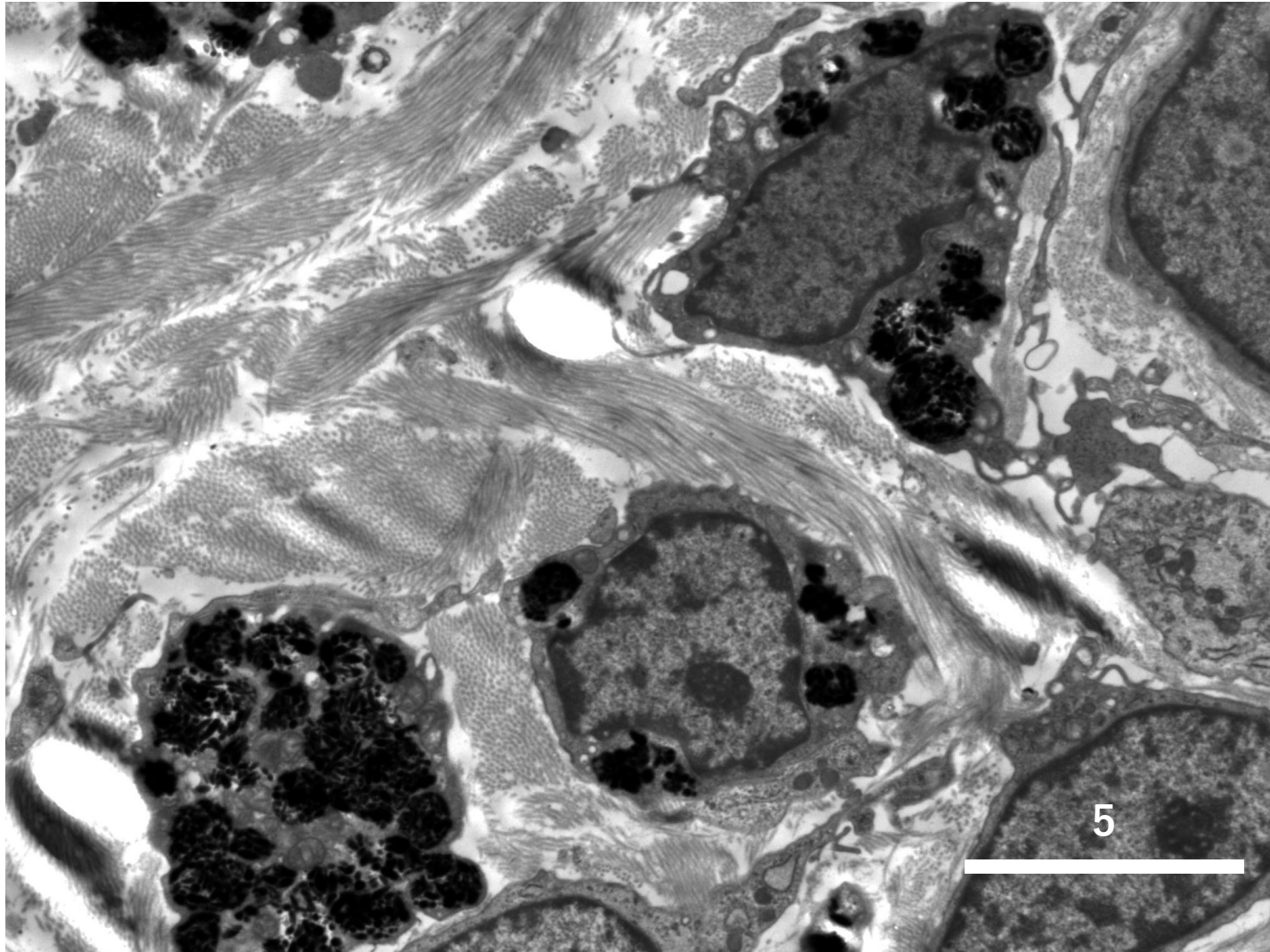
Inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. Mild acanthosis, hyperkeratosis with hypergranulosis and mild dermal mononuclear infiltration are noted (Toluidine blue in thick section for EM study).



