Vascular Anomalies

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31.1 Definition

Vascular anomalies are a heterogeneous group of congenital disorders that are subcategorized based on their histology, biological behavior, and clinical presentation into vascular tumors, e.g., hemangiomas, and vascular malformations [1].

31.2 Epidemiology

Vascular anomalies are frequently seen in the pediatric age occurring in about 4.5% of all children [2, 3].

31.2.1 Hemangiomas

Hemangiomas are the most common tumor occurring in infancy. In a recent retrospective study, they occurred in about 1.97 per 100 person-years [4] although their incidence was previously estimated up to 10% [5]. Hemangiomas have been related to decrease in gestational age at birth and to a lower birth weight, along with an increase in pregnancy complications [4].

31.2.2 Vascular Malformations

Vascular malformations are very heterogeneous and occur in approximately 0.3% of the general population, excluding the salmon patch which shows a prevalence up to 50% [6, 7].

31.3 Etiology

31.3.1 Hemangiomas

Hemangiomas are localized or regional areas of abnormal vascular development and proliferation. Their pathogenesis remains unclear, but several factors may take part in the development of hemangiomas, including endothelial cells arising from disrupted placental tissue embedded in fetal soft tissues during gestation or birth, endothelial progenitor and stem cells in the circulation, and cytokines and genetic alterations in growth factor receptors [5].

31.3.2 Vascular Malformations

Vascular malformations are localized defects of vascular morphogenesis and structure without evidence of endothelial cell proliferation, involving capillaries, veins, lymphatic vessels, arteries, or a combination of these. Dysfunction in signaling processes that regulate migration, differentiation, maturation, adhesion, and survival of the cells of vascular walls is thought to play a pathogenic role.

31.4 Clinical Features

31.4.1 Hemangiomas

Most hemangiomas have a typical presentation and growth pattern developing in three clearly defined stages: proliferation, quiescence, and involution. They can be fully developed

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at birth (congenital hemangiomas) or present shortly after birth and grow rapidly (infantile hemangiomas). Infantile hemangiomas are classified as focal, multifocal, segmental, and indeterminate depending on their morphology, extent, or distribution and as superficial, deep, and mixed depending on their location in the dermis and/or hypodermis. Superficial lesions appear as early telangiectatic patches that after a few weeks begin to turn red and swell reaching variable sizes (Figs. 31.1a and 31.2a). Sometimes, they can be destructive and life-threatening. During proliferation, ischemia, necrosis, ulceration, bleeding, and secondary infection may occur. Deep lesions appear as warm, ill-defined, light blue-purple masses with minimal or no overlying skin changes, sometimes making the diagnosis difficult. Mixed hemangiomas have both superficial and deep components.

31.4.2 Vascular Malformations

In contrast to hemangiomas, vascular malformations are present at birth and never completely regress. They are commonly classified depending on flow characteristics (high/low flow) and which type of anomalous vessel is involved in capillary malformation (i.e., salmon patches, port-wine stain), venous malformation, lymphatic malformation (i.e., lymphangioma circumscriptum), and arteriovenous malformation.

The most common types of capillary malformations are salmon patches. They usually appear as irregular, pinkish, or reddish macules or patches located on the midface (forehead, glabella, eyelids, nose, philtrum) and occiput and may lighten and disappear with time. Another variant of capillary malformations is the port-wine stain, which usually appears as red-violaceous patches and, over years, may darken in color and thicken (Figs. 31.3a and 31.4a). The involvement of maxillary and/or ophthalmic dermatome may be signs of a complex syndrome (Sturge-Weber syndrome).

31.5 Differential Diagnosis

31.5.1 Hemangiomas

Hemangiomas may sometimes mimic many other types of vascular anomalies such as pyogenic granuloma, tufted angioma, infantile hemangiopericytoma, diffuse neonatal hemangiomatosis, as well as neuroblastoma, myofibromatosis, and lipoblastoma. Moreover, early lesions may sometimes be misdiagnosed as vascular malformations.

31.5.2 Vascular Malformations

The main differential diagnosis of vascular malformations is with hemangiomas.

31.6 Diagnosis

Most hemangiomas and vascular malformations can be diagnosed clinically based on patient history and lesion appearance. Histopathology and imaging techniques, such as magnetic resonance imaging and ultrasound, can be helpful to better evaluate deep lesions [8].

31.7 Dermatoscopy

31.7.1 Hemangiomas

Dermatoscopy reveals the presence of sharply demarcated and red round-oval structures resembling lacunae (Figs. 31.1b, c and 31.2b). A polymorphous vascular pattern consisting of red globular/circulated/comma-like/wavy/dilated/linear vessels has been described. Blue-whitish septa usually separate the lacunae from each other. If any of these lacunae develops a thrombus, they will appear black [9–12].

