SKIN

BRIEF ARTICLE

Primary Cutaneous Marginal Zone Lymphoma: An Interesting Presentation

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ABSTRACT

Primary cutaneous marginal zone lymphoma (PCMZL) is a subtype of extranodal marginal zone B-cell lymphoma which may involve mucosal sites. Approximately 8% of non-Hodgkin lymphomas can be classified as marginal zone lymphomas. PCMZL presents more frequently in the fifth and sixth decades as red papules, plaques, or localized nodules typically located on the trunk, arms, or head. Male patients present twice as often as female patients, and the median age is 55 years old. Diagnosis of PCMZL is made by skin biopsy, containing reticular dermis and fat, which can assist in distinguishing it from a reactive or inflammatory process. By definition, a primary cutaneous lymphoma affects the skin without evidence of disease elsewhere in the body when diagnosed. Marginal zone lymphoma (MZL) can include mucosal sites and extend to the lymph nodes and bone marrow, making MZL disseminated in approximately one-third of patients. Treatment of PCMZL includes surgery, radiotherapy, and topical therapy. The five-year overall survival rate for PCMZL is estimated to be 96.6% with a 10-year overall survival of 90.5%. A 70-year-old female presented for a facial rash present for 4 months. The patient reported the rash started as a small, pruritic, erythematous patch. The patch later evolved into an erythematous plaque covering bilateral cheeks and extending to the upper cutaneous lip. The results of the biopsy were consistent with primary cutaneous marginal zone lymphoma. The case is of interest because of the acute presentation in an older female with facial lesions only.

INTRODUCTION

Primary cutaneous marginal zone lymphoma (PCMZL) is a subtype of extranodal marginal zone B-cell lymphoma which involves mucosal sites.[1] PCMLZ presents more frequently in the fifth and sixth decades as red papules, plaques, or localized nodules commonly located on the trunk, arms, or head.[1] This is the case of a 70-year-old female who presented for care with a facial rash present for 4 months.

CASE REPORT

A 70-year-old female presented to dermatology for a facial rash present for 4 months. The patient reported the rash started as a small, pruritic, erythematous patch. The patch later evolved into an erythematous plaque covering half the surface area of the left cheek and a quarter of the area of the right cheek with two papules on either side above the superior vermillion border of the lip (**Figures 1 and 2**). In addition to the rash, the

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patient reported occasional chest pain and shortness of breath. Family history was positive for sarcoidosis. A 6 mm punch biopsy of the upper cutaneous lip was performed to initially rule out sarcoidosis, Sweet's syndrome, or tumid lupus. The results of the biopsy were consistent with primary cutaneous marginal zone lymphoma (low grade small cell B-cell lymphoma).

FISH analysis was positive for trisomy 12 and negative for trisomy 3, trisomy 18, BCL6 rearrangement, IGH rearrangement, or MALT1 rearrangement. Immunostain results of lymphocytic infiltrate showed most cells positive for CD23; remnant germinal cells positive for Ki-67; positive for CD20, CD79a, BCL2; few cells positive for CD138, Kappa ISH, and Lambda ISH; follicular dendritic cell meshworks disrupted for CD21; negative for CD3, CD5, CD10, CD43, BCL6 (few remnant germinal centers positive), and MUM1. Flow cytometry of the peripheral blood was negative for increased blasts, monotypic B-cells, or abnormal T-cell populations. PET scan revealed FDG avid bilateral facial lesions consistent with lymphoma or an inflammatory process without evidence of distal FDG avid disease (**Figure 3**).

Treatment was initiated with four rituximab infusions given weekly over one month. Patient noticed thinning of plaques and decreased erythema after first treatment. Treatment plan includes following up with patient after rituximab and potential radiation treatment pending results of rituximab infusions.

