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Observation

Disseminated Mantle-Cell Lymphoma Presenting as a Petechial Maculopapular Eruption

Cutaneous involvement by mantle-cell lymphoma (MCL) is rare. We describe an unusual case of advanced-stage MCL that presented with a diffuse petechial maculopapular eruption mimicking a hypersensitivity reaction.

Report of a Case | A man in his early 50s presented with a petechial maculopapular eruption with rapid dissemination to the entire body including the face and ears. Peripheral lymphadenopathy was noted. He also had an elevated white blood cell count of 26,900/μL. (To convert white blood cell count to ×10^9/L, multiply by 0.001.) He had mild pruritus with no other constitutional symptoms. Skin biopsies showed a prominent perivascular and perivascular infiltrate of small to medium-sized monomorphic lymphocytes with focal erythrocyte extravasation but without evidence of vasculopathy or vasculitis. Immunohistochemical analysis revealed CD20+, CD5+, cyclin-D1+, CD3-, CD10-, and CD23+, infiltrate, consistent with cutaneous involvement by MCL (Figure). Staining for skin-homing molecules revealed a cutaneous lymphocyte-associated antigen (CLA), L-selectin (CD62L)+ infiltrate.

Laboratory investigations showed peripheral blood and bone marrow involvement by MCL with chromosomal translocation t(11;14), and positron emission tomographic/computed tomographic scans revealed diffuse lymphadenopathy. The patient was started on intensive chemotherapy followed by allogeneic stem cell transplant.

Discussion | Mantle-cell lymphoma is a B-cell lymphoma that arises from pregerminal naïve B cells populating the mantle zone of lymphoid follicles. The genetic hallmark is the chromosomal translocation t(11;14)(q13;q32) that results in aberrant cyclin-D1 expression. Histologically, MCL is characterized by small to medium-sized CD5+, CD10+, CD23+, cyclin-D1+ lymphoid cells with irregular nuclear contours, dispersed chromatin, inconspicuous nucleoli, and scant cytoplasm. It is typically marked by an aggressive clinical course.

We describe a patient with a diffuse eruption as initial manifestation of systemic MCL. Cutaneous involvement is rare, seen in 2% to 6% of MCL cases, and is associated with concurrent lymph node and systemic disease. It usually presents as isolated nodules or plaques. Our case is unique in its presentation as a petechial eruption that rapidly spreads to involve most of the body, face, and unusually, the ears. To our knowledge, there has been only 1 other case in which MCL presented as a lesion on the ear, and the widespread distribution in our case raised alternative clinical possibilities such as a hypersensitivity reaction or viral infection.

The histopathologic findings of the skin biopsy were consistent with cutaneous involvement by MCL. The slight perivascular erythrocyte extravasation correlated with the petechial presentation. Given the disseminated systemic disease, the cutaneous involvement likely reflected the patient’s high tumor burden.

Cutaneous involvement by MCL may be the result of tissue-specific adhesion molecules on malignant B cells inducing their migration into the skin. Chemokine receptors, selectins, and integrins are all involved in cutaneous homing, facilitating lymphocyte migration via interactions with vascular endothelium and the local microenvironment. Of these, CLA, which binds to E-selectin on endothelial cells, plays a major role in skin-homing of memory T cells. Data on CLA expression on B lymphocytes is scarce. A single case report demonstrated CLA expression by cutaneous involvement of lymphoblastoid MCL that was not substantiated in other cases. Our case was CLA+ but L-selectin-.

L-selectin is a lymph node-homing molecule that is constitutively expressed on central memory T cells to facilitate their migration by rolling on vascular endothelium and subsequent entry into lymphoid tissues, hence facilitating their skin homing as well. L-selectin is expressed on B lymphocytes that recirculate between peripheral blood and secondary lymphoid tissues. B cells that are not part of this recirculating pool tend to lack L-selectin expression and migrate preferentially to the spleen. Interestingly, L-selectin expression has not been reported in MCL or other B-cell neoplasms and may correlate with the high tumor burden that facilitated lymphocyte homing to the skin. In our case, the perivascular distribution seen on histologic analysis likely resulted from L-selectin binding to vascular endothelial cells. However, we cannot rule out that local cytokines and other adhesion molecules contributed to this unusual presentation.

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A and B, Clinically, a diffuse petechial maculopapular eruption is seen on the trunk (A) and thigh (B). C and D, Histologic sections show a superficial to deep dermal perivascular infiltrate (C) (hematoxylin-eosin, original magnification ×20) with areas of red blood cell extravasation (D) (hematoxylin-eosin, original magnification ×200). E, The infiltrate is composed of small to medium-sized lymphocytes with irregular nuclei, condensed chromatin, and inconspicuous nucleoli (E) (hematoxylin-eosin, original magnification ×400). F and G, The atypical lymphocytes are uniformly positive for CD20 (F) (original magnification ×400) and cyclin-D1 (G) (original magnification ×400). H, L-selectin is also expressed by the lymphoma (original magnification ×400).
Reflectance Confocal Microscopy Features of Degos Disease

Degos disease, otherwise known as 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...