Reflectance Confocal Microscopy Features of Degos Disease

Degos disease, otherwise known as a malignant atrophic papulosis, is a rare occlusive vasculopathy characterized by pathognomonic cutaneous lesions and frequently fatal systemic involvement. The cause of Degos disease is unknown, and there is currently no effective treatment. Cutaneous lesions of Degos disease have a typical histologic appearance consisting of wedge-shaped necrosis of the dermis. Reflectance confocal microscopy (RCM) is a new in vivo skin imaging technique. The resolution of emerging images is close to that of conventional microscopy (approximately 1 μm), with a penetration depth up to 200 μm allowing the morphologic observation of the normal and abnormal dermis. Our treatment of a patient with Degos disease prompted us to investigate the RCM features of his skin lesions.

Report of a Case | A 50-year-old white man with a medical history of chronic hepatitis C virus and diabetes mellitus presented with recurrent skin lesions of 2 months’ duration. Cutaneous examination showed about 10 small papules 5 to 10 mm in diameter with white centers surrounded by erythematous borders, nonpruritic and painless, located principally on the trunk and the lower extremities (Figure 1). The patient had no neurologic or abdominal symptoms, and physical examination was otherwise unremarkable.

Dermoscopy of several typical lesions showed the same pattern of a white central structureless area crowned with telangiectasias or hairpin vessels (Figure 2). Examination of the same lesions with RCM showed a loss of epidermal structures and in the superficial dermis an abnormal aspect of collagen fibers that appeared highly refractile and grouped in a mass instead of the normal aspect of moderately refractile bundles in the surrounding normal skin (Figure 3A). In addition, capillaries were hardly visible in the lesion (Figure 3B), contrasting with increased and dilated capillaries in normal skin (Figure 3C). This was confirmed by dynamic examination using the video mode, which showed a decreased blood flow in the lesion compared with normal skin.

Histologic analysis confirmed the diagnosis of Degos disease, showing an ulcerated epidermis overlying a wedge-shaped area of necrosis in the dermis, with eosinophilic and densified collagen fibers and a decrease of dermal capillaries (Figure 4). Careful examination at the edge of the necrotic area confirmed the presence of capillary hyperplasia in the periphery of the lesion.

Discussion | Erythematous papules with a white atrophic center are pathognomonic of Degos disease and reflect a specific histologic image of wedge-shaped necrosis. Herein we provide an RCM description of Degos disease skin lesions. In the central atrophic regions of the papules, we found a reproducible pattern of dermal necrosis with collagen densification and loss of dermal capillaries. These RCM features correlated closely with the underlying histologic changes. In addition, RCM disclosed that the telangiectatic ring, previously characterized as...
a *crown of thorns* in dermoscopy,\(^3\) corresponded to increased and dilated dermal capillaries, a new finding that was confirmed by histologic analysis and, to our knowledge, not described previously.\(^4\) In conclusion, as with the clinical and pathologic presentations, the RCM features associated with cutaneous lesions of Degos disease seem to exhibit very distinctive RCM features.

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Sibling Cases of Hailey-Hailey Disease Showing Atypical Clinical Features and Unique Disease Course

Hailey-Hailey disease (HHD), a well-characterized autosomal dominant hereditary disease, is caused by mutation in *ATP2C1* gene and clinically shows characteristic erosive lesions predominantly on the intertriginous areas.\(^1-3\) We herein report sibling cases of HHD with novel mutations in the *ATP2C1* gene that showed unique and atypical clinical phenotypes mimicking seborrhoeic dermatitis, pemphigus vulgaris, or pemphigus foliaceus as well as considerable alterations during the disease course.

Report of Cases | Case 1. A 61-year-old Japanese man with intractable pulmonary arterial hypertension, pulmonary fibrosis, and emphysema was receiving oxygen supply from a nasal...