Man with recurrent necrotic papules

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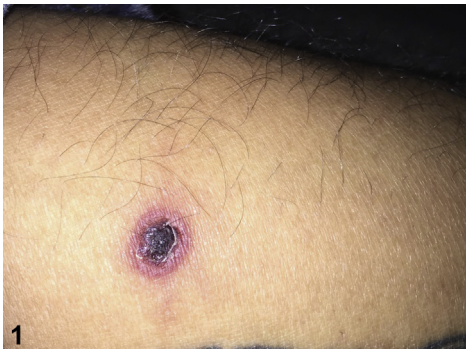

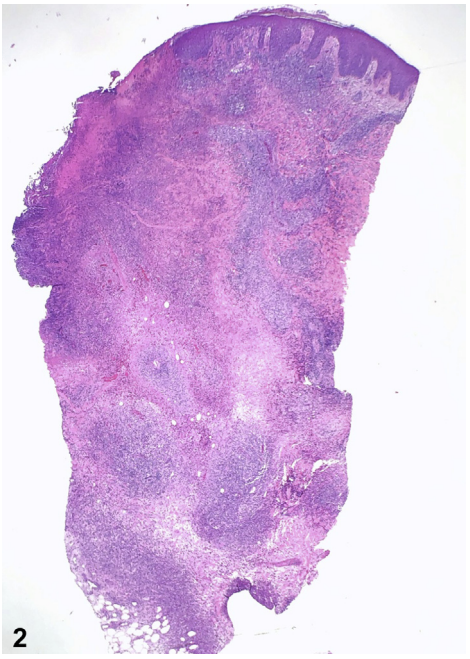

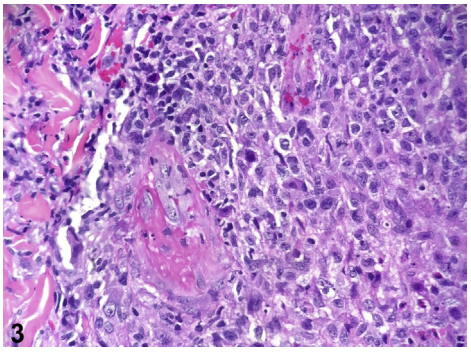

A 33-year-old man and 13-year-old-boy presented with lesions on the extremities and the back. The lesions began as erythematous nodules, became ulcerated with central eschar, and healed without scarring, only to recur in new locations (Fig 1). A biopsy revealed a diffuse, angiocentric, nodular, lymphocytic infiltrate of large atypical cells that coexpressed CD3, CD4, and CD30, associated with vascular thrombosis, necrosis, and

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ulceration (Figs 2 and 3). Results for further laboratory evaluation and evaluation for systemic involvement were negative. In the adult, new lesions continued to develop and resolve. At subsequent follow-up appointments, the child remained completely healthy, with no signs of recurrence.

Question 1: Which of the following is the most likely diagnosis?

A. Hodgkin lymphoma

B. Anaplastic large cell lymphoma

C. Extranodal natural killer/T-cell lymphoma, nasal type

D. Lymphomatoid papulosis (LyP) type A

E. LyP type E

Answers:

A. Hodgkin lymphoma—Incorrect. Although the Reed-Sternberg cell of Hodgkin lymphoma is $CD30^+$, the clinical description of recurrent lesions that spontaneously resolve is most consistent with LyP. Additionally, cutaneous involvement of Hodgkin lymphoma is rare.

B. Anaplastic large cell lymphoma—Incorrect. Although anaplastic large cell lymphoma is $CD30^+$, the distinguishing feature of LyP is that of recurrent lesions that spontaneously resolve.

C. Extranodal natural killer/T-cell lymphoma, nasal type—Incorrect. Extranodal natural killer/T-cell lymphoma, nasal type is angiocentric. However, the clinical and histologic descriptions are consistent with LyP.

