

The dermoscopic variability of Degos disease at different stages of progression

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A 34-year-old woman presented with an 8-month standing eruption disseminated on the trunk and anterior upper limbs. Following their initial appearance, the skin lesions were described to occur in crops. On clinical examination, lesions of recent onset were small, dome-shaped, pale-red papules surrounded by an erythematous halo (Figure 1a), while papules of longer duration displayed a central ulceration covered by a yellowish crust and were surrounded by a whitish rim (Figure 2a). Finally, porcelain-white scars with a reddish or hyperpigmented halo were noted on the site of healed lesions.

A 44-year-old man presented with widespread, bilateral and symmetrical skin lesions with a 6-month history. Similarly to the previous patient, the eruption consisted of lesions

in three different evolution stages: reddish papules of recent onset, measuring 5 to 10 mm in diameter and not disappearing with pressure; reddish papules with a purple or necrotic center with or without central crust; and porcelain-white scars surrounded by an erythematous halo (Figure 3a).

Dermoscopic examination in both patients revealed three different patterns, each one corresponding to a different evolution stage. Specifically, papules of recent onset were dermoscopically characterized by the combination of a reddish-to-purple background and purpuric dots (Figure 1b). At an intermediate progression stage, the papules displayed a targetoid pattern with yellowish, purple or necrotic center surrounded by an erythematous halo (Figure 2b). Finally,

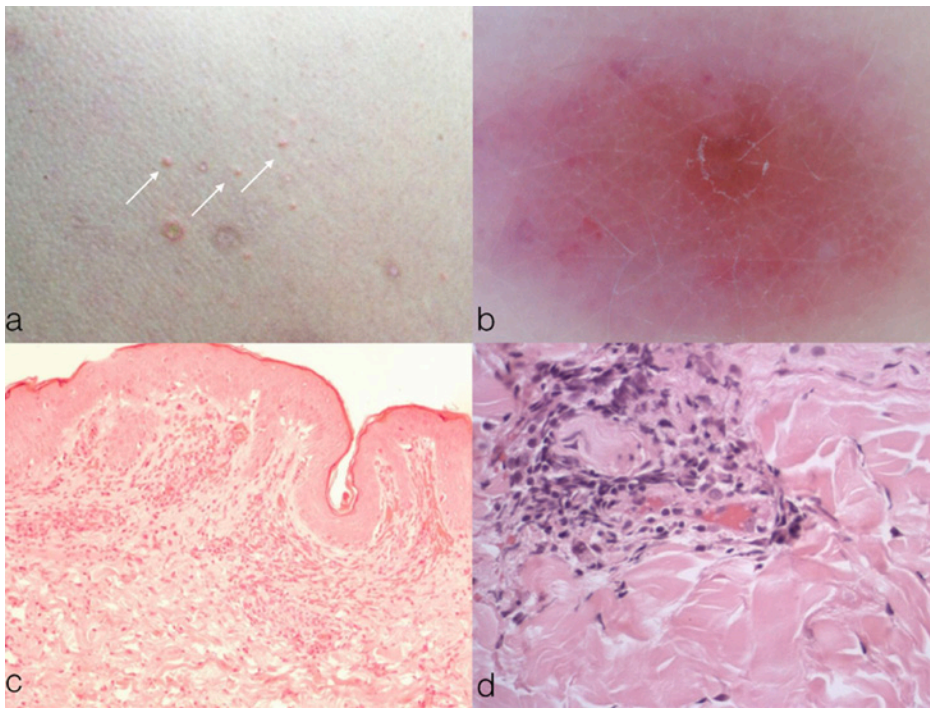


Figure 1. Dome-shaped reddish papules of recent onset (white arrows) on the thigh of a 34-year-old woman (a). Dermoscopic examination revealed a reddish-to-purple background and purpuric dots (b). Histopathologically, the lesions were characterized by focal basal vacuolization, necrotic keratinocytes and moderate superficial perivascular lymphocytic infiltration in the dermis (c-H&E X100). A higher magnification revealed intravascular thrombi without fibrinoid necrosis of the wall and some extravasated red blood cells (d-H&E X200). [Copyright: ©2014 Pérez Anker et al.]

