Lymphatic Malformations

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33.1 Definition

Superficial lymphatic malformations (LMs) include a group of vascular malformations of the lymphatic network involving the skin and the subcutaneous tissue. A recent reclassification of International Society for the Study of Vascular Anomalies (ISSVA) in 2014 divides common cystic lymphatic malformation in microcysts, macrocysts, and mixed forms according to the vessels size [1].

33.2 Epidemiology

Microcystic LM accounts for 4% of all vascular tumors, representing the 25% of all benign vascular tumors in children. About 50% of them are observed at birth or manifests by 2 years of age.

33.3 Etiology

Anomalous lymphatic cisterns grow apart from the regular vascular network, probably arising from a primitive lymphatic sac.

33.4 Clinical Features

Microcystic LM clinically presents as a cluster of translucent and clear vesicles (compared to frog spawn) that tend to increase in number and size (Fig. 33.1a). Such lesions may turn purplish or black colored as a result of blood leakage or frank hemorrhage (Figs. 33.2a and 33.3a). The most involved sites are the extremities, trunk, axillae, and tongue.

Macrocystic LM manifests as palpable tender subcutaneous masses with a superimposed thickened skin. Superinfection is not uncommon [1].

33.5 Differential Diagnosis

Differential diagnosis mainly includes angiokeratoma, warts, molluscum contagiosum, epidermal naevi, and herpes zoster.

33.6 Diagnosis

Diagnosis is usually made clinically although confirmation by ultrasound or magnetic resonance imaging can be necessary in order to evaluate the depth and extension of the lesion.

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33.7 Dermatoscopy

Dermatoscopy reveals a whitish or yellowish lacunar pattern (Fig. 33.1b). Frequently, based on the possible presence of blood, scattered reddish areas within the yellowish lacunae (Fig. 33.2b) or sole reddish lacunae (Fig. 33.3b) may be observed. In some cases, the variable quantity of blood content within the lacunae generates a wide spectrum of color transition from yellow to purple, which is described as hypopyon-like pattern [2–5].

33.8 Histopathological Correlation

The yellowish lacunae histopathologically correspond to irregular lymphatic channels proliferating under a thinned epidermis (Fig. 33.4) [6]. The reddish areas are related to blood cells extravasation [5].