DISCUSSION

Primary cutaneous marginal zone lymphoma (PCMZL) is a subtype of extranodal marginal zone B-cell lymphoma which involves mucosal sites.[1] Approximately 8% of non-Hodgkin lymphomas can be classified as marginal zone lymphomas.[1] PCMLZ presents more frequently in the fifth and sixth decades as red papules, plaques, or localized nodules most commonly located on the trunk, arms, or head.[1] Male patients present twice as often as female patients, and the median age is 55 years old.[2]

Diagnosis of PCMLZ is made by skin biopsy, containing reticular dermis and fat, which can assist in distinguishing it from a reactive or inflammatory process.[3] B-cells in the marginal zone express CD markers CD20, CD22, CD79a, and BCL-2, but do not generally express CD5, CD10, or BCL-6.[1]

By definition, primary cutaneous lymphoma affects the skin without evidence of disease elsewhere in the body when diagnosed.[1] After diagnosis, initial staging involves thorough physical exam, blood cell counts, tissue sampling, and CT scans.[1] Marginal zone lymphoma can include mucosal sites and extend to the lymph nodes and bone marrow, and approximately one-third of patients have disseminated marginal zone lymphoma.[1] Treatment of PCMLZ includes surgery, radiotherapy, and topical therapy. Patients with solitary lesions are candidates for surgery or radiotherapy, and such measures are often curative.[3] For patients with multifocal disease, topical therapy is recommended. This spectrum of therapy includes class I glucocorticoids, topical mustard, topical imiquimod, intralesional triamcinolone, intralesional interferon, and cryotherapy.[2,4,5,6] The five-year overall survival rate for PCMZL is estimated to be 96.6% with a 10-year overall survival of 90.5%.[1]

Conflict of Interest Disclosures: None

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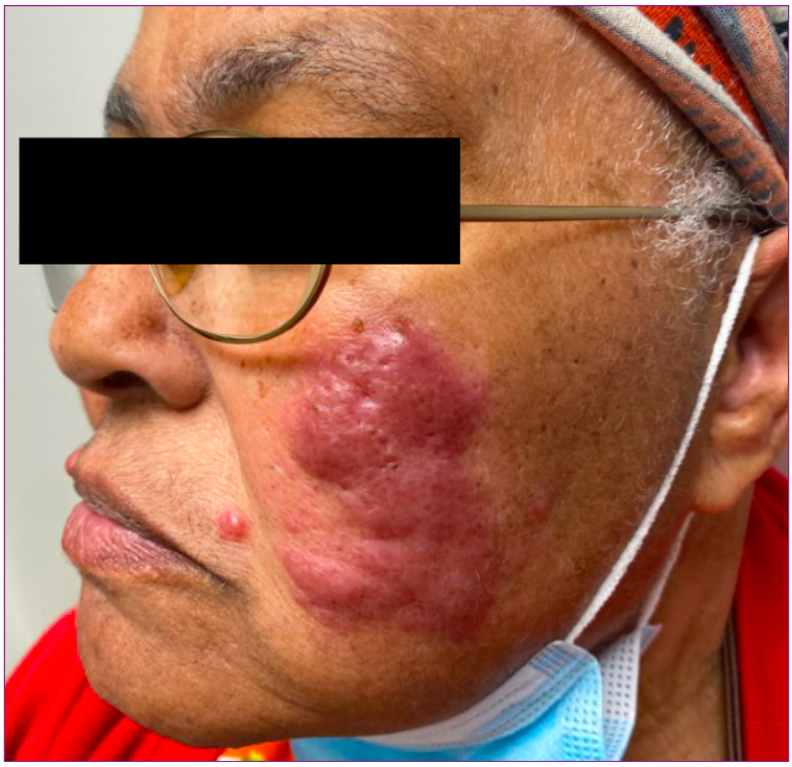

Figure 1. Erythematous plaque on the left cheek and one papule on above the left superior vermillion border of the lip.