31.7.2 Vascular Malformations

Few studies reported dermatoscopic features of vascular malformation. Lesions may reveal three dermatoscopic patterns: *type 1* or superficial, characterized by a vascular structure of red-dotted, globular vessels (Fig. 31.3b); *type 2* or deep, revealing red dilated, linear vessels forming irregular networks (Fig. 31.4b, c); and *type 3*, showing round/sacular/glomerular structures [13, 14]. Nonvascular structures have also been described, including a perifollicular pale halo and a gray-whitish veil [15, 16]. Some authors have highlighted the importance of dermatoscopy in the prognostic evaluation of treatment response. Superficial lesions have been related to laser therapy to a better response than deeply located vessels [12, 15–17].

31.8 Histopathological Correlation

31.8.1 Hemangiomas

The vascular pattern seen at dermatoscopy correlates with ectatic vascular structures in the papillary and subpapillary dermis.

31.8.2 Vascular Malformations

These lesions are characterized by the presence of ectatic vessels at different depth. The red round globular vessels seen at dermatoscopy are related to superficial ectatic vertical papillary vessels, whereas the red linear vessels correspond to deeper dilated horizontal subpapillary capillaries. Enlargement of the horizontal plexus over time correlates to the roundish vascular structures seen at dermatoscopy.

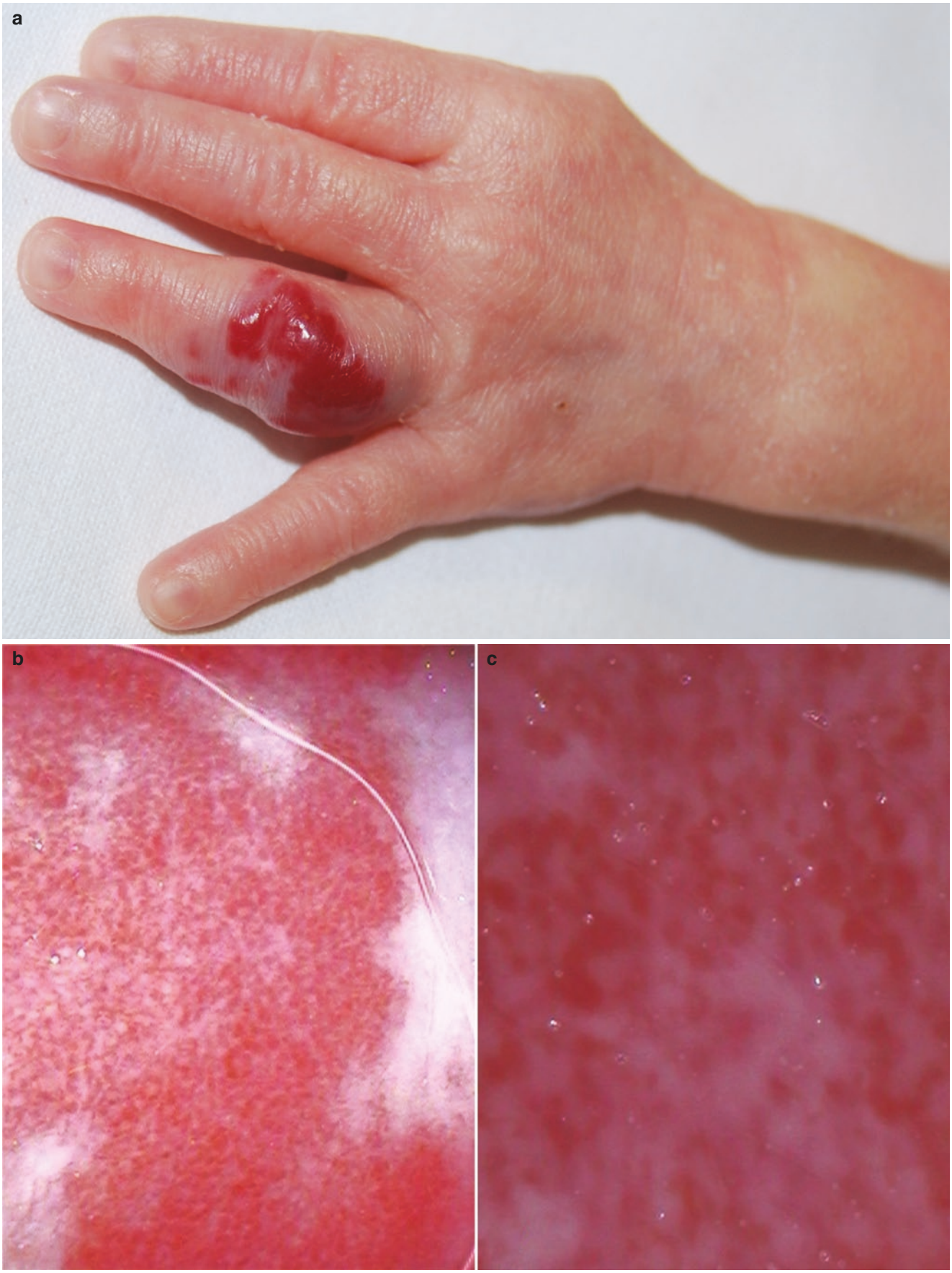

Fig. 31.1 (**a**) Hemangioma. Reddish tumor involving the forth finger in a 1-month-old girl. (**b**) Dermatoscopy (×10): diffuse, red round-oval structures resembling lacunae with interposed whitish septa. (**c**) High-magnification dermatoscopy (×100): enhanced visualization of dilated vessels