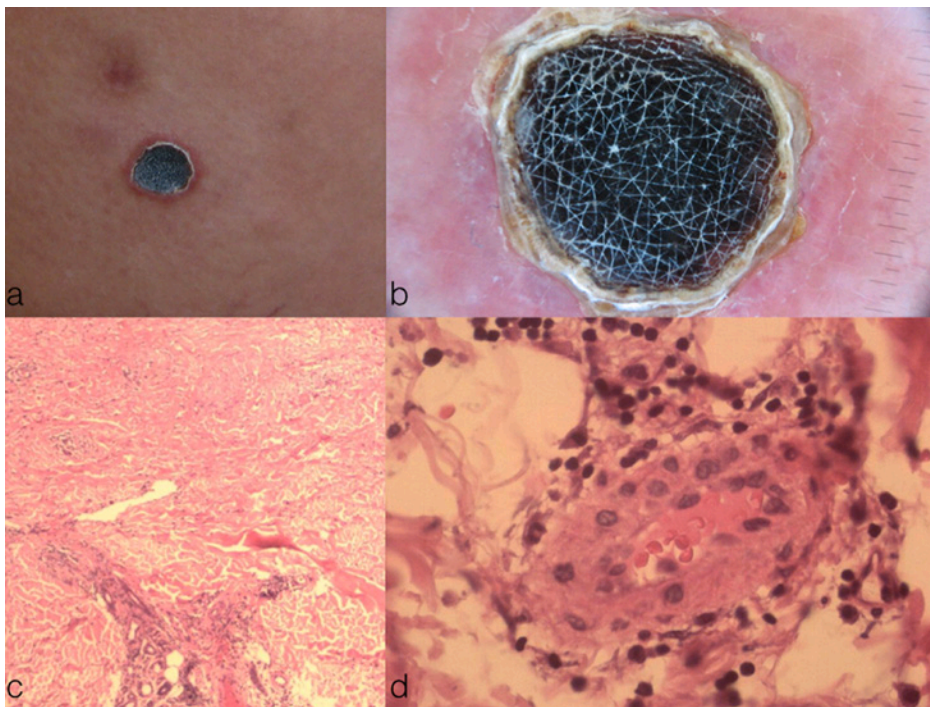


Figure 2. Longer standing papules in the same patient with central ulceration or necrosis (a), dermoscopically typified by a necrotic center surrounded by an erythematous halo (b). Histopathology showed extensive epidermal necrosis with foci of re-epithelialization and superficial and deep perivascular inflammatory infiltrate in the dermis (c-H&E X100). At a higher magnification, numerous extravasated red cells, some intraluminal thrombi and capillary congestion without leukocytoclasia was seen (d-H&E X200). [Copyright: ©2014 Pérez Anker et al.]

dermoscopy of healed lesions revealed a whitish structureless center surrounded by a rim of short, thin and slightly curved vessels (Figure 3b, c).

A biopsy was taken from three lesions, one of each progression stage. The histopathologic findings corresponded well to the dermoscopic criteria (Figures 1c & d, 2c & d, 3d). The histopathologic examination in both patients was overall suggestive of Degos disease (DD).

