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Dermoscopic features of livedoid vasculopathy

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Abstract

Livedoid vasculopathy (atrophie blanche) is a form of thrombotic vasculopathy. It is characterized by small ulcers that become crusted, and heal after several months to produce white atrophic scars. The most commonly affected sites are the lower legs, in particular the dorsum of the feet and ankles. To date, the dermoscopic features of livedoid vasculopathy have not been clearly described in the literature. In this observational study, we sought to evaluate the dermoscopic patterns of livedoid vasculopathy and determine whether the dermoscopic features are associated with certain histopathological characteristics. We evaluated 9 patients with livedoid vasculopathy by dermoscopy. Skin biopsy specimens were stained with hematoxylin and eosin for histopathologic examination, and dermoscopic features were correlated with histopathological characteristics. In the majority of patients with livedoid vasculopathy, examination with dermoscopy revealed central crusted ulcers or ivory white areas associated with peripheral pigmentation in a reticular pattern. In addition, increased vascular structures including linear and glomerular vessels were found. On histopathological examination, the central ivory white areas correlated with dermal fibrosis, the reticular pigmentation corresponded to epidermal basal layer hyperpigmentation or melanin within melanophages in the dermal papillae, and the vascular structures correlated with dilatation and proliferation of capillaries in the upper dermis. In summary, the most common dermoscopic features of livedoid vasculopathy identified in this study were central crusted ulcers or ivory white scar-like areas associated with peripheral reticular pigmentation and increased vascular structures. The characterization of dermoscopic criteria for livedoid vasculopathy may improve the accuracy in the clinical diagnosis and follow-up of this disease.

Abbreviations: IgA = immunoglobulin A, IgG = immunoglobulin G, IgM = immunoglobulin M.

Keywords: dermoscopy, diagnostic accuracy, glomerular vessels, Livedoid vasculopathy

1. Introduction

Livedoid vasculopathy (also known as livedo vasculitis or atrophie blanche) is characterized by the appearance of erythematous or purpuric papules, which develop into small crusted ulcers that eventually heal to form white atrophic scars.[1–3] The ulcers are often painful, may heal poorly, and multiple recurrences are common. Fully developed lesions of livedoid vasculopathy are characterized by atrophic plaques with hyperpigmented borders and telangiectactic vessels. The lower legs, in particular the dorsum of the feet and ankles, are most often affected.[4] The disease is more common in middle-aged women. Venous stasis can also be associated with this disease. The severity of the disease may have a seasonal variation, with the painful ulcers often aggravating in the winter period.

Dermoscopy is a noninvasive imaging tool that allows the visualization of morphological features (such as pigmentation pattern and vascular structure), which are too small to be seen by the naked eye.[5–7] It has been shown to improve the accuracy in diagnosis of various pigmented and nonpigmented skin lesions. Dermoscopy enables the assessment of the entire skin lesion in a short amount of time, compared to skin biopsy in which only a small area of the skin lesion is assessed. The dermoscopic features of malignant melanoma have been investigated by various studies.[8–15] The dermoscopic patterns of cutaneous inflammatory disorders have yet to be clarified fully. In this study, we sought to determine the dermoscopic features of livedoid vasculopathy and correlate these features with the histopathologic findings.

2. Methods

Nine patients with biopsy-proven livedoid vasculopathy who presented to the Dermatology department of Kaohsiung Medical University Hospital were included in this study. Dermoscopic assessment was performed for all livedoid vasculopathy skin lesions in the 9 patients. This study was approved by the institutional review board of Kaohsiung Medical University Hospital.

All skin lesions were examined by board-certified dermatologists who had >10 years of experience with dermoscopy. Dermoscopic examination was performed using a DermLite Foto polarized light dermoscope (10× magnification; 3Gen, Dana

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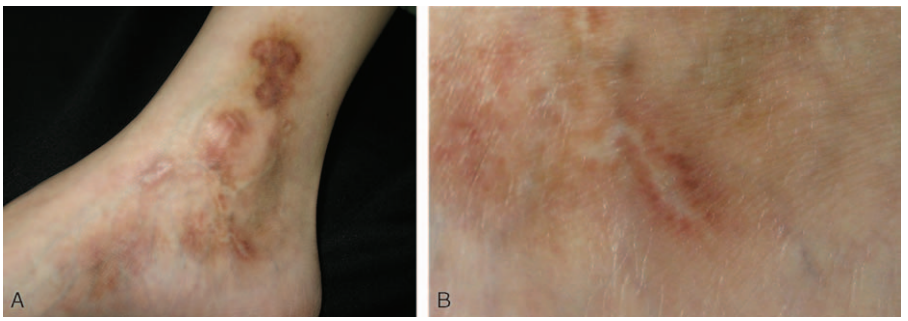

Figure 1. (A) Clinical image of livedoid vasculopathy lesions on the lower leg. (B) Close-up view showing white atrophic scars with a hyperpigmented border.

