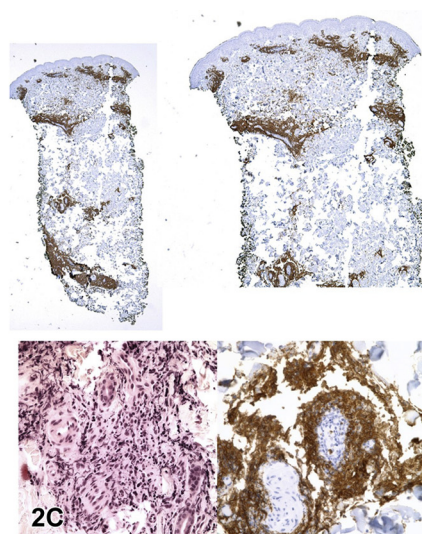
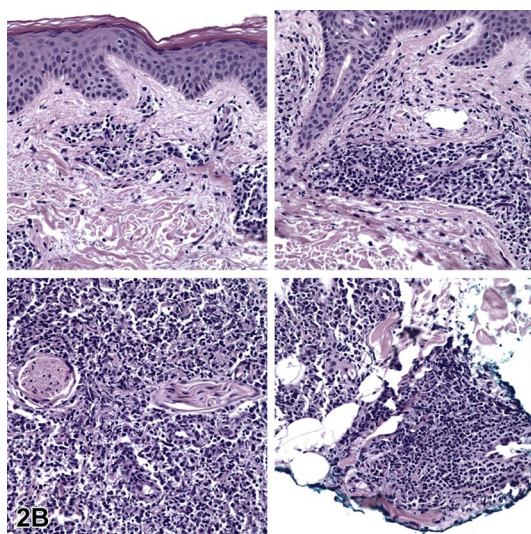
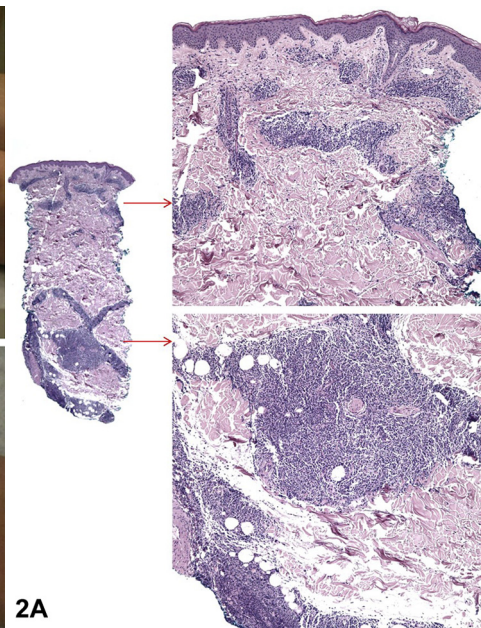


Figurate erythema for 20 years



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CASE

A 49-year-old man from New England presented with a 20-year history of pink, nonscaly, arcuate and figurate plaques on the right thigh, unresponsive to midpotency topical steroids (Fig 1, A). In the year before presentation, 2 nodules developed within the plaques (Fig 1, B). The patient denied having fevers, night sweats, pruritus, or weight loss. He denied history of tick bites. No lymphadenopathy was appreciated. Biopsies (Fig 2, A-C) of the new nodule and longstanding plaque were obtained.

Question 1: What is the most likely diagnosis?

- A. Granuloma annulare (GA)
- B. B-cell lymphoma
- C. Erythema annulare centrifugum (EAC)
- D. Erythema migrans (EM)
- E. Erythema gyratum repens (EGR)

Answers:

A. GA — Incorrect. Although GA commonly presents as coalescing nonscaly papules forming annular plaques, histopathology results show granulomatous inflammation comprising palisading histiocytes and mucin.

B. B-cell lymphoma — Correct. This is a case of diffuse large B-cell lymphoma (DLBCL). If the skin is involved, DLBCL commonly manifests on the legs as papules, nodules, or indurated plaques.¹ It is suggested that annular-appearing DLBCL may represent transformation of an indolent follicular lymphoma to DLBCL,² which may have occurred here given the prolonged clinical course. Histopathology (Fig 2, A) results show papillary to reticular dermal perivascular lymphocytic infiltrate. At higher magnification (Fig 2, B), the infiltrate comprises intermediate- to large-sized atypical lymphocytes with round to irregular nuclei, dispersed vesicular chromatin, occasional single nucleoli, and moderate amounts of pale eosinophilic cytoplasm. Immunohistochemistry (Fig 2, C) shows CD20⁺ B cells that stain positively for Bcl-2, Bcl-6, and MUM-1, and negative for CD10. The MIB-1 (Ki-67) proliferative index is approximately 80%. Staging for this disease revealed no extracutaneous disease, thus the patient was thought to have primary cutaneous DLBCL-leg type (PCDLBCL-LT).

