A Case of Primary Cutaneous Marginal Zone Lymphoma

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Introduction

Primary cutaneous marginal zone B-cell lymphoma (PCMZL) is a form of primary cutaneous B-cell lymphoma (PCBCL) that typically manifests in adulthood with an incidence of fewer than one case per million people annually (1). Clinically, this disease presents as asymptomatic, erythematous papules, plaques, or nodules, located on the trunk, extremities, head, or neck (2,3). Histopathological examination reveals patchy, nodular, or diffuse lymphocytic infiltrates in the dermis and subcutaneous fat. Immunohistochemistry (IHC) shows positivity for CD20, CD79a, and BCL-2, and negativity for CD10 and BCL6 (2,3). After the diagnosis is made, systemic involvement needs to be ruled out before proceeding with management options such as surgical excision, radiation therapy, or topical modalities. Relapse rates are high, and patients need to followed longitudinally by dermatology and oncology.

Case Presentation

A 76-year-old Caucasian male with a history of actinic keratosis and basal and squamous cell carcinomas presented to the dermatology clinic with two asymptomatic well-circumscribed, pink to erythematous papules approximately 0.7mm in size on the upper back and left upper arm (Figure 1) that had been present for several months. The patient reported that he initially developed 3 spots on his left upper arm but two spots resolved without intervention and one persisted. He had no systemic symptoms, recent medication changes, or mucosal involvement.

Saucerization biopsy showed a dense nodular proliferation of atypical lymphocytes in the dermis, and IHC exhibited strong positivity for CD20, CD79a, and BCL-2, confirming the diagnosis of cutaneous marginal zone lymphoma. The patient was referred to oncology for further workup and recommendations.

Extensive laboratory studies were remarkable for elevated serum kappa light chains but were otherwise normal. Positron emission tomography-computed tomography (PET-CT) scan showed no metabolically active disease, and after excluding systemic involvement, the diagnosis of primary cutaneous marginal zone lymphoma was confirmed. The patient was scheduled for excisions of both sites with three millimeter margins. On the day of surgery, the patient revealed a new onset, painless erythematous nodule on the left hip (Figure 2). Biopsy demonstrated dense dermal infiltration of atypical lymphocytes showing perivascular, periadnexal, and interstitial extension (Figure 3) with strong positivity for CD79a and CD20, and some expression of BCL2 (Figure 4), consistent with a new cutaneous marginal zone lymphoma, which was also excised with clear margins. The patient is being followed by oncology and dermatology every 3-6 months.

Clinical Images

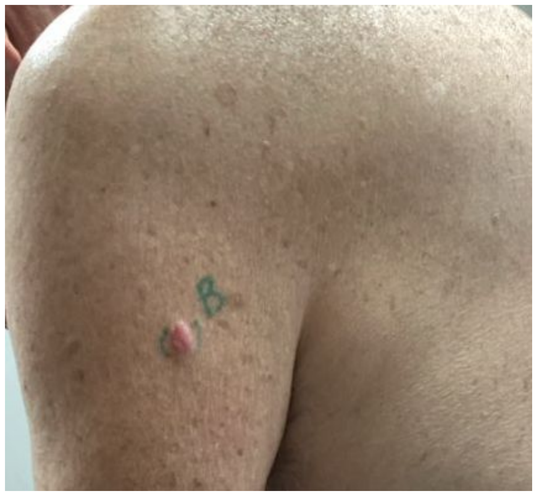


Figure 1. Well-circumscribed erythematous-pink papule on patient's left posterior upper arm.

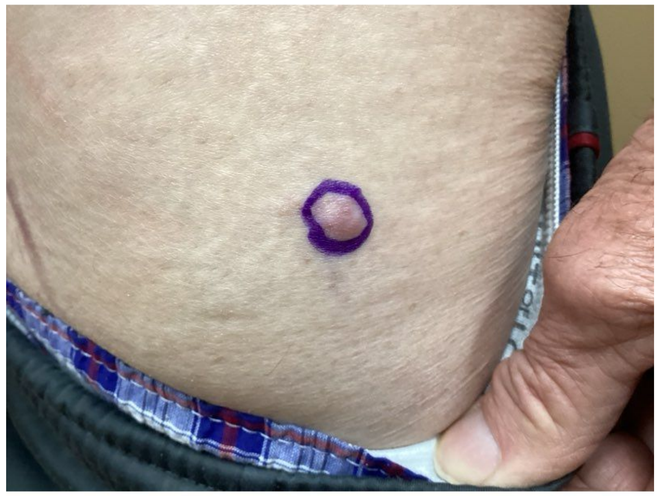
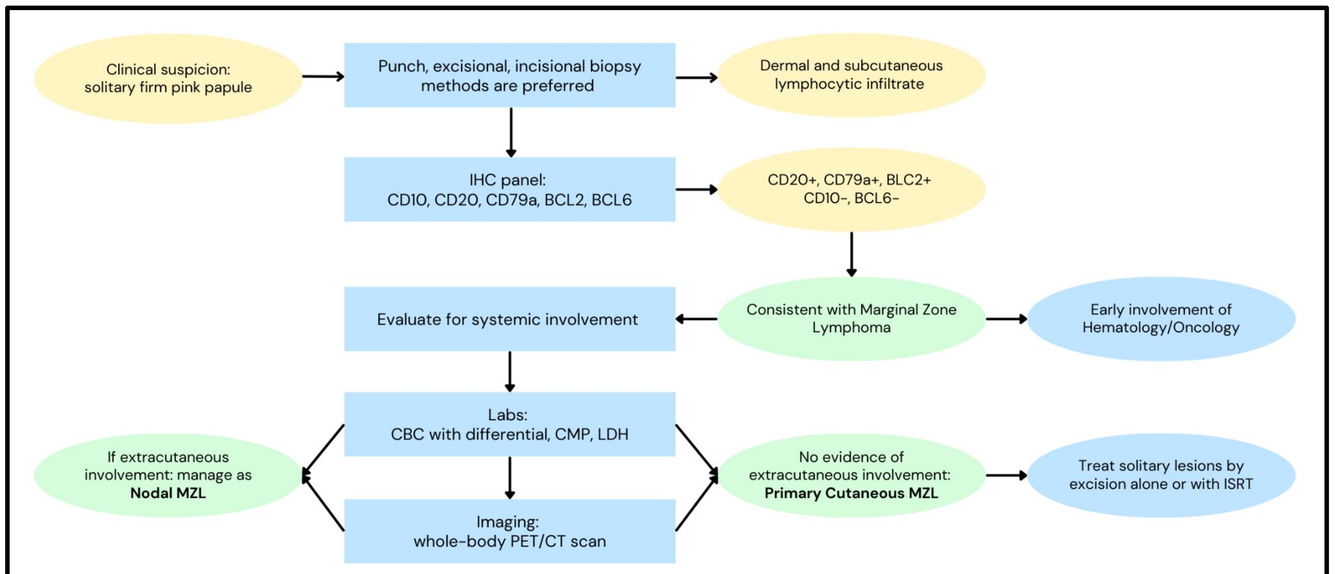


