Self-healing eschar-like erythematous nodules

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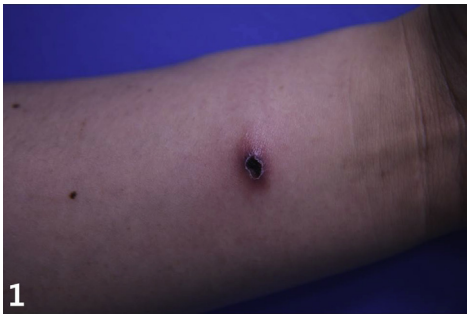

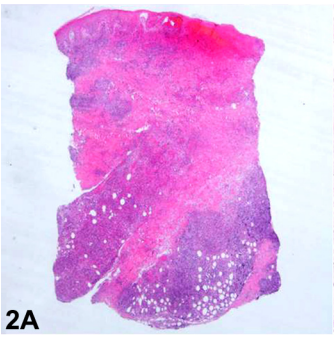

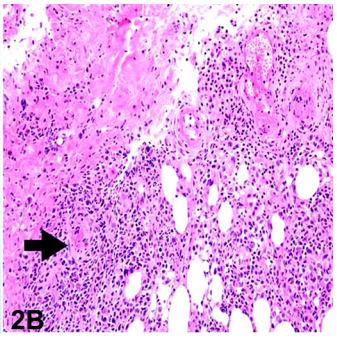

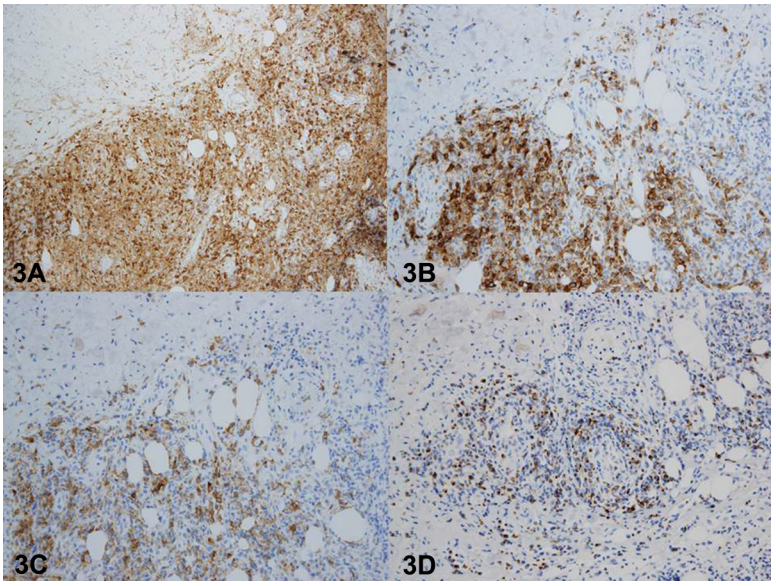

A 34-year-old man presented with a 4-year history of relapsing self-healing eschar-like erythematous nodules on his arms (Fig 1). The lesions would appear in crops of 2 to 3 lesions at a time. No lymphadenopathy or systemic symptoms were present, and a complete blood cell count and liver and kidney function tests were within normal limits.

Discussion: Histopathology of the lesion showed profound epidermal and superficial dermal necrosis (Fig 2, *A*) caused by vessel wall destruction (Fig 2, *B*) by atypical lymphoid cells (*arrow*). These cells were positive for CD3, CD4 (Fig 3, *A*), CD8, CD30 (Fig 3, *B*), CD56 (Fig 3, *C*), and granzyme B (Fig 3, *D*), but negative for CD20, CD79a, ALK, and Epstein–Barr virus–encoded RNA in situ hybridization. The Ki-67 labeling index was nearly 90%.

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Question 1: What is the most likely diagnosis?

A. Sweet syndrome

B. Mucormycosis

C. Nasal type extranodal natural killer/T-cell lymphoma

D. Cutaneous anaplastic large cell lymphoma

E. Lymphomatoid papulosis type E

A. Sweet syndrome—Incorrect. The lesions in patients with Sweet syndrome can be bullae or pustules that typically heal without scarring. The $CD30^+$ variants can be found, but neutrophils are usually the dominant infiltrate.

B. Mucormycosis—Incorrect. This infection develops as small erythematous macules, purpura, or erosions with eschar formation. It most commonly affects immunosuppressed patients, and no evidence of infections was identified in serologic and histopathologic evaluations.

