# Cutaneous Vascular Diseases

# 14

Cheng Tan, Hong-Mei Jing, Li-Tao Zhang, Dong-Lai Ma, and Ru-Zhi Zhang

**Abstract**

Various cutaneous vascular diseases are presented in this chapter: eruptive recurrent pigmented telangiectasia, telangiectasia macularis eruptiva perstans, unilateral nevoid telangiectasia superimposed on the Bier spots, Mondor's disease, arteriosclerosis obliterans, and eruptive pseudoangiomatosis.

C. Tan, M.D.
Department of Dermatology, Affiliated Hospital of Nanjing University of Chinese Medicine, Nanjing, China
e-mail: tancheng@yeah.net

H.-M. Jing
Department of Dermatology, Hospital of Traditional Chinese Medicine in Anyang City, Anyang, China

L.-T. Zhang, M.D.
Department of Dermatology, Tianjin Changzheng Hospital, Tianjin, China
e-mail: zhanglitao@medmail.com.cn

D.-L. Ma, M.D.
Department of Dermatology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

R.-Z. Zhang, M.D. (✉)
Department of Dermatology, The Third Affiliated Hospital of Suzhou University, Changzhou, China


W.-Y. Zhu et al. (eds.), *Atlas of Skin Disorders*, https://doi.org/10.1007/978-981-10-8037-1_14

## 14.1 Eruptive Recurrent Pigmented Telangiectasia [1]

- Eruptive recurrent pigmented telangiectasia (ERPT) is a newly described skin disorder that appeared initially as red, dome-shaped papules.
- Pathological feature consists of conspicuous dilation of the superficial blood vessels without blood vessel proliferation.
- Clinically, ERPT occurs annually and shows cyclical morphological changes from red papules to pigmented macules and finally clears spontaneously.

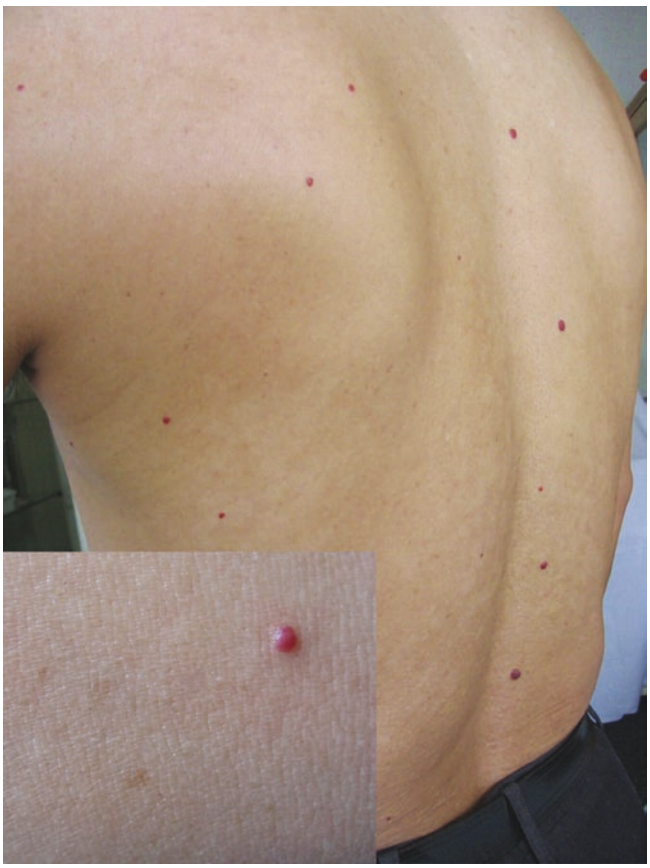

**Fig. 14-1-1** Bright red, dome-shaped papules measuring 1–4 mm on the trunk. Similar-sized oval or guttate pigmented macules distributed around the red papules (left low) (Reproduced with the permission from [1])