Figure 2. Well-demarcated erythematous to violaceous nodule on patient's left anterior thigh



Discussion

Primary cutaneous marginal zone lymphoma (PCMZL) is the second most common primary cutaneous B-cell lymphoma (PCBCL) accounting for 24-31% of cases (2). The diagnosis is made via biopsy, preferably using a punch, incisional, or excisional technique, while avoiding the shave method as it may miss primarily dermal infiltrates. Immunohistochemistry stains positive for CD20, CD79a, and BCL2, and stain negative for CD10 and BCL6 (2,3). After a histopathologic diagnosis is made, patients must undergo a comprehensive workup to rule out systemic involvement including a history and physical exam, total body skin exam, laboratory studies (CBC with differential, CMP, and LDH), imaging (whole-body PET/CT scan), and referral to oncology (2). Additionally, if indicated, a more comprehensive workup may include peripheral blood flow cytometry, bone marrow biopsy, SPEP/quantitative immunoglobulins, HIV, Hepatitis B and C testing, and fertility preservation (2). In our case, the patient was referred to oncology and underwent extensive laboratory testing and imaging and once systemic involvement was excluded, the diagnosis was confirmed. Systemic symptoms and extracutaneous involvement are rare findings and according to a 2013 retrospective analysis of 137 patients, Servitje et al. reported that only 4% of cases showed extracutaneous dissemination (4). After confirmation that the disease is limited to the skin, additional imaging during follow-up is not needed, only clinical follow-up is recommended at least every 6 months or at the onset of new lesions or symptoms (2).

Pathology

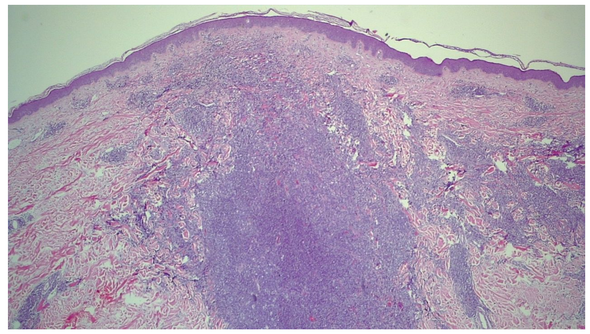


Figure 3. H&E of specimen demonstrating dense nodular proliferation of atypical lymphocytes in superficial and deep dermis consistent with atypical lymphoid infiltrate consistent with cutaneous marginal zone lymphoma.

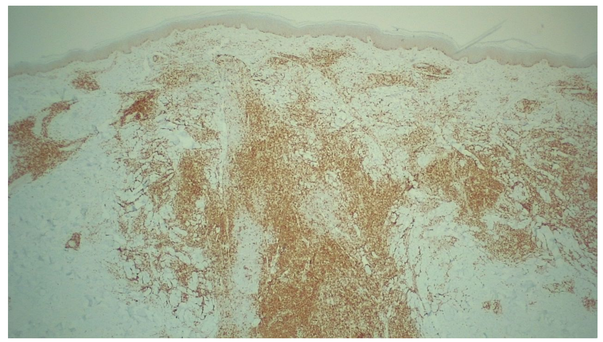


Figure 4. IHC strong positivity for Bcl-2 within lymphocytic infiltrate of left anterior hip

Discussion cont.

The preferred initial treatment for small, solitary lesions or early-stage PCMZL, such as in our case, is excision alone or involved-site radiation therapy (ISRT), with or without excision (2). Other treatment options include observation, and skin-directed therapies such as topical corticosteroids, imiquimod, bexarotene, nitrogen mustard, intralesional steroids, or rituximab. Despite different treatment modalities, disease relapse occurs in the majority of patients and those with multiple lesions have higher relapse rates compared to those with solitary lesions (4). However, PCMZL has an excellent prognosis with a 5-year survival of approximately 99% (2). Dermatologists and other healthcare providers should know this rare yet indolent cutaneous neoplasm that requires long-term follow-up. This case underscores the importance of early detection, careful monitoring, and individualized workup and treatment in PCMZL to optimize outcomes and patient care.

References

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4. Servitje O, Muniesa C, Benavente Y, et al. Primary cutaneous marginal zone B-cell lymphoma: Response to treatment and disease-free survival in a series of 137 patients. *J Am Acad Dermatol*. 2013;69(3):727-734.