The clinical recognition of DD is often troublesome, since the disease is characterized by polymorphic and rather unspecific cutaneous manifestations [1,2]. As highlighted by

our cases, clinical examination usually reveals lesions at different progression stages, ranging from light pink papules that do not fade with pressure to necrotic papules surrounded by erythema and whitish scars [1-3]. A delayed diagnosis might significantly deteriorate the patient's prognosis, since DD is a potentially fatal obliterative arteritis syndrome with an unpredictable physical course. Specifically, in some affected patients the disease remains restricted to the skin and follows a long benign course, while others die due to fulminating peritonitis or cerebral infarctions within the first few years [3,4]. Effectively, the diagnosis of DD should always be followed by

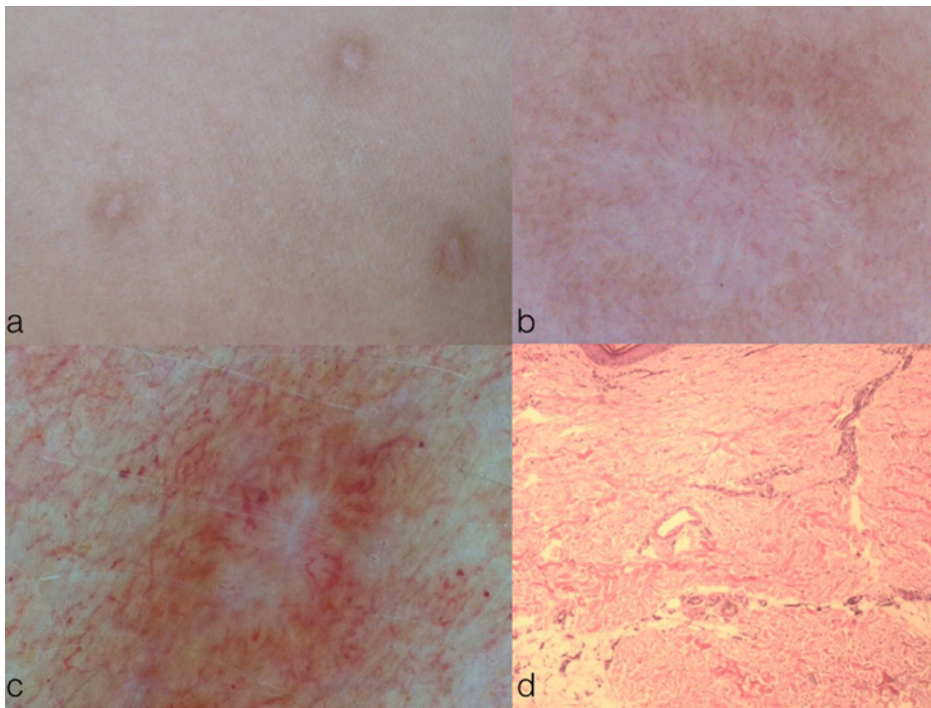


Figure 3. Porcelain-white scars with a reddish or hyperpigmented halo on the forearm of a 44-year-old man (a). Dermoscopic examination showed a central whitish structureless area (b), sometimes surrounded by a rim of short, thin, slightly curved vessels (c). Histopathology revealed an atrophic epidermis and dermal fibrosis with absence of pilosebaceous units. [Copyright: ©2014 Pérez Anker et al.]

monitoring for systemic involvement, and treatment should be adjusted accordingly [3,4]. In our patients, laboratory, imaging and endoscopic examinations did not reveal any signs of systemic involvement. In the first patient, a treatment with 100 mg of aspirin daily was initiated, followed by remission of the lesions and no relapse during a 4-year follow up. The second patient was already under anticoagulant medication because of heart failure, and no additional treatment was initiated.

Dermoscopy has been shown to enhance the recognition of several inflammatory skin diseases, especially when the macroscopic morphology is not typical enough to allow a clinical diagnosis [5-7]. A dermoscopic “crown of thorns,” consisting of linear and hairpin vessels surrounding a central scar, has been previously described in DD [8-10]. Our cases highlight that the latter pattern can be seen in late-stage lesions of DD, while early and intermediate lesions exhibit different dermoscopic findings, as shown in Figures 1 to 3.

Our findings provide an indication that dermoscopy might enhance the clinical recognition of DD, by revealing three different patterns which correspond to lesions at different evolution stages and mirror the underlying histopathologic alterations.

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