SKIN

BRIEF ARTICLE

Unusual Presentation of Marginal Zone Lymphoma

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ABSTRACT

Primary cutaneous B-cell lymphomas (pcBCLs) are non-Hodgkin lymphomas that present with exclusive cutaneous disease at the time of diagnosis. The lesions may be indolent, subtle, or waxing and waning, making the diagnosis challenging. The ear is an uncommon location, and its frequency of involvement remains unknown. A retrospective analysis of a single patient with primary cutaneous marginal zone lymphoma of the ear. The patient was a 44-year-old male who presented with erythematous tender nodules on the left superior posterior helix. Shave biopsy revealed mixed dermal lymphoid infiltrate with eosinophils and telangiectasias. The patient was diagnosed with angiolymphoid hyperplasia with eosinophilia. Multiple treatment regiments were trialed but failed due to lack of therapeutic response, relapse, and clinical progression. The patient ultimately underwent Mohs surgery with biopsy findings that demonstrated CD-20 positive cells. Evaluation with B-cell histopathology and gene rearrangement studies confirmed the diagnosis of marginal zone lymphoma. This case report shows a unique presentation of marginal zone lymphoma of the ear, initially diagnosed as angiolymphoid hyperplasia with eosinophilia. Our findings demonstrate a relatively rare presentation for this disease and highlight the challenges in diagnosing primary cutaneous marginal zone lymphoma.

INTRODUCTION

Primary cutaneous B-cell lymphomas (pcBCLs) are non-Hodgkin lymphomas that present on the skin, without evidence of extracutaneous disease at the time of diagnosis. Primary cutaneous marginal zone lymphoma (PCMZL) is one of the most common classifications of pcBCLs.[1] It presents as pink to red papules, pustules, or nodules, that may be either solitary or grouped. The lesions may be indolent, subtle, or waxing and waning, making the diagnosis challenging.[2] The most frequent locations include the trunk, arms, head, or neck. The ear is an uncommon location, and its frequency of involvement remains unknown.[1,3,4] The following case report shows a unique presentation of PCMZL with exclusive concurrent bilateral auricular involvement.

CASE REPORT

A 44-year-old male with a history of untreated stage 3 chronic lymphocytic leukemia (small cell), presented to the dermatology clinic for a 4-month history of pink to erythematous

tender nodules on the helices (**Figures 1 and 2**). A shave biopsy was performed showing infiltrate with eosinophils and telangiectasias, compatible with angiolymphoid hyperplasia with eosinophilia. Based on the clinical presentation and initial biopsy results, a diagnosis of angiolymphoid hyperplasia with eosinophilia was suggested but not confirmed.

The patient returned to the clinic several months later with recurrence and involvement of left and right superior posterior helices. He was subsequently treated with intralesional triamcinolone, betamethasone dipropionate 0.05% cream, and pulsed dye laser treatment. Despite initial improvement, the patient returned with recurrent, worsening lesions. He displayed 9 lesions with involvement of bilateral ear lobes and helices. After being lost to follow-up for a period of several years, the patient presented once again with no improvement and with significantly worsening pain. After failing laser treatment, the decision to pursue Mohs surgery for removal of the nodules was made. Histopathologic sections from additional biopsies demonstrated a superficial and deep dense lymphoid infiltrate consisting of small uniform-appearing lymphocytes with a well-defined grenz zone and a histologic predominance of CD-20 positive cells (**Figures 3 and 4**). There were no vascular irregularities or eosinophilia noted. Staining for Bcl-2 was diffusely positive, and Bcl-6 showed weak positive patchy staining. A CD-10 stain was negative, while CD-31 highlighted background blood vessels. Kappa and lambda light chains did not support clear cut evidence of clonality, so B-cell gene rearrangement studies were performed. These were positive which led to the diagnosis of a marginal zone lymphoma, and the patient was subsequently referred to oncology.

