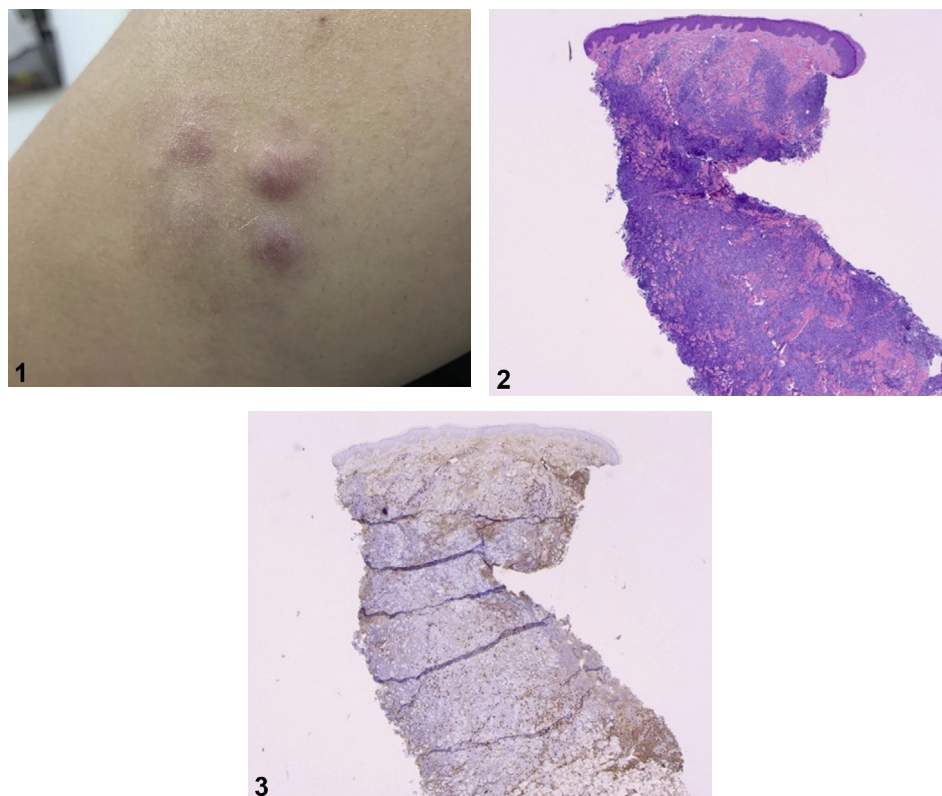


Asymptomatic pink nodules on a 28-year-old patient



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A 28-year-old Asian-American man with no past medical history presented with enlarging pink nodules and skin texture changes on the left triceps lasting for approximately 1 year (Fig 1). The patient denied any previous trauma or injury to the site. The patient's symptoms and history were reviewed extensively. Physical exam revealed no lymphadenopathy. A punch biopsy showed a dense lymphoid infiltrate consisting of small to medium-sized lymphocytes containing abundant plasma cells (Fig 2). A panel of immunohistochemistry stains demonstrated many CD3⁺ T cells and vast collections of CD20⁺ B cells, with a much greater number of T cells.

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The plasma cells expressed lambda light chain to a much greater degree than kappa light chain (Fig 3). The patient was referred to oncology for further work up.

Question 1: What is the most likely diagnosis if whole body positron emission tomography scan were unremarkable?

- A. Primary cutaneous mantle cell lymphoma
- B. Primary cutaneous marginal zone lymphoma (PCMZL)
- C. Cutaneous pseudolymphoma
- D. Lymphomatoid papulosis
- E. Primary cutaneous follicular center lymphoma

Answers:

A. Primary cutaneous mantle cell lymphoma – Incorrect. Mantle cell lymphoma typically stains CD5⁺ and CD10⁻. It is associated with a t(11,14) translocation.¹

B. Primary Cutaneous Marginal Zone Lymphoma (PCMZL) – Correct. PCMZL accounts for approximately 40% of all cutaneous B-cell lymphomas and approximately 10% of all primary cutaneous lymphomas. It typically presents as pink to red nodules to plaques on the arms or trunk. It is a B-cell lymphoma with numerous T cells.¹

C. Cutaneous pseudolymphoma – Incorrect. The kappa-to-lambda ratio is normally two-thirds kappa to one-third lambda. The imbalance of this ratio suggests that B-cell clonality is present. In pseudolymphoma, there is no disproportionate change in the light chains. Thus, a true lymphoma is more likely in this patient's case.^{2,3}

D. Lymphomatoid papulosis – Incorrect. This is a benign proliferation of T cells, presenting as recurrent, self-resolving erythematous papules or nodules. It stains CD30⁺ and CD3⁺, CD4⁺, and CD5⁺ (common T-cell markers).³

E. Primary cutaneous follicular center lymphoma – Incorrect. Primary cutaneous follicular center lymphoma typically stains CD10⁺ and Bcl-6⁺ and Bcl-2⁻. A t(14,18) translocation is associated with the systemic form predominantly. The head and neck are most commonly affected.³

Question 2: What is true regarding the dermatopathologic findings in PCMZL?

A. Histologic findings often show nodular and diffuse infiltrates of lymphocytes in the dermis.

Follicles contain neoplastic cells in the periphery region that lack the polarity of germinal center with a reduced and sometimes absent mantle zone. There are reduced tangible body macrophages in the follicles.