D. Lymphomatoid papulosis (LyP) type A—Incorrect. The clinical presentation of recurrent and relapsing cutaneous lesions is observed in all subtypes of LyP. Type A is characterized by a small number of $CD30^+$ T cells scattered within a dense lymphoid infiltrate without the angiocentric or angioinvasive features that define LyP type E.[1]

E. LyP type E—Correct. LyP type E is a rare variant of LyP[2] and shares the characteristic clinical features of recurrent crops of ulcerative nodules that spontaneously resolve. Histopathologic examination reveals an angioinvasive infiltrate of atypical large $CD30^+$ T cells.

Question 2: After initial diagnosis, which of the following would be a reasonable next step in management?

A. Lymph node biopsy

B. Complete blood cell count and chest radiography

C. Bone marrow biopsy

D. Intralesional interferon alfa

E. Multiagent chemotherapy

Answers:

A. Lymph node biopsy—Incorrect. A lymph node biopsy would be reasonable if a clinically enlarged lymph node greater than 1.5 cm were palpable or detected on radiologic imaging.

B. Complete blood cell count and chest radiography—Correct. Patients with LyP, including LyP type E, often have an indolent and spontaneously resolving course. Minimally invasive studies to rule out systemic lymphomas may be considered and include complete blood cell counts with differential diagnosis, blood chemistry, or lactate dehydrogenase. Second lymphoid malignancies can occur concomitantly or precede the onset of LyP.[3-5]

C. Bone marrow biopsy—Incorrect. A bone marrow biopsy may be considered for individuals who have evidence of bone marrow involvement, as observed in initial serologic studies.

D. Intralesional interferon alfa—Incorrect. LyP subtypes, including LyP type E, do not routinely require therapy. If patients have extensive or scarring lesions, topical steroids, phototherapy, or methotrexate may be useful initial interventions.

E. Multiagent chemotherapy—Incorrect. Multiagent chemotherapy is not indicated for LyP and should be avoided because of an inability to provide a durable response, as well as adverse effects and long-term complications.[4]

Question 3: Which of the following is most specific for this disease?

A. Erythematous nodules that ulcerate with a central eschar

B. Diffuse angiocentric nodular lymphocytic infiltrate of large atypical cells with thrombotic vascular changes

C. Resolving and recurring lesions with a $CD30^+$ infiltrate

D. Prompt resolution after topical or systemic antibiotics

E. Lesions that do not recur after resolution

Answers:

A. Erythematous nodules that ulcerate with a central eschar—Incorrect. The finding of erythematous nodules that progress to ulcerated central eschar is nonspecific and is observed in a wide variety of dermatologic conditions such as pressure sores, nonmelanoma skin cancers, pityriasis lichenoides et varioliformis acuta, and cutaneous anthrax.

B. Diffuse angiocentric nodular lymphocytic infiltrate of large atypical cells with thrombotic vascular changes—Incorrect. The angiocentric histologic features of LyP type E often mimic aggressive lymphomas, including gamma/delta T-cell and extranodal natural killer/T-cell lymphoma, nasal type. The diagnosis of LyP type E can be suspected by histologic findings but requires immunohistochemical stains and clinical correlation.

C. Resolving and recurring lesions with a $CD30^+$ infiltrate—Correct. Immunohistochemical staining demonstrating $CD30^+$ cells combined with clinically resolving and recurrent lesions is required to diagnose LyP.[1]

D. Prompt resolution after topical or systemic antibiotics—Incorrect. The finding of lesions that resolve after antibiotic therapy is nonspecific and may be observed in infectious or inflammatory conditions. The lesions of LyP type E are known to have a waxing and a waning course, which makes the resolution of the lesions more likely caused by the natural progression of the disease rather than from treatment.

E. Lesions that do not recur after resolution—Incorrect. The self-resolving and recurrent nature of the disease is the most characteristic clinical feature. Lack of recurrence after resolution is inconsistent with a diagnosis of LyP.

Abbreviation used:

LyP: lymphomatoid papulosis

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