DISCUSSION

This report describes an unusual clinical presentation of marginal zone lymphoma. Marginal zone lymphoma is a non-Hodgkin, B-cell lymphoma, that originates in the post-germinal marginal zone. It contains B cells markers such as CD19, CD20, CD22, and notably lacks CD5, CD10, and CD23.[5] Biopsy results demonstrate polymorphous infiltrate of small lymphocytes, reactive T cells, lymphoplasmacytic cells, and plasma cells.[2,6] Although uncommon, the diagnosis of marginal zone lymphoma with eosinophilia can be made.

Based on clinical presentation, the initial differential diagnosis included angiolymphoid hyperplasia with eosinophilia (ALHE), cutaneous chronic lymphocytic leukemia (CLL), and cutaneous mantle cell lymphoma. Clinical manifestations of ALHE include pink to brown solitary, firm, dome-shaped papules or nodules that may appear on the postauricular area, face or scalp. Typical distribution comprises multiple areas on the head, neck, and ear; exclusive bilateral postauricular involvement would be uncommon.[7] Histopathology includes epithelioid-like cells with abundant clear cytoplasm, and rarely displays cellular mitoses or atypia.[8]

Cutaneous involvement of CLL was included in the differential as a result of its “cauliflower-ear” appearance, similar in display to this patient’s lesions. The patient respectively had a history of untreated stage 3 chronic lymphocytic leukemia. Diagnosis of CLL of involves CD5 and CD23 positivity, both of which were not present in our biopsy results.[9] Mantle cell lymphoma was considered, as it displays a wide variety of presentations. It contains a predilection for male sex and frequently presents on the face as papules,

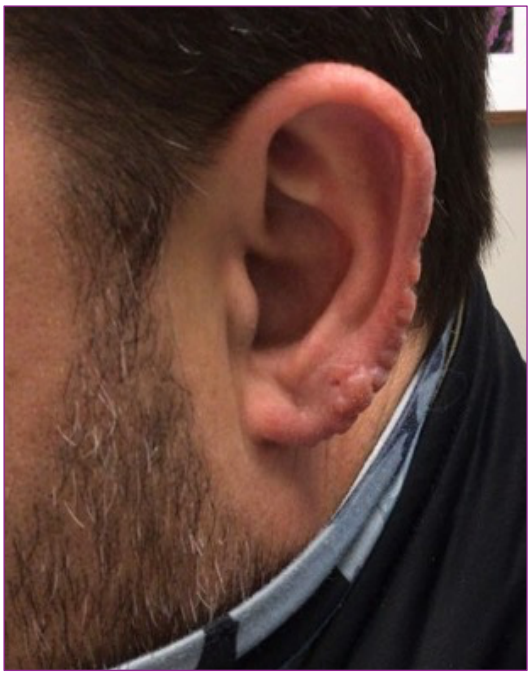

Figure 1. Pink to erythematous nodules on the left helix.

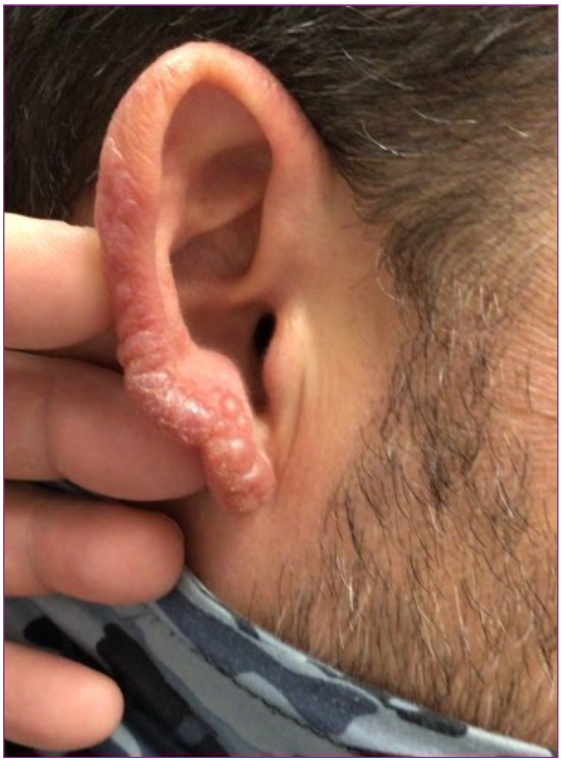

Figure 2. Pink to erythematous nodule on the right helix.