B. Histologic findings often show nodular and diffuse infiltrates of monocytoïd B cells in the dermis and subcutaneous region. Lymphoid nodules contain central darker normal cells and paler peripheral neoplastic marginal zone cells. Numerous plasma cells are seen and some with Dutcher bodies. Intracytoplasmic monoclonal immunoglobulin kappa or lambda restriction is often seen.

C. Histologic findings often show dense and diffuse sheets of infiltrates with large and round lymphocytes with prominent nucleoli. A grenz zone is often appreciated.

D. Histologic findings often show diffuse and sheets of myeloid blasts in the dermis with a grenz zone present. The histologic pattern varies with type. Neoplastic infiltrates could be perivascular, interstitial, nodular, or diffuse. A thorough history and clinical presentation would be helpful to establish the correct diagnosis.

E. Proliferation of large atypical B cells with large cytoplasm and prominent nucleoli are observed in dilated blood vessels in the dermis and subcutaneous tissues.

Answers:

A. Histologic findings often show nodular and diffuse infiltrates of lymphocytes in the dermis. Follicles contain neoplastic cells in the periphery region that lack the polarity of the germinal center with a reduced and sometimes absent mantle zone. There are reduced tangible body macrophages in the follicles. – Incorrect. This is the dermatopathology description of primary cutaneous follicle center cell lymphoma.

B. Histologic findings often show nodular and diffuse infiltrates of monocytoïd B cells in the dermis and subcutaneous region. Lymphoid nodules contain central darker normal cells and paler peripheral neoplastic marginal zone cells. Numerous plasma cells are seen and some with Dutcher bodies. Intracytoplasmic monoclonal immunoglobulin kappa or lambda restriction is often seen. – Correct. This is the dermatopathology description of PCMZL.

C. Histologic findings often show dense and diffuse sheets of infiltrates with large and round lymphocytes with prominent nucleoli. A grenz zone is often appreciated. — Incorrect. This is the dermatopathology description of cutaneous diffuse large B-cell lymphoma, leg type.

D. Histologic findings often show diffuse and sheets of myeloid blasts in the dermis with a grenz zone present. The histologic pattern varies with type. Neoplastic infiltrates could be perivascular, interstitial, nodular, or diffuse. A thorough history and clinical presentation would be helpful to establish the correct diagnosis. — Incorrect. This is the dermatopathology description of leukemia cutis.

E. Proliferation of large atypical B cells with large cytoplasm and prominent nucleoli are observed in dilated blood vessels in the dermis and subcutaneous tissues. — Incorrect. This is the dermatopathology description of intravascular diffuse large B-cell lymphoma.

Question 3: Which of the following is true regarding this diagnosis?

A. Immunohistochemistry reveals CD5⁺, CD10⁺, Bcl-6⁺, Bcl-2⁻, CD20⁻, and CD79a⁻.

B. Immunohistochemistry reveals CD20⁺, CD5⁺, CD10⁺, Bcl-6⁺, Bcl-2⁻, and CD79a⁻.

C. The prognosis of PCMZL is poor with a high rate of metastasis to internal organs.

D. The first-line treatment of PCMZL is wide excision with sentinel lymph node mapping.

E. In comparison to PCMZL, patients with marginal zone lymphoma are usually older, have systemic symptoms, show expression of IgM, and have a predominant Th1 cytokine inflammatory background.

Answers:

A. Immunohistochemistry usually reveals CD5⁺, CD10⁺, Bcl-6⁺, Bcl-2⁻, CD20⁻, and CD79a⁻. — Incorrect. CD5 indicates a T-cell lymphoma. Our patient's diagnosis is PCMZL, which is of B-cell origin.

B. Immunohistochemistry usually reveals CD20⁺, CD5⁺, CD10⁺, Bcl-6⁺, Bcl-2⁻ and CD79a⁻. — Incorrect.

Bcl-2 would be positive in PCMZL because it is an oncogene responsible for preventing apoptosis. Furthermore, CD79a is a marker for plasma cells and B cells, so it would also be positive.

C. The prognosis of PCMZL is poor with a high rate of metastasis to internal organs. — Incorrect. PCMZL is a low-grade malignancy and the prognosis is excellent. Studies show low numbers of cases with extracutaneous disease progression from PCMZL. The five-year survival rate is 98%-100%.

D. The first-line treatment of PCMZL is wide excision with sentinel lymph node mapping. — Incorrect. This is the treatment approach for primary merkel cell carcinoma. In general, the treatment approach to localized PCMZL is local radiation therapy with a radiation dose of 24 Gy. Surgical excisions could be considered for patients with localized lesions and anatomical locations not amenable to radiation therapy.

E. In comparison to PCMZL, patients with marginal zone lymphoma are usually older, have systemic symptoms, show expression of IgM, and have a predominant Th1 cytokine inflammatory background. — Correct. Marginal zone lymphoma patients tend to be older, have systemic symptoms and show expression of IgM. Patients with PCMZL tend to be younger, with no systemic systems, express IgG, IgA, or IgE, and have a predominant of Th1 cytokine inflammatory background.

Abbreviation used:

PCMZL: primary cutaneous marginal zone lymphoma

Conflicts of interest

None disclosed.

REFERENCES

1. Kempf W, Denisjuk N, Kerl K, Cozzio A, Sander C. Primary cutaneous B-cell lymphomas. *J Dtsch Dermatol Ges.* 2012;10(1):12-22.
2. Elston D, Ferringer T, Ko C, Peckham S, High W, DiCaudo D. *Dermatopathology.* 3rd Edition. Elsevier; 2018.
3. Lima M. Cutaneous primary B-cell lymphomas: from diagnosis to treatment. *An Bras Dermatol.* 2015;90(5):687-706.