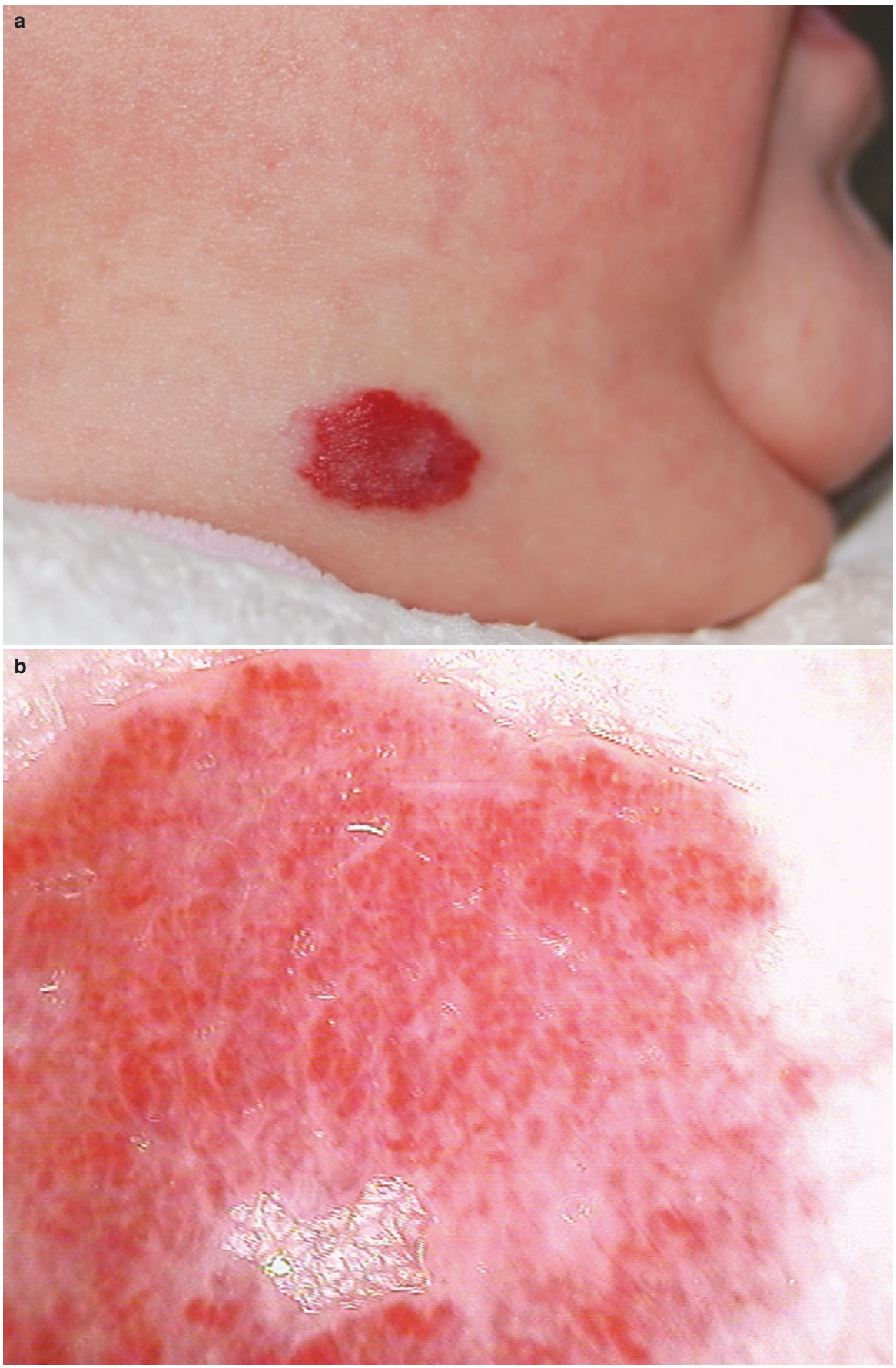

Fig. 31.2 (**a**) Hemangioma. Reddish plaque on the face in a 1-year-old girl. (**b**) Dermatoscopy (×10): red round-oval structures resembling lacunae and whitish septa