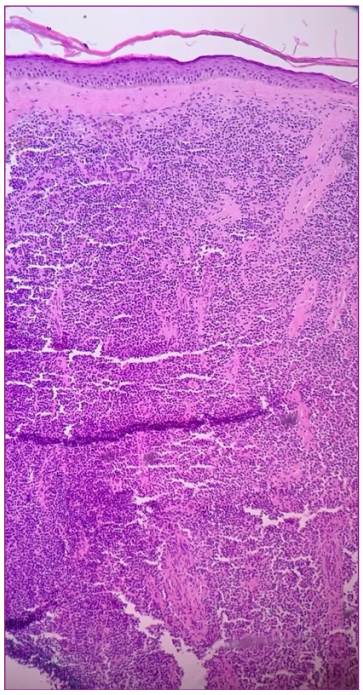

Figure 3. A superficial and deep dense lymphoid infiltrate consisting of small uniform-appearing lymphocytes and a well-defined grenz zone. (Hematoxylin and eosin, 10x).

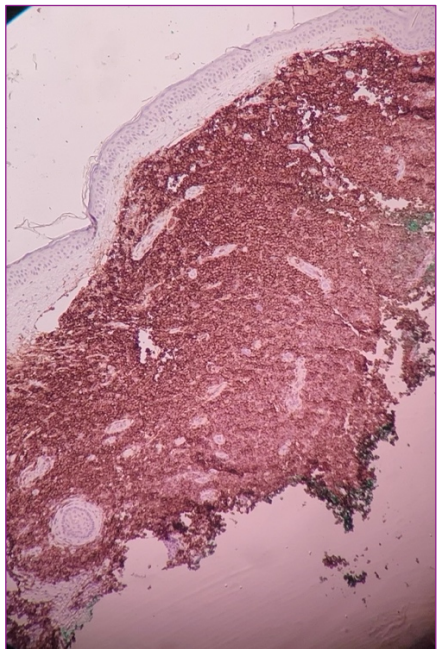

Figure 4. A dense lymphoid infiltrate consisting predominantly of CD20-positive B-cells. (CD20 immunostain, 10x)

plaques, or subcutaneous nodules.[10] It is identified though CD20 and cyclin D1 positivity. Although CD20 positivity was present on biopsy, cyclin D1 was not investigated. Immunophenotypically, CLL and mantle cell lymphoma display several overlapping features with marginal zone lymphoma that were seen in this report. This includes positive CD20 and BCL-2, as well as negative CD10. Ultimately, B-cell gene rearrangement studies led to diagnosis of marginal zone rather than cutaneous chronic lymphocytic or mantle cell lymphoma.

Local radiotherapy or surgical excision are the main treatments for PCMZL.[11] Chlorambucil therapy and radiotherapy are effective for treating PCMZL with multifocal lesions.[11]

CONCLUSION

This case describes an unusual but indolent presentation of marginal zone lymphoma. Its ambiguous clinical features led to delays in disease diagnosis, management, and treatment. As a result, clinicians should perform more thorough investigation in patients presenting with cutaneous features similar to those described in this report.

Consent: Consent for the publication of all patient photographs and medical information was provided by the authors at the time of article submission to the journal stating that all patients gave consent for their photographs and medical information to be published in print and online and with the understanding that this information may be publicly available.

Conflict of Interest Disclosures: None

Funding: None

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