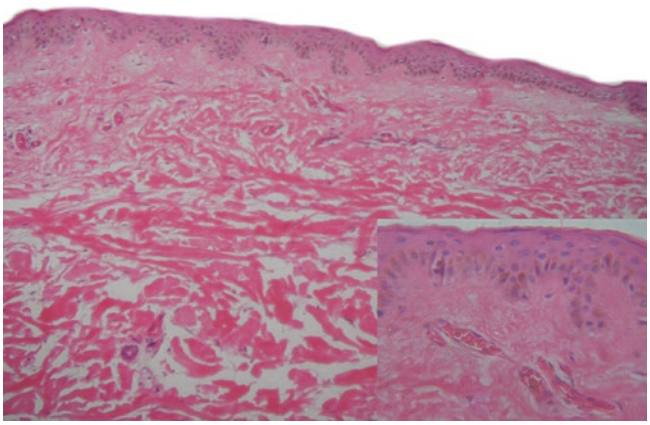

**Fig. 14-1-2 14-1-3** Superficial plexus are prominently dilated. There is no vessel proliferation, and perivascular lymphocytic infiltration is unremarkable ((1), HE stain, ×100). Dilated superficial blood vessels laden with red blood cells. The basement layer is slightly hyperpigmented ((2), HE stain, ×400) (Reproduced with the permission from [1])

## 14.2 Telangiectasia Macularis Eruptiva Perstans [2]

- Telangiectasia macularis eruptiva perstans (TMEP) is an extremely unusual form of cutaneous mastocytosis, preferentially occurring on the torso and proximal extremities, with occasional systemic involvement.
- The representative manifestations of TMEP are small, irregular reddish-brown telangiectatic macules covered by a tan to brown setting without pruritic phenomena. The diameter of the isolated lesion is approximately 2–4 mm.
- Adults are easily affected. Cold stimulation, shellfish, and aspirin may induce relapse. Skin biopsy is crucial for the diagnosis, revealing a slight increase in mast cells with infiltration of the epidermis.
- Treatment usually involves a focus on the relief of symptoms, phototherapy (PUVA and UVB), histamine receptor antagonists, and injectable epinephrine, for example.