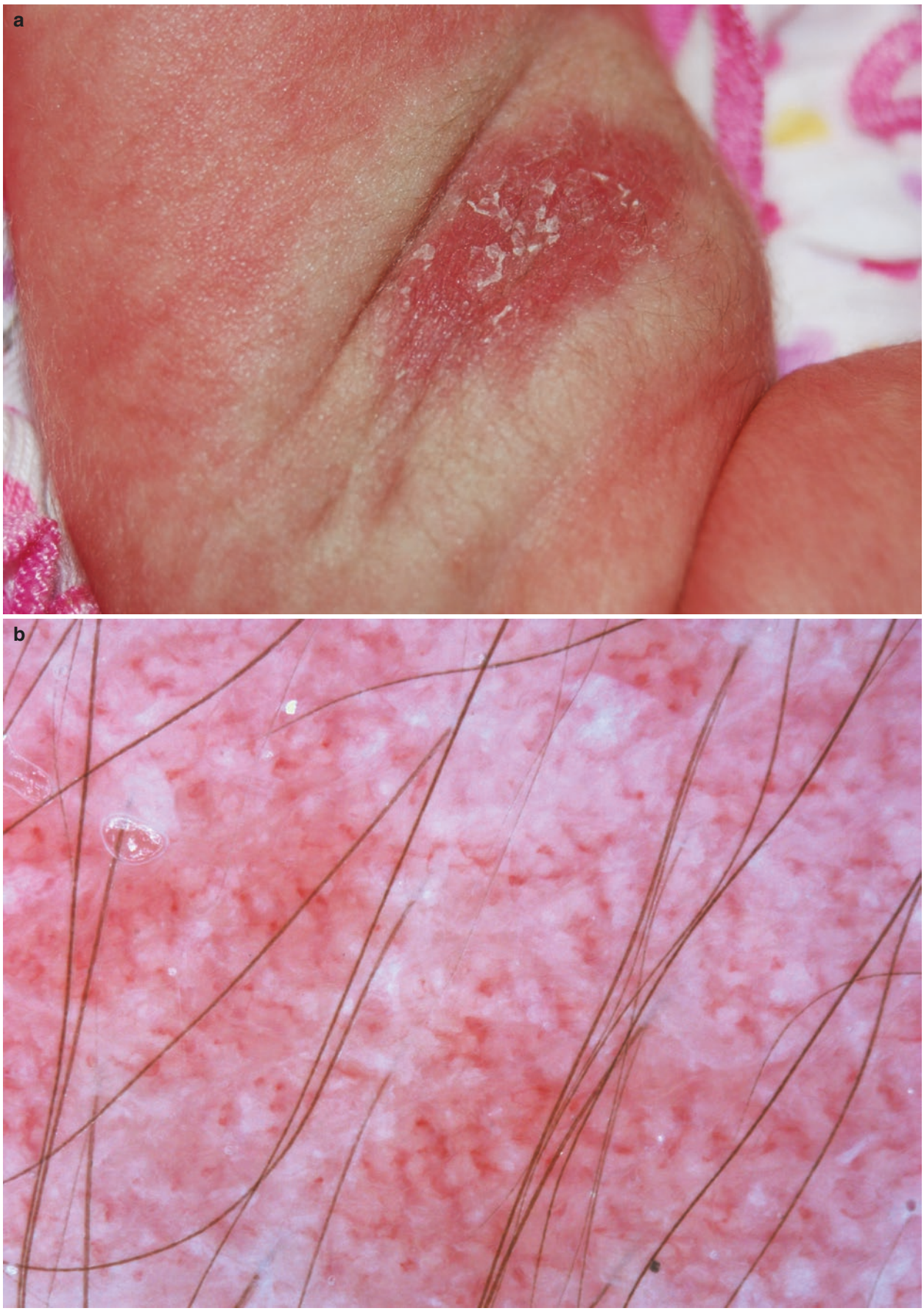

Fig. 31.3 (**a**) Vascular malformation. Reddish macule of the arm in a 2-month-old girl. (**b**) High-magnification dermatoscopy (×100): red-dotted and globular vessels (*type 1* pattern)