Point, CA) attached to a Nikon Coolpix 995 digital camera (3× optical zoom; Nikon, Tokyo, Japan). The DermLite Foto dermoscope contains polarization filters, and therefore immersion liquids do not need to be applied during skin examination. All dermoscopic images were projected onto a 30-inch liquid crystal display screen for more detailed visualization.

All skin biopsy specimens were stained with hematoxylin-eosin for histopathologic evaluation. Evaluation of pathology specimens was performed by 2 dermatologists who were experienced with dermatopathology. For correlation of dermoscopic features and pathological characteristics, skin biopsies were performed at the time of dermoscopy, in the same area of dermoscopic examination.

3. Results

3.1. Clinical features of patients with livedoid vasculopathy

All 9 patients were Taiwanese women with livedoid vasculopathy. They presented with multiple small ulcers on the bilateral lower legs, which healed to form white atrophic scars (Fig. 1). The demographic and clinical features of the patients are presented in Table 1.

3.2. Dermoscopic features of patients with livedoid vasculopathy

We found several common features of livedoid vasculopathy by dermoscopy. First, the center of the lesions showed either shallow crusted ulcers (5/9 patients) or ivory-white atrophic scar-like areas (8/9 patients; Fig. 2). Second, the periphery of the lesions was characterized by hyperpigmentation in a reticular pattern (8/9 patients; Fig. 3). In comparison to the pigment network seen in melanocytic lesions, the brownish lines in the reticular pigmentation among our patients with livedoid vasculopathy were thinner and more closely positioned and the holes in the network were smaller and more regular in shape and size. Third, increased vascular structures in the periphery of the lesions were seen, including telangiectatic linear vessels (9/9 patients) and glomerular vessels (7/9 patients;).

3.3. Histopathology of livedoid vasculopathy lesions

Histopathology of livedoid vasculopathy lesions showed epidermal thinning or ulceration. Hyperpigmentation of the basal layer of the epidermis and melanin within dermal melanophages were seen in some lesions. In the superficial and mid-dermis, vessels containing intraluminal hyaline thrombi were found. Dilated vessels and an increased number of dermal blood vessels were also seen in some parts of the lesion (Fig. 6). There was a mild perivascular infiltrate of lymphocytes, but no vascular destruction or fibrinoid necrosis was observed. Extravasated erythrocytes were frequently found. The dermis in some lesions showed moderate to dense fibrosis and scarring.

3.4. Correlation of dermoscopic features and pathologic findings

The central ivory white area corresponded to dermal fibrosis following the healing of vasculitic ulcers. The reticular pigmentation correlated with hyperpigmentation of the basal layer of the epidermis or melanin within melanophages in the dermal papillae. The linear and glomerular vessels corresponded to dilatation and proliferation of capillaries in the upper dermis.

4. Discussion

The pathogenesis of livedoid vasculopathy is not well defined. Previously, the disease was regarded as a form of vasculitis. However, livedoid vasculopathy is now considered to be caused

Table 1

Demographic, clinical, and dermoscopic characteristics of patients with livedoid vasculopathy.

Patient	Age	Sex	Location	Ulcer	Ivory white area	Reticular pigmentation	Linear vessels	Glomerular vessels
1	46	Female	Lower legs + feet	−	+	+	+	+
2	54	Female	Lower legs + feet	+	+	+	+	+
3	41	Female	Lower legs	−	+	+	+	−
4	33	Female	Lower legs	+	+	+	+	+
5	40	Female	Feet	−	+	+	+	+
6	37	Female	Lower legs + feet	+	−	+	+	−
7	41	Male	Lower legs + feet	+	+	+	+	+
8	42	Female	Feet	+	+	−	+	+
9	39	Female	Lower legs	−	+	+	+	+