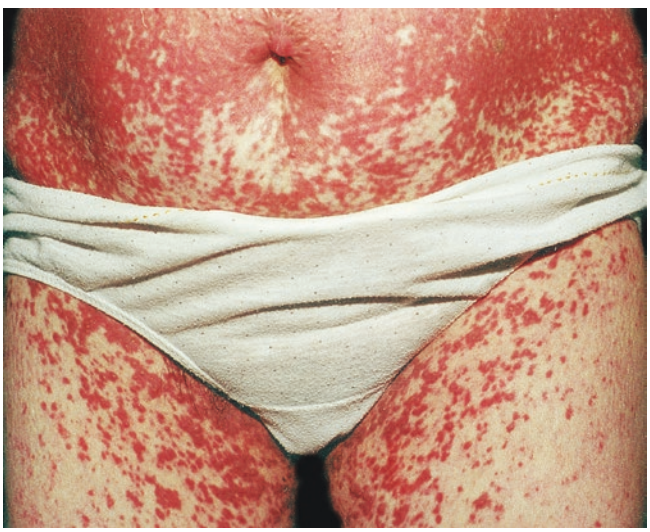

**Fig. 14-2-1** Numerous red-tan macules and papules 0.3–0.5 cm in diameter are scatted on the abdomen and lower limbs

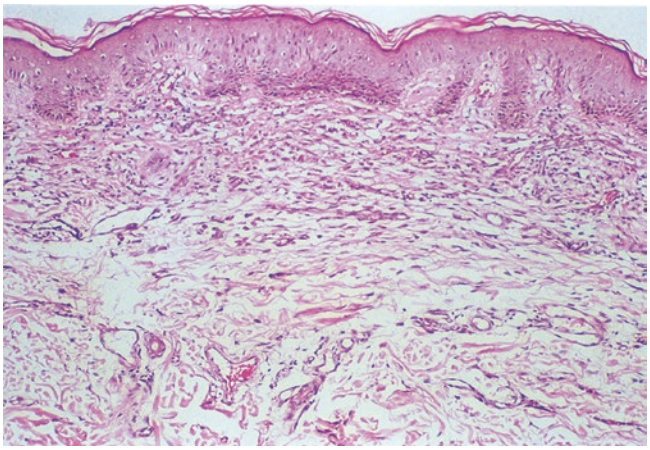

**Fig. 14-2-2** Hyperkeratosis and hyperpigmentation of the epidermal basal layer and an increased number of mast cells in the papillary dermis (HE stain, ×100)

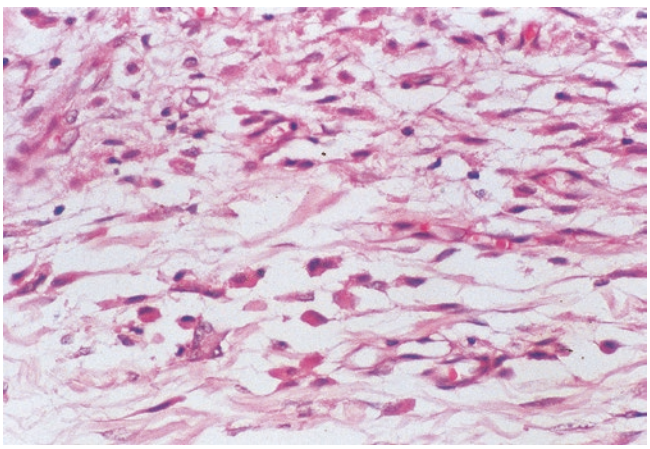

**Fig. 14-2-3** Numerous mast cells infiltrated in the dermis (HE stain, ×400)

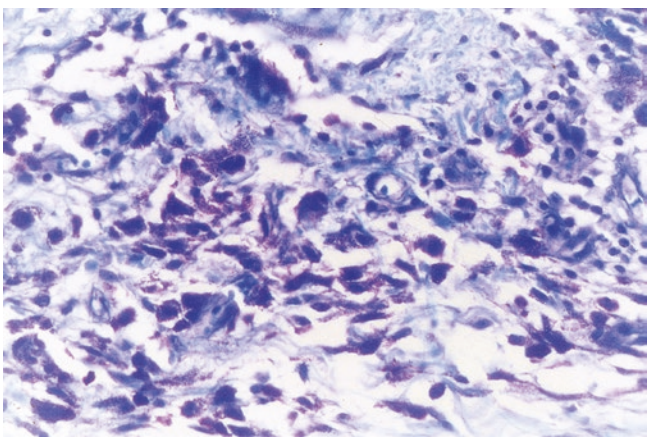

**Fig. 14-2-4** Metachromatic granules are found in mast cells (Giemsa stain, ×400)

## 14.3 Unilateral Nevoid Telangiectasia Superimposed on Bier Spots [3]

- Twin spotting represents two different visible neighboring spots originating from two mutant cell clones on a genetically normal background.
- Twin spotting can be categorized as allelic and nonallelic. The allelic category comprises vascular twin nevi, cutis tricolor, segmental lentiginosis with ipsilateral nevus depigmentosus, partial lipomatosis and partial lipohypoplasia, paired hairless and hairy nevus, and paired melanotic and achromic macules. Phakomatosis pigmentovascularis (types I–V), phakomatosis pigmentokeratotica, unilateral nevoid telangiectasia and Becker's nevus, nevus depigmentosus with intralesional lentiginosis and Becker's nevus, didymosis melorheosebacea, nevoid hypertrichosis and melanosis, and didymosis aplasticosebacea are examples of nonallelic forms.
- Vascular twin spotting defines two adjacent vascular birthmarks with different biological functions. Nevus anemicus and nevus flammeus are examples. Several cases of unilateral nevoid telangiectasia superimposed on Bier's spots reinforce the notion that it is a type of vascular twin spot.
- Examination of unilateral nevoid telangiectasia (UNT) reveals a unilateral distribution of numerous telangiectasias on the trunk, shoulders, and arms along with curtain dermatomes. UNT syndrome is applied with concurrent cirrhosis, malignancies, hepatitis B or C infections, and hyperthyroidism.
- Negative Darier's sign helps to differentiate the condition from cutaneous mastocytosis.
- UNT may have an increased number of estrogen receptors compared with the normal skin, although serum estrogen and progesterone are normal in most cases.