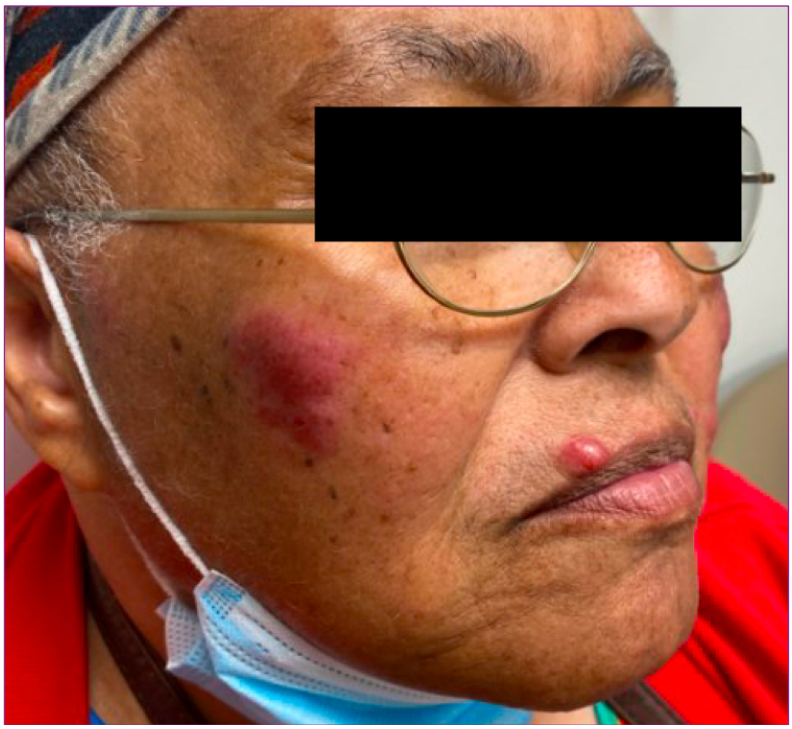

Figure 2. Erythematous plaque on the right cheek and one papule on above the right superior vermillion border of the lip.

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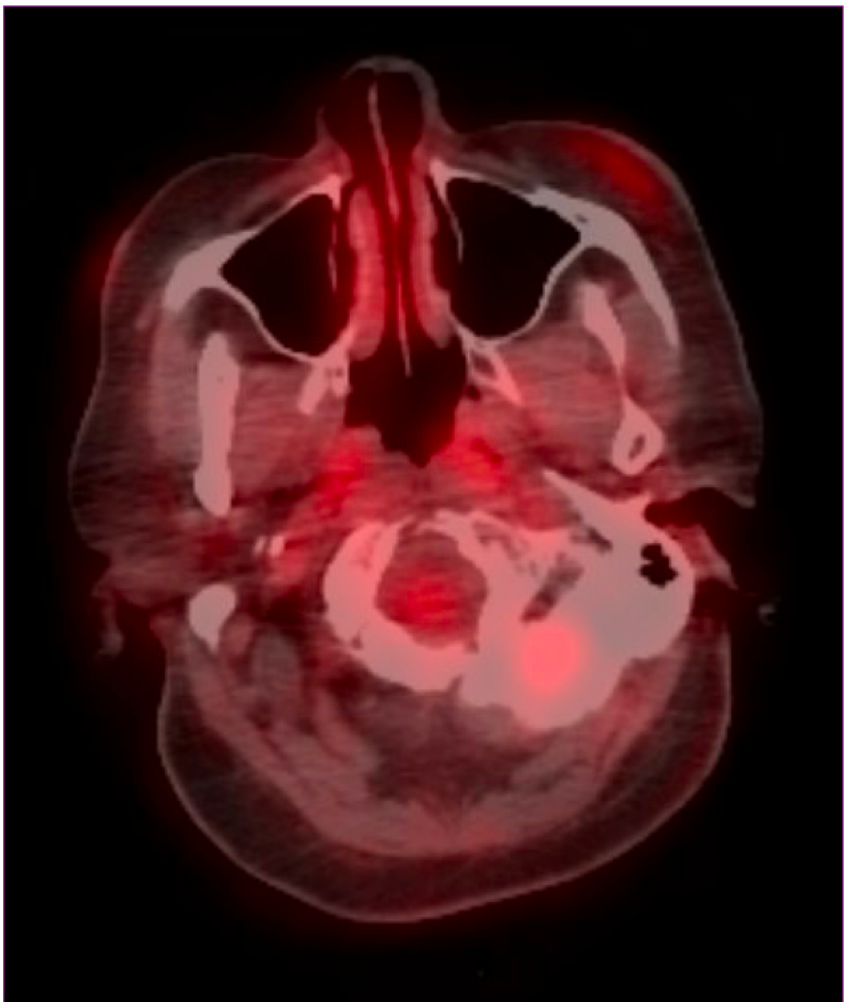

Figure 3. PET scan of head.

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