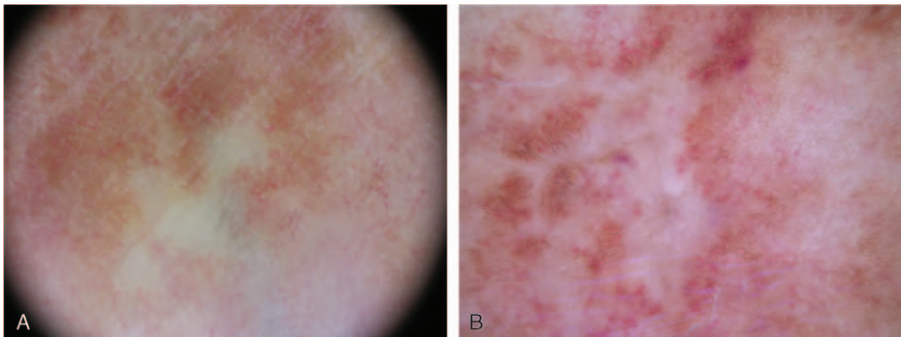

Figure 2. (A, B) On dermoscopy, most of the lesions were characterized by central crusted ulcer or whitish scar-like area with peripheral pigmentation and increased vascular structures.

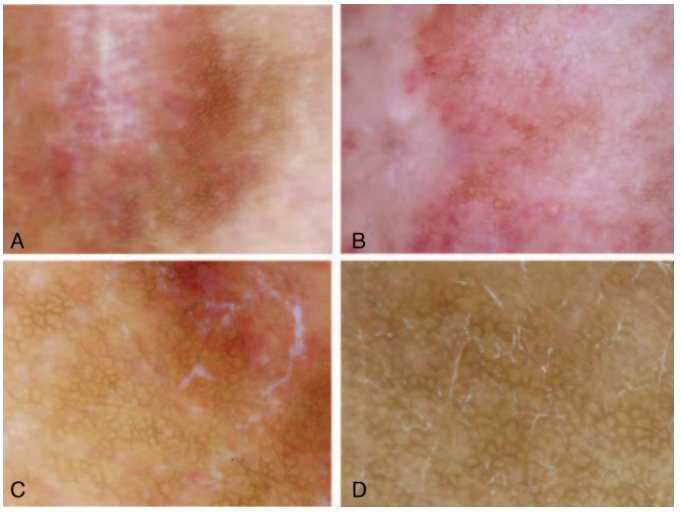

Figure 3. (A–D) High-power dermoscopic views of peripheral pigment network (reticular pigmentation).

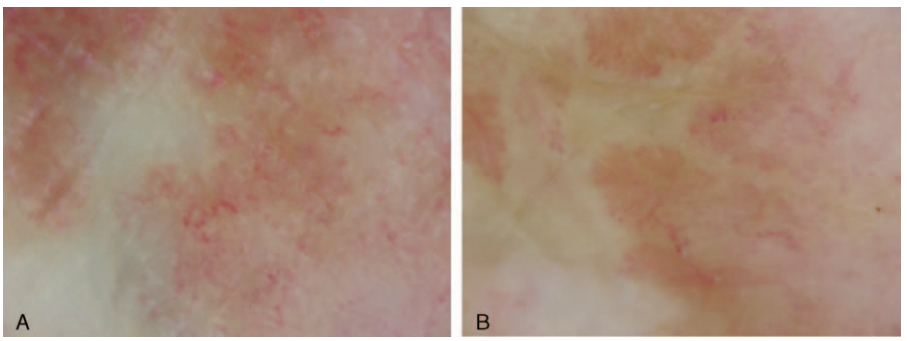

Figure 4. (A, B) High-power dermoscopic views of linear vessels.

by a hypercoagulable state, leading to obstruction of small dermal blood vessels by intraluminal thrombi.[16–20] In addition, the platelets may be characterized by increased aggregation ability.[21–23] This may lead to skin ischemia with ulceration. Furthermore, the presence of immunoglobulins (IgM, IgG, and IgA) and complement within the vascular walls suggest that an immunological process may be involved in the pathogenesis.[24]