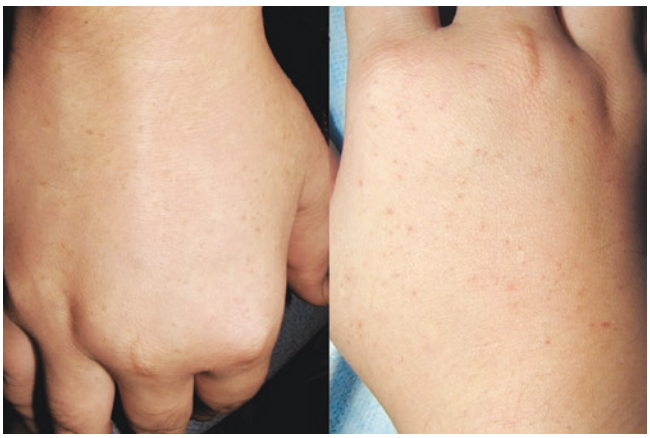

**Fig. 14-3-1** Multiple pin-sized wiry and spiderlike telangiectasia with peripheral pale macule unilaterally distributed on her right hand (Reproduced with the permission from [4])

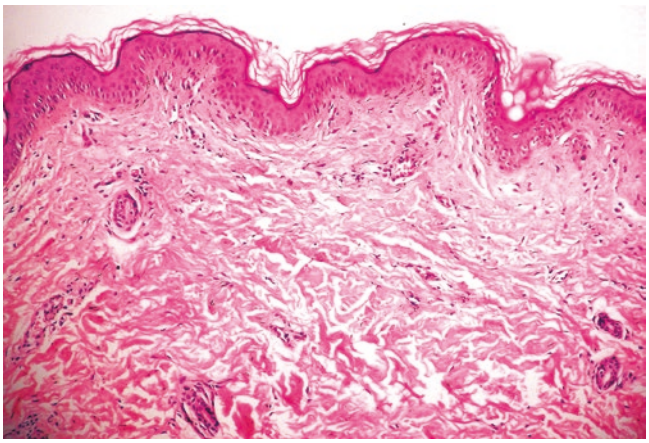

**Fig. 14-3-2** Skin biopsy revealed vascular dilation with mild perivascular infiltration of mononuclear cells in the papillary dermis (HE stain, ×100) (Reproduced with the permission from [4])

## 14.4 Mondor's Disease [5]

- Mondor's disease is characterized by a hypodermal, solid, painful cord companied by depression of the overlying skin. The aggravated pain factors include pressure on the lesion, breathing, or movement of the homolateral arm. There is no redness, increased temperature, or edema around the lesion.
- The pain and tenderness will be spontaneously relieved within a few days to several weeks. Treatment is focused on symptomatic relief.
- Mondor's disease, which mostly occurs in women aged 30–60 years, is an uncommon, innocuous, and self-limited disorder, also known as superficial thrombophlebitis of the chest. It affects the lateral thoracic vein.