C. Nasal type extranodal natural killer/T-cell lymphoma—Incorrect. This type is an aggressive lymphoma with angiocentric and angiodestructive infiltrates of $CD3^+$, $CD8^{+/-}$, $CD56^+$, and necrosis, but lack expression of CD30. In rare cases, a subset of cells in the extranodal natural killer/T-cell lymphoma, nasal type can express CD30. Moreover, it is linked to Epstein–Barr virus in virtually all cases.

D. Cutaneous anaplastic large cell lymphoma—Incorrect. The angiocentric variants of anaplastic large cell lymphoma manifest as solitary or grouped ulcerated nodules that do not spontaneous regress eventually lead to regional lymphadenopathy in 10% of patients.[1]

E. Lymphomatoid papulosis type E—Correct. Type E is recently introduced subtype of lymphomatoid papulosis (LyP), which presents with eschar-like lesions and is characterized by angiocentric invasion of dermal blood vessels by $CD30^+$ atypical lymphocytes, resulting in necrosis of the vessel walls. Because of the dense dermal infiltration of large atypical lymphocytes with high mitotic activity and angiodestructive invasion, it is challenging to distinguish from aggressive cutaneous lymphomas.[2]

Question 2: What type of lymphomatoid papulosis must be considered when atypical T cells express overt angioinvasion and angiodestruction?

A. Type A

B. Type B

C. Type C

D. Type D

E. Type E

A. Type A—Incorrect. This type is characterized by the presence of large pleomorphic or anaplastic $CD30^+$ T cells scattered in or in small clusters within the background of eosinophilic and neutrophilic granulocytes, histiocytes, and small lymphocytes.

B. Type B—Incorrect. This type shows epidermotropic infiltrates of small- to medium-sized lymphoid cells, with a variable extent of CD30 expression.

C. Type C—Incorrect. This type demonstrates that a nodular dense infiltrate of cohesive sheets of pleomorphic or anaplastic $CD30^+$ cells is present, and it usually contains only a few eosinophilic or neutrophilic granulocytes.

D. Type D—Incorrect. This type displays an epidermotropic infiltrate of $CD8^+$ and $CD30^+$ small- to medium-sized lymphoid cells.

E. Type E—Correct. This is a newly introduced subtype, characterized by oligolesional papules that evolve into necrotic eschar-like lesions with spontaneous regression and microscopically by typical angiocentric invasion of dermal blood vessels by $CD30^+$ atypical lymphocytes resulting in necrosis of the vessel walls.[2]

Question 3: Which immunohistochemical marker can differentiate type E lymphomatoid papulosis from other mimickers?

A. CD30

B. CD4

C. CD8

D. CD56

E. None

A. CD30—Incorrect. LyP is a primary cutaneous $CD30^+$ T cell lymphoproliferative disorder that is clinically characterized by a variable number of self-healing papulonodular lesions. This disorder has a waxing and waning course and represents the second most common form of cutaneous T-cell lymphoma.[2] Many subtypes of LyP express CD30.

B. CD4—Incorrect. Immunohistochemically, the neoplastic cells of LyP are typically $CD4^+$ lymphocytes.[2] Therefore, this marker does not distinguish type E LyP from the others.

C. CD8—Incorrect. LyP type E is characterized by oligolesional papules that rapidly ulcerate and evolve into large necrotic eschar-like lesions with a diameter of 1 to 4 cm and an angiocentric and angiodestructive infiltrate of small- to medium-sized atypical lymphocytes expressing CD30 and frequently CD8.[2] Type D may also have $CD8^+$ cells.

D. CD56—Incorrect. CD56, a monoclonal antibody against the neural cell adhesion molecule, was initially identified as a surface molecule of $CD16^+$ natural killer cells with the morphology of large granular lymphocytes. Although cutaneous lymphomas expressing CD56, such as skin infiltration of acute myeloid leukemia, nasal type extranodal natural killer/T-cell lymphoma, and blastic natural killer cell lymphoma, are generally characterized by a highly aggressive clinical course and poor prognosis,[3] a few cases of $CD56^+$ LyP type E, including our case, showed an excellent prognosis paradoxically.[2,4,5]

E. None—Correct. Unfortunately, there is not 1 single immunohistochemical marker that distinguishes type E LyP from the other subtypes. The clinical presentation of eschar-like lesions that wax and wane without associated lymphadenopathy are key to the diagnosis. Careful history taking, a comprehensive physical examination, and histopathologic examination with a full histochemical profile are key to the diagnosis.

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