The histopathological features of livedoid vasculopathy may vary depending on the age of the skin lesion. Microscopically, early lesions of livedoid vasculopathy showed an increased number of blood vessels in the dermis, which often contain intraluminal hyaline thrombi. The small vessels in the superficial and mid-dermis are mostly involved, but deeper vessels may be rarely affected. The walls of affected blood vessels may also contain fibrin material. Small areas of ulceration may be found in the upper dermis secondary to vascular occlusion and infarction. A mild perivascular infiltrate of lymphocytes may be seen, but typical features of vasculitis such as fibrinoid necrosis of the vessel

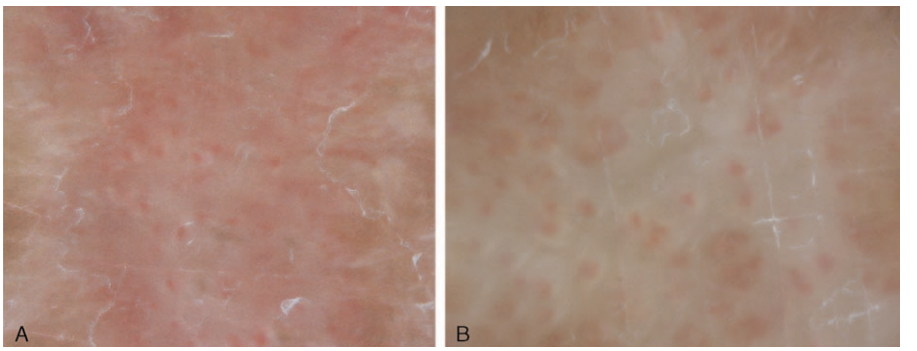

Figure 5. (A, B) High-power dermoscopic views of glomerular vessels.

walls are not present. A small number of neutrophils may be seen at the base of the ulcer. Extravasated erythrocytes are frequently observed in the superficial dermis. Increased numbers of dermal blood vessels are often present in the perilesional skin. In later stages of livedoid vasculopathy, the epidermis may be atrophic, and dense scarring may be found in the dermis.[25,26]

In our dermoscopic examination of 9 patients with livedoid vasculopathy, we found several common features. The center of the lesions was characterized by shallow crusted ulcers or ivory white scar-like areas, whereas the periphery of the lesions showed hyperpigmentation in the form of reticular pigmentation and increased vascular structures. Previously, Criado et al[27] performed dermoscopic examination on a patient with livedoid vasculopathy, which showed white scar and purpuric macule. The white scar was similar in appearance to our cases, whereas the purpuric macule may represent an early preulcerative lesion.

In addition, dermoscopic examination of 3 cases of venous stasis dermatitis by Zaballos et al[28] revealed glomerular-like vessels and a scaly surface. However, it should be noted that ivory white scar-like areas and reticular pigmentation seen in livedoid vasculopathy lesions were not found in these 3 cases of chronic venous stasis.

In our cases, the central ivory white area corresponds to dermal fibrosis following healing of vasculitic ulcers. Previously, whitish areas under dermoscopy had been seen in dermatofibromas, corresponding to increased collagen and fibrosis in the upper dermis.[29–36] The whitish areas in dermatofibromas had been previously noted to be more prominent under polarized dermoscopy compared to those under nonpolarized dermoscopy, which could be a result of greater penetration of light into the deeper layers of the dermis using polarized dermoscopy.[37,38]

In livedoid vasculopathy lesions, the peripheral pigmentation is arranged in the form of reticular pigmentation in many places.