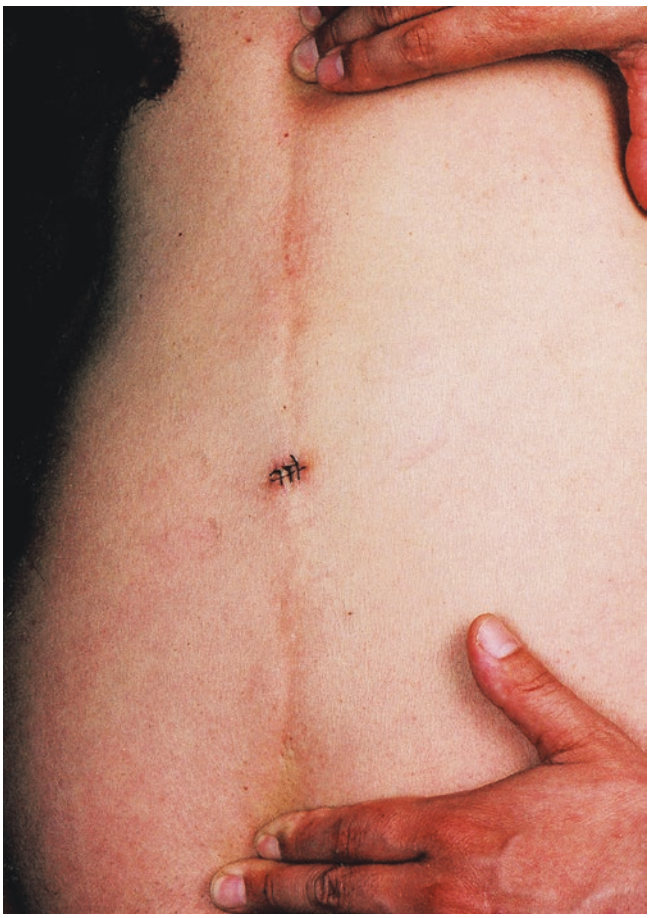

**Fig. 14-4-1** Strip hollow on the left breast

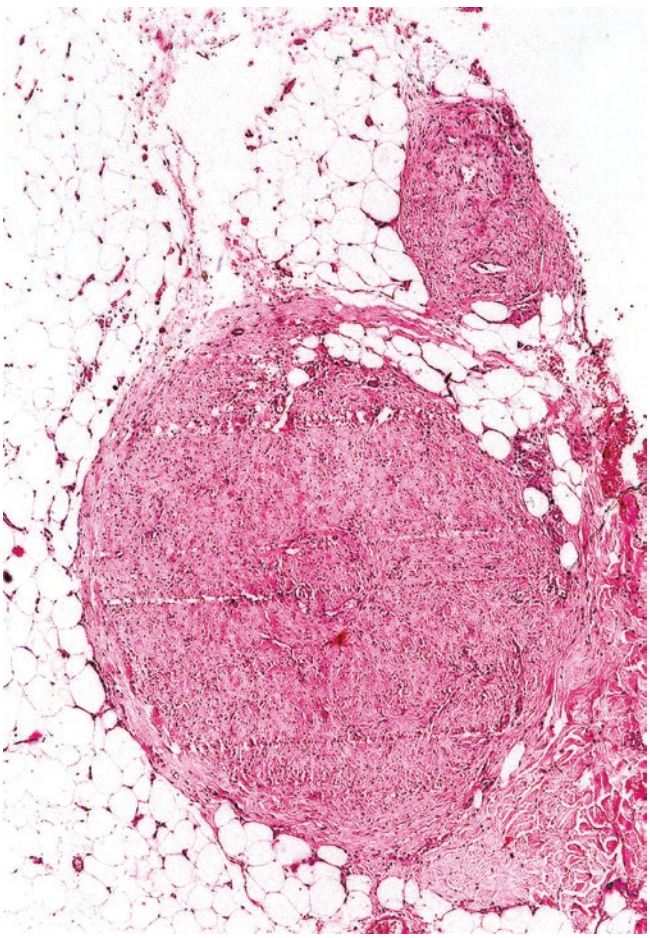

**Fig. 14-4-2** Tube structures with thrombosis in the dermis and vein wall thickening and fibrosis (HE stain, ×100)

## 14.5 Arteriosclerosis Obliterans [6]

- Arteriosclerosis obliterans (ASO) is one of the most frequent peripheral vascular diseases, characterized by claudication and rest pain. It usually affects the arteries of the lower extremities, as well as those of the heart, brain, kidney, and viscera.
- Skin ulcers due to ischemia appear along the peripheral arteries in ASO, usually leading to amputation. The pulsation of impaired arteries is attenuated and sometimes cannot even be felt. The temperature of the skin in the relevant area is usually reduced.
- Risk factors of ASO include age, male gender, smoking, hyperlipidemia, hypertension, diabetes mellitus, chronic renal failure, and hyperhomocysteinemia.