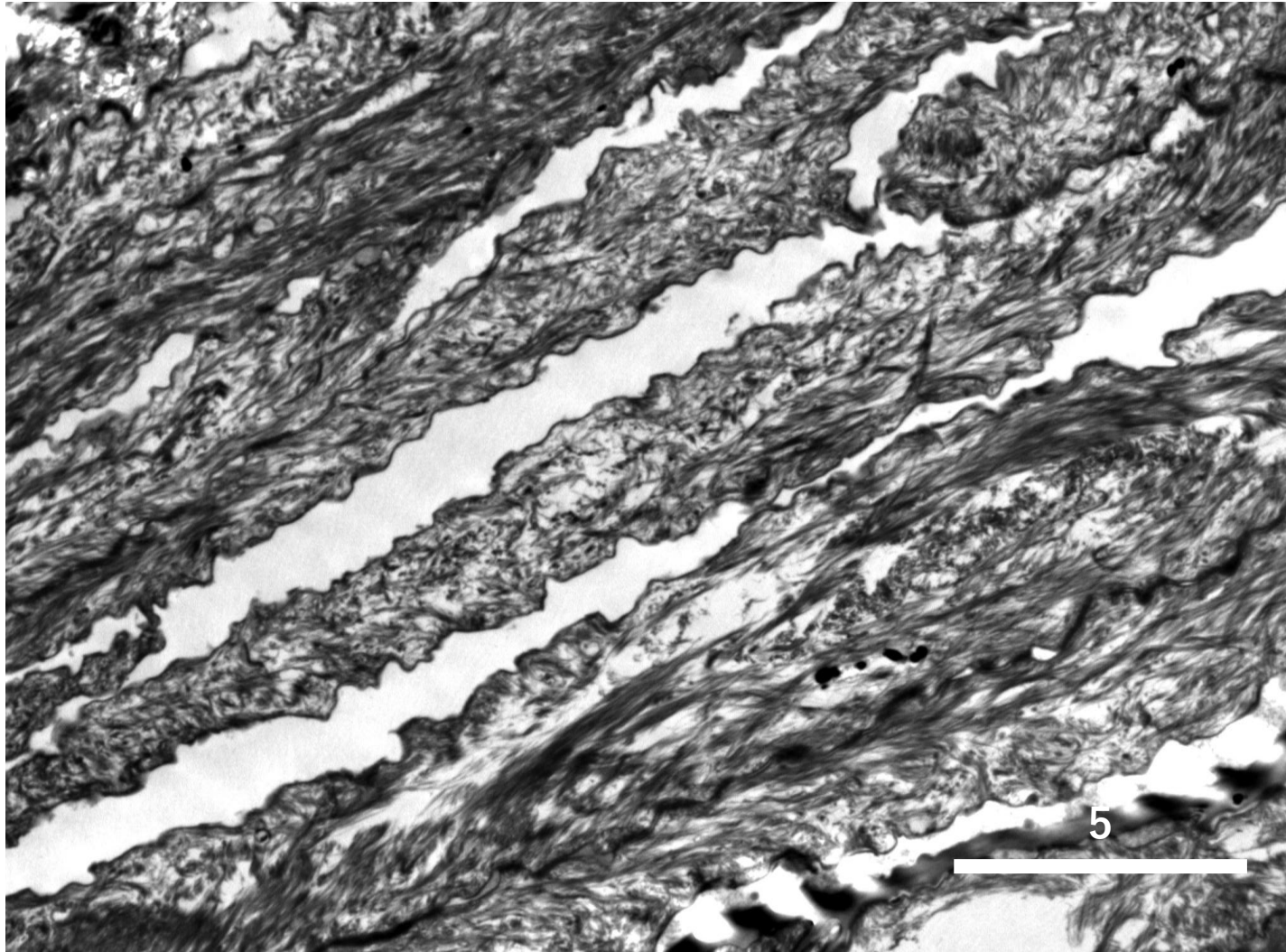
Ultrastructure of inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. In the upper dermis, mild infiltration of lymphocytes and macrophages is noted. Melanocytes are seen in the basal layer of the epidermis (TEM-1).



Ultrastructure of inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. In the upper dermis, mild infiltration of lymphocytes and macrophages is noted. Capillary vessels are increased in number (TEM-2).



Ultrastructure of inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. In the upper dermis, macrophages phagocytizing melanin pigments are noted (TEM-3).



Ultrastructure of inflammatory linear verrucous epidermal nevus on the left lower leg of a 7 y-o girl. The thickened cornified layer anucleated keratinocytes are rich in tonofilaments (TEM-4).