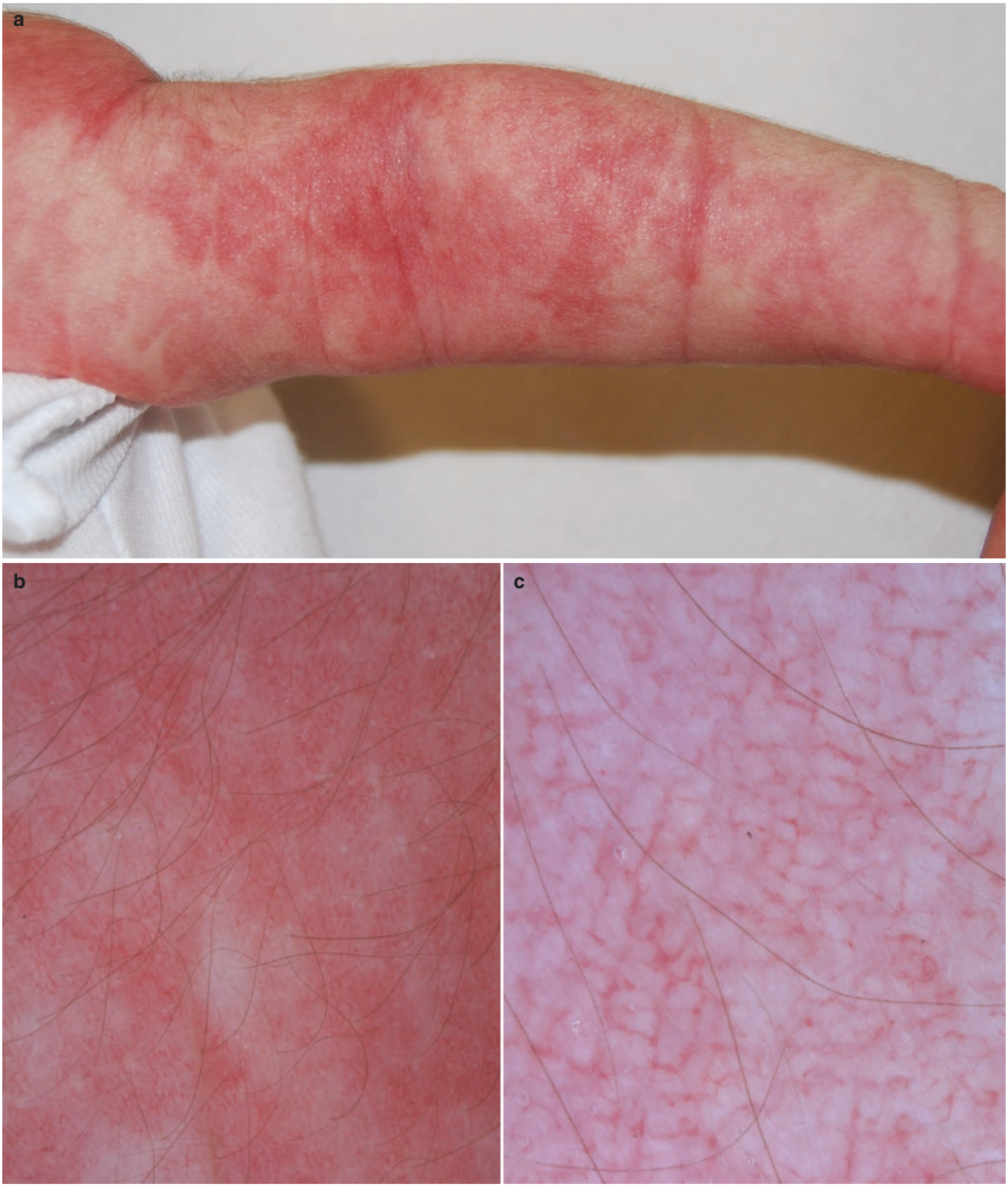

Fig. 31.4 (**a**) Vascular malformation. Reddish macule involving almost the entire arm in a 3-month-old boy. (**b**) Dermatoscopy (×10): diffuse dilated, linear vessels and some perifollicular pale halos (*type 2* pattern). (**c**) High-magnification dermatoscopy (×100): linear vessels forming an irregular network

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