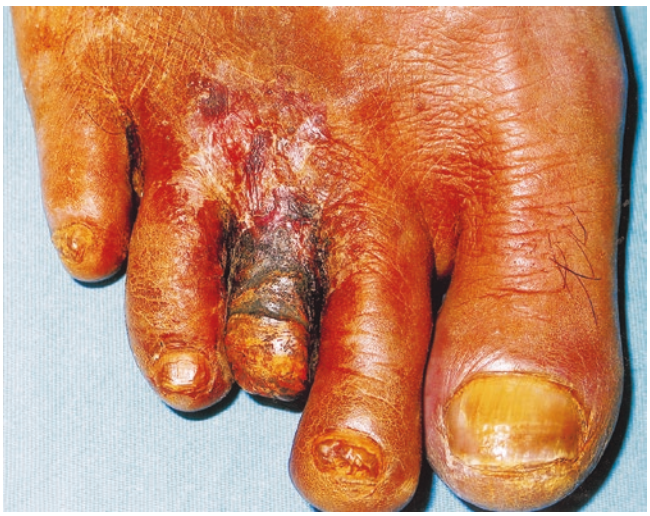

**Fig. 14-5-1** The toe of the right foot is purple, and the third toe is necrotic and shortened

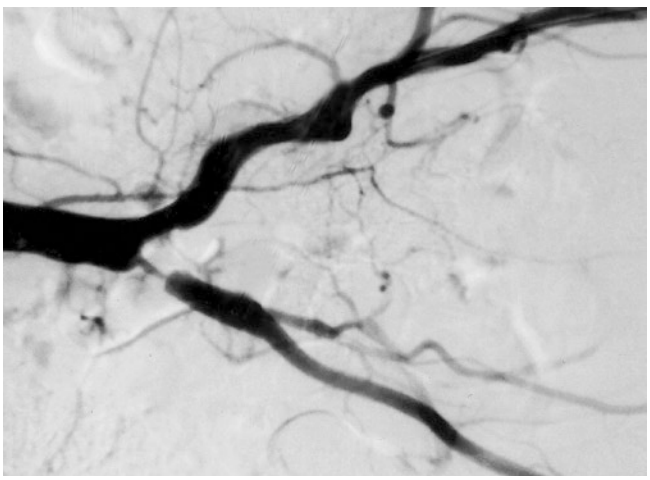

**Fig. 14-5-2** The narrowing of the hypogastric artery from the arteria femoralis

## 14.6 Eruptive Pseudoangiomatosis [7]

- Eruptive pseudoangiomatosis (EP) is characterized by degenerative erythematous papules with a faded halo.
- Pathological findings are hobnail-shaped endothelial cells in the dilated blood vessels. The blood vessels are not proliferative.
- EP can be misdiagnosed as erythema punctatum Higuchi following mosquito bites. Although dilation of dermal blood vessels is similarly present, the absence of hobnail endothelial cells distinguishes the condition from E.
- Most EP patients persist from 1 week to 5 months. It is advisable to use 1% pimecrolimus cream if necessary.