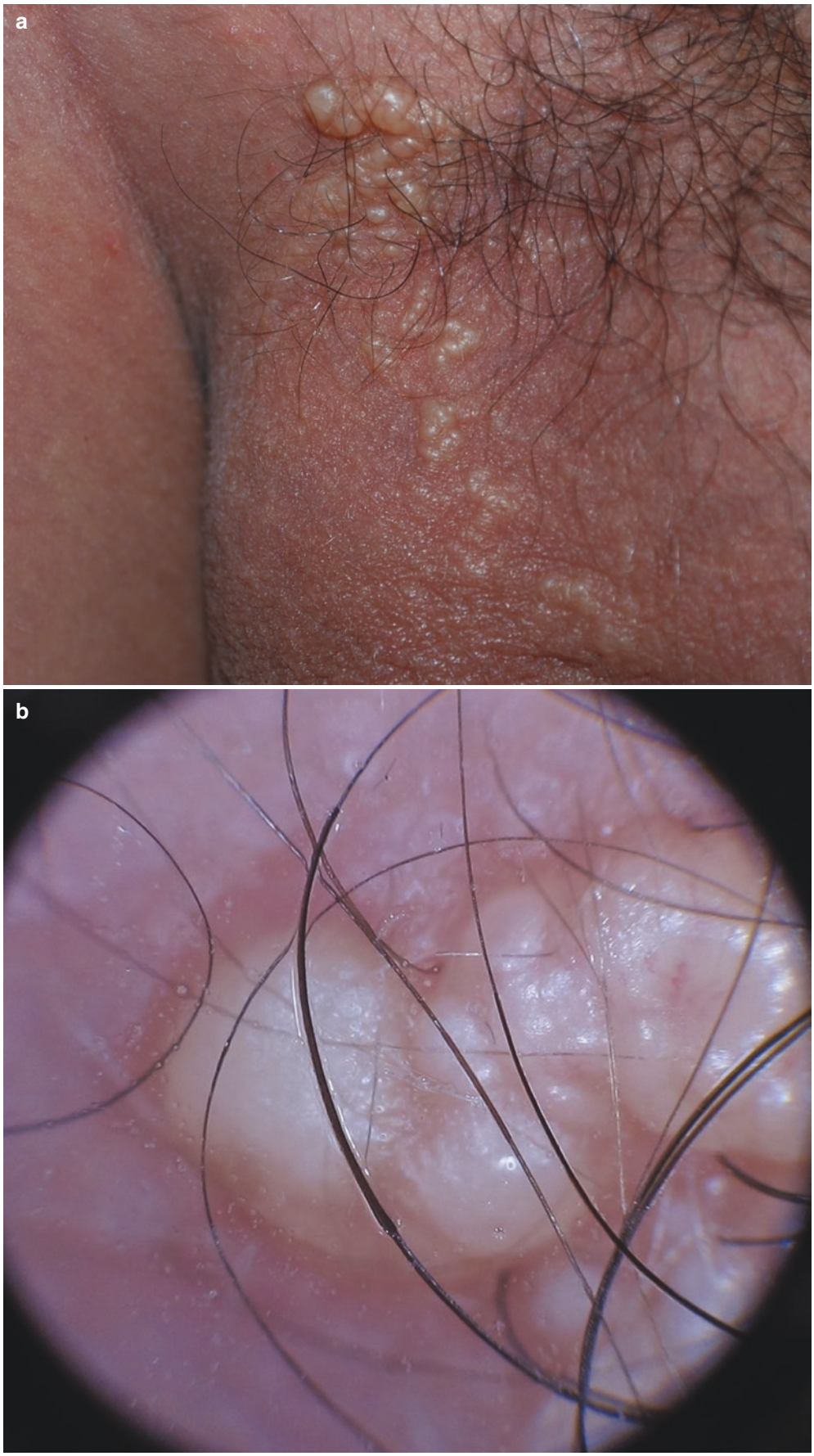

Fig. 33.1 (**a**) Microcystic lymphatic malformation in a boy. (**b**) Dermatoscopy (×10): whitish lacunae surrounded by pale septa