C. EAC — Incorrect. EAC may have a chronic course and shows tight perivascular cuffing, as in this case, but would otherwise have normal-appearing lymphocytes.

D. EM — Incorrect. EM is a common figurate presentation for Lyme disease in New England; however, nodularity is not a feature of EM, nor is

the lengthy course. Histopathology findings are nonspecific with perivascular lymphocytic infiltrate and occasional plasma cells.

E. EGR — Incorrect. EGR is a paraneoplastic eruption that presents as diffuse erythematous figurate bands with a “wood-grained” appearance. Histopathology findings are nonspecific with mild hyperkeratosis, parakeratosis, acanthosis, and spongiosis.

Question 2: A subset of PCDLBCL-LT patients with cutaneous presentation may progress to have extracutaneous disease. To which of the sites is PCDLBCL-LT most likely to spread extracutaneously?

- A. Liver
- B. Lungs
- C. Regional lymph nodes
- D. Spleen
- E. Small intestine

Answers:

A. Liver — Incorrect. Although lymphomas including PCDLBCL-LT can spread to the liver, this is not the most common extracutaneous site of spread PCDLBCL-LT.

B. Lungs — Incorrect. Although lymphomas, including PCDLBCL-LT, can spread to the lungs, this is not the most common extracutaneous site of spread PCDLBCL-LT.

C. Regional lymph nodes — Correct. Regional lymph nodes are a common site to which PCDLBCL-LT can spread extracutaneously, typically months after initial cutaneous presentation.³

D. Spleen — Incorrect. Although lymphomas including PCDLBCL-LT can spread to the spleen, this is not the most common extracutaneous site of spread PCDLBCL-LT.

E. Small intestine — Incorrect. Although lymphomas including PCDLBCL-LT can spread to the small intestine, this is not the most common extracutaneous site of spread for PCDLBCL-LT.

Question 3: What is the current standard treatment for DLBCL/PCDLBCL-LT?

- A. ABVD regimen
- B. Locoregional therapy
- C. Intralesional corticosteroids
- D. R-CHOP regimen
- E. Stanford V regimen

Answers:

A. ABVD regimen — Incorrect. ABVD regimen (doxorubicin, bleomycin, vinblastine, dacarbazine) is a common regimen used in the treatment of Hodgkin lymphoma.

B. Locoregional therapy — Incorrect. Because of the clinically aggressive behavior of DLBCL, it is recommended patients receive systemic chemotherapy rather than local treatment such as locoregional therapy, which has shown success in treating patients with indolent solitary or regional primary cutaneous B-cell lymphomas such as primary cutaneous marginal zone lymphoma or primary cutaneous follicle center B-cell lymphoma.

C. Intralesional corticosteroids — Incorrect. Because of the clinically aggressive behavior of DLBCL, it is recommended patients receive systemic chemotherapy rather than local treatment such as intralesional corticosteroids, which has shown success in treating patients with indolent solitary or regional primary cutaneous B-cell lymphomas such as primary cutaneous marginal zone lymphoma or primary cutaneous follicle center B-cell lymphoma.

D. R-CHOP regimen — Correct. These lymphomas have more aggressive clinical behavior, thus standard recommendations for patients is multidrug therapy with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisone).^{4,5} The patient underwent 6 cycles of R-CHOP plus radiation resulting in complete radiographic remission and resolution of all visible cutaneous findings.

E. Stanford V regimen — Incorrect. Stanford V regimen (doxorubicin, mechlorethamine, vincristine, vinblastine, bleomycin, etoposide, prednisone) is a common regimen used to treat Hodgkin lymphoma.

Abbreviations used:

DLBCL: diffuse large B-cell lymphoma

EAC: erythema annulare centrifugum

EGR: erythema gyratum repens

EM: erythema migrans

GA: granuloma annulare

PCDLBCL-LT: primary cutaneous DLBCL-leg type

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