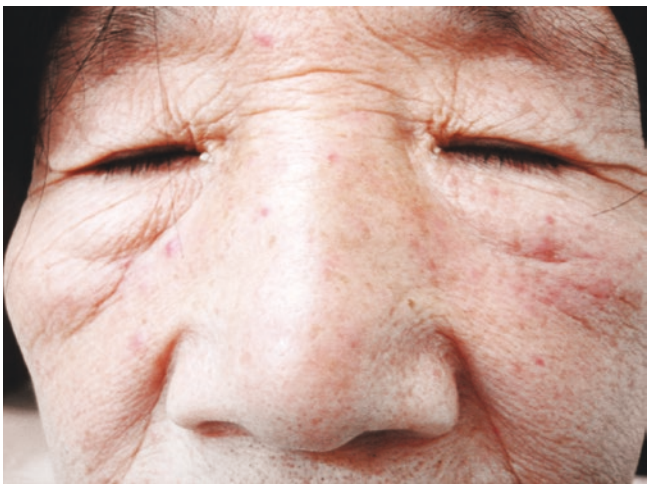

**Fig. 14-6-1** Disseminated, smooth, and angioma-like papules with diameters up to 2–5 mm mainly located on the face (Reproduced with the permission from [8])

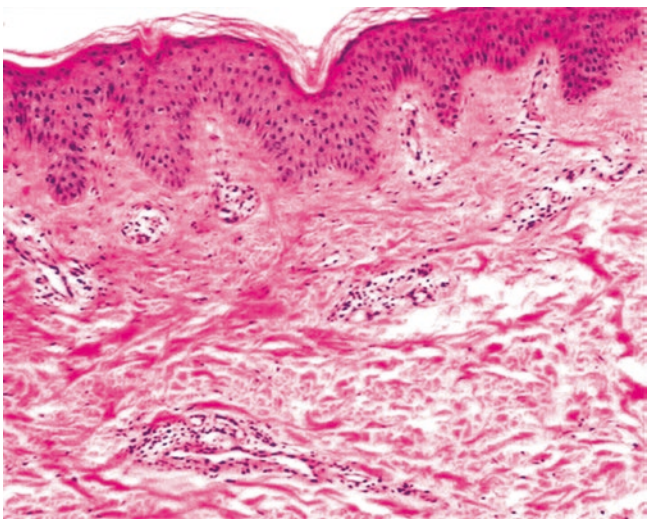

**Fig. 14-6-2** The epidermis was normal. The blood vessels in the superficial dermis were dilated and surrounded mainly by a lymphocyte infiltration (HE stain, ×100) (Reproduced with the permission from [8])

## References


1. Tan C, Zhu WY, Lai RS. Eruptive recurrent pigmented telangiectasia: a new clinical entity? Br J Dermatol. 2006;154(4):777–8. https://doi.org/10.1111/j.1365-2133.2005.07116.x.
2. Altiner A, Tzu J, Patel R, Meehan S, Sanchez M. Telangiectasia macularis eruptiva perstans. Dermatol Online J. 2011;17(10):7.
3. Lang N, Happle R, Enk AH, Haenssle HA. Unilateral nevoid telangiectasia in two adolescents. J Dtsch Dermatol Ges. 2017;15(6):660–2. https://doi.org/10.1111/ddg.13072.
4. Tan C, Zhu WY. Unilateral nevoid telangiectasia superimposed on the Bier spots: another demonstration of vascular twin spotting. J Dtsch Dermatol Ges. 2011;9(5):389–90. https://doi.org/10.1111/j.1610-0387.2010.07584.x.
5. Manimala NJ, Parker J. Evaluation and treatment of penile thrombophlebitis (mondor's disease). Curr Urol Rep. 2015;16(6):39. https://doi.org/10.1007/s11934-015-0512-z.
6. Miyahara T, Shigematsu K. Epidemiology, etiology, pathology, and pathophysiology of arteriosclerosis obliterans. Nihon Rinsho. 2016;74(Suppl 2):324–7.
7. Houberdon M, Meric M, Hatte D. Eruptive pseudoangiomatosis. Rev Prat. 2012;62(6):764.
8. Tan C, Zhu WY, Min ZS. A case of recurrent eruptive pseudo-angiomatosis that responded well to pimecrolimus 1% cream. Dermatology. 2009;218(2):181–3. https://doi.org/10.1159/000182267.