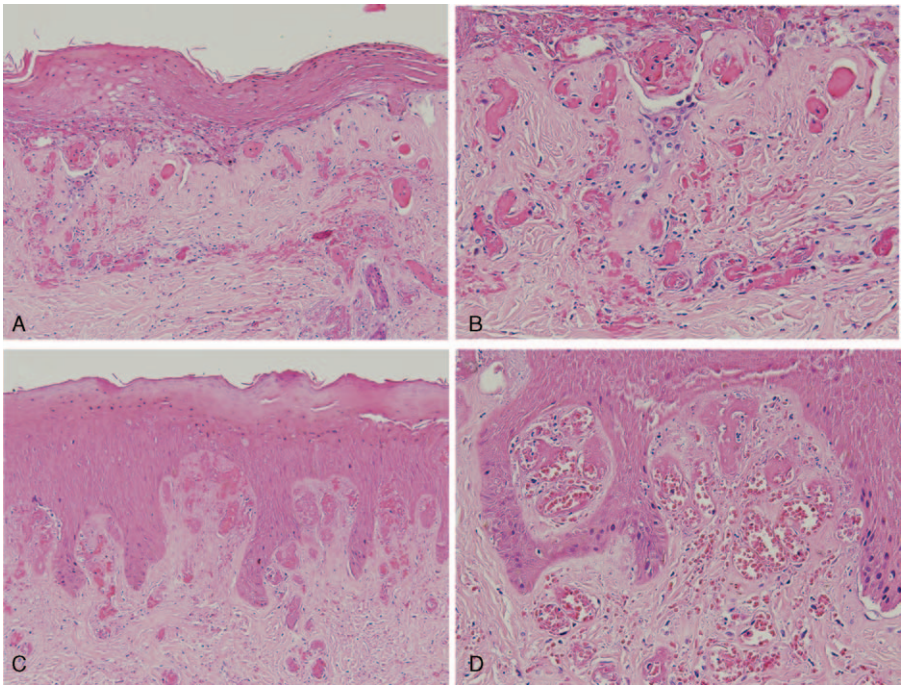

Figure 6. (A, B) Histopathology of livedoid vasculopathy skin lesions revealed epidermal thinning or ulceration, and moderate to dense fibrosis and scarring in the dermis. Vessels containing intraluminal hyaline thrombi could be found in the superficial and mid-dermis. (C, D) Dilated vessels and an increased number of dermal blood vessels were also seen in some parts of the lesion. Magnification: 100× (A, C), 200× (B, D).

Pigment network is a characteristic of benign and malignant melanocytic lesions, such as lentigines, nevi, and melanomas.[39] In benign nevi, the pigment network corresponds to the clusters of nevus cells in the rete ridges. More recently, the pigment network has also been observed in a variety of nonmelanocytic tumors, such as dermatofibromas and mastocytosis (urticaria pigmentosa).[31,40,41] Previously, we also observed reticular pigmentation in pigmented Bowen's disease.[42] In our patients with livedoid vasculopathy, the pigment network or reticular pigmentation may correspond to hyperpigmentation of the basal layers of the epidermis or melanin within melanophages in the dermal papillae.

In comparison to the pigment network seen in melanocytic lesions (such as melanocytic nevus), we found that the brownish lines in the pigment network among our patients with livedoid vasculopathy were thinner and more closely positioned and the holes in the network smaller and more regular in shape and size. The dermoscopic differences between the pigment network in livedoid vasculopathy and melanocytic lesions are understandable, as the pigment network in livedoid vasculopathy corresponds histopathologically to increased melanin pigment in the basal layers of the epidermis, whereas the pigment network in melanocytic lesions corresponds histopathologically to nevus cells in the rete ridges.[43]

The dermoscopic vascular pattern of livedoid vasculopathy as observed in our cases was characterized by telangiectatic linear and glomerular vessels. The prominence of vascular structures observed surrounding the central ivory white plaque might be because of dilatation and proliferation of vessels in response to thrombosis and ischemia of the central area. Glomerular vessels are a particular type of dotted vessels, which resemble the convoluted vascular structures of the renal glomerulus, and correspond histopathologically to the presence of grouped, convoluted, and dilated capillaries in the papillary dermis.[44,45] Previously, glomerular vessels have been described in Bowen's disease.[46–50] In addition, it has been found in chronic venous stasis.[28] The mechanisms responsible for the formation of glomerular vessels in chronic venous stasis and livedoid vasculopathy may be similar. Both these diseases are associated with chronic severe ischemia of the involved skin, which may promote angiogenesis and lead to an increased number of dermal blood vessels in the surrounding skin.

There are some limitations in this study, including the small number of patients and lack of a control group to confirm the specificity of the findings. Further studies involving a larger number of patients are warranted to determine the sensitivity and specificity of the dermoscopic features.

In this report, we described the dermoscopic features of a series of patients with livedoid vasculopathy. We found that reticular pigmentation, which had been previously described mostly in melanocytic lesions, could also be clearly seen in livedoid vasculopathy. In addition, glomerular vessels, which had only been described in Bowen's disease and chronic venous stasis, could be found in livedoid vasculopathy lesions as well. The identification of dermoscopic features of livedoid vasculopathy will lead to increased accuracy in the diagnosis and follow-up of this disease.

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