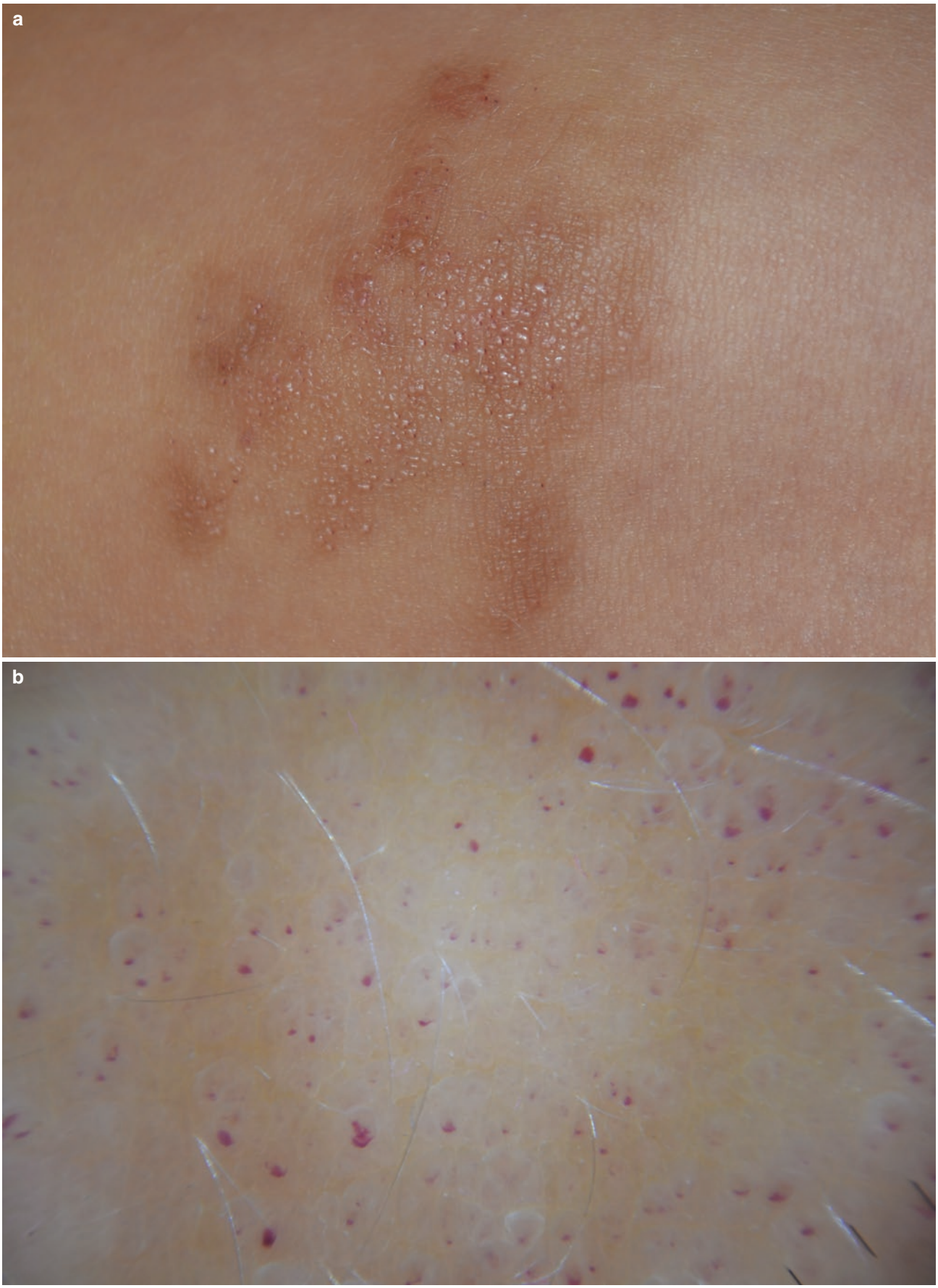

Fig. 33.2 (**a**) Microcystic lymphatic malformation in a 4-year-old girl. (**b**) Dermatoscopy (×10): yellowish lacunae associated with scattered red areas (Courtesy of Giuseppe Micali, MD)

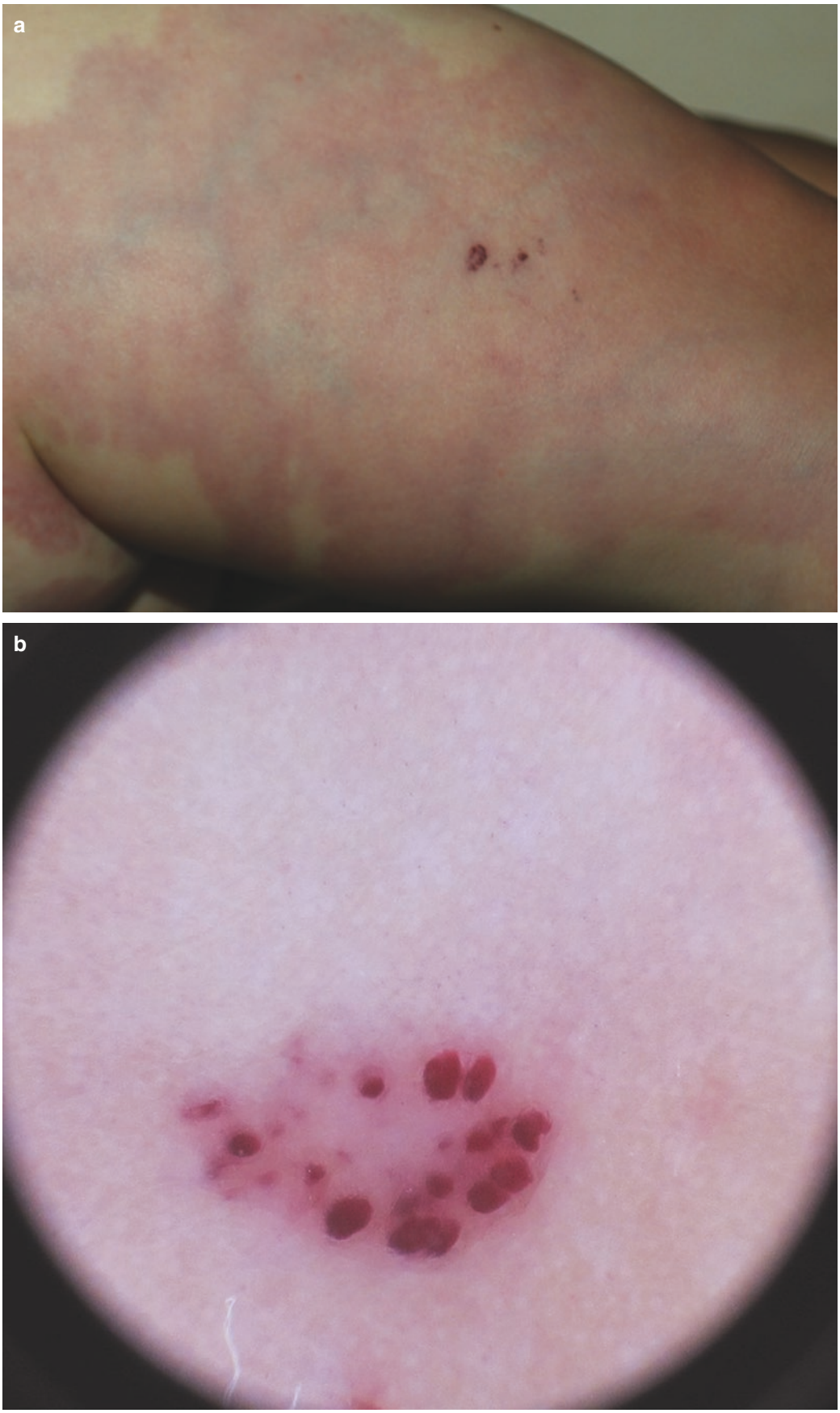

Fig. 33.3 (**a**) Combined vascular malformation: superficial lymphatic malformation in the context of a capillary malformation. (**b**) Dermatoscopy (×10): reddish lacunae

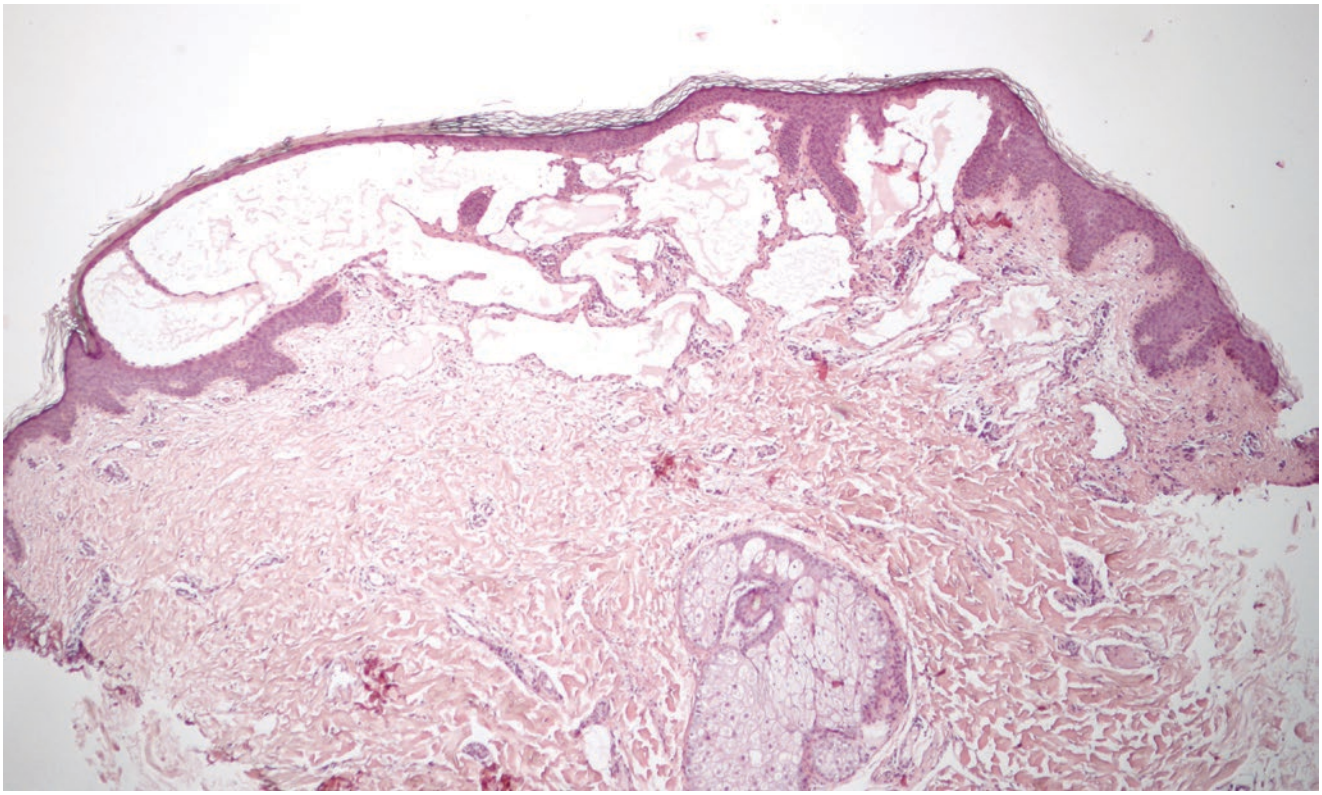

Fig. 33.4 Microcystic lymphatic malformation. Histopathology showing thin epidermis with underlying proliferating and irregular lymphatic channels [H&E staining; magnification, ×40]

References

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