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Case presentation

A case report of primary cutaneous marginal zone lymphoma treated with intralesional steroids

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Abstract

Importance: Primary cutaneous marginal zone lymphoma (PCMZL) is a low-grade malignant B-cell lymphoma that appears in the skin without any extracutaneous manifestations. Therapeutic mainstays for PCMZL have primarily included radiotherapy and surgery. Intralesional steroids have been found to resolve the lesions caused by PCMZL, but there is a dearth of literature regarding this therapy indicating that this is not a commonly favored treatment option.

Observations: We present a case of 60-year-old woman who presented with PCMZL on her right eyebrow. Three years later, after two courses of radiation and one relapse, the patient presented with new lesions on both arms, consistent once again with PCMZL. At this time, therapy with nine rounds of intralesional triamcinolone was attempted. After each round of triamcinolone, gradual improvement of the lesions was noted until complete resolution occurred.

Conclusions and Relevance: This case reminds practitioners that intralesional corticosteroids can be effectively and safely used to treat localized PCMZL. Intralesional steroids are less invasive, cheaper, and easier to administer than the majority of other recommended therapies for PCMZL. Thus, although they are often overlooked, intralesional corticosteroids should be more often considered when developing a treatment plan for localized PCMZL.

Keywords: lymphoma, corticoteroids, cutaneous marginal zone lymphoma

Background

Primary cutaneous marginal zone B-cell lymphoma (PCMZL) is one of the major types of primary cutaneous B-cell lymphoma. It is a low-grade malignant B-cell lymphoma that appears in the skin with no extracutaneous manifestations and represents 2-16% of all cutaneous lymphomas [1]. PCMZL is thought to be the cutaneous counterpart of extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue, also known as MALT lymphoma [2]. There have been a variety of therapeutic options for PCMZL. Surgery and radiotherapy have been the most common treatments. Rituximab has also been used [3]. The following is a case report of the use of intralesional triamcinolone in PCMZLshowing resolution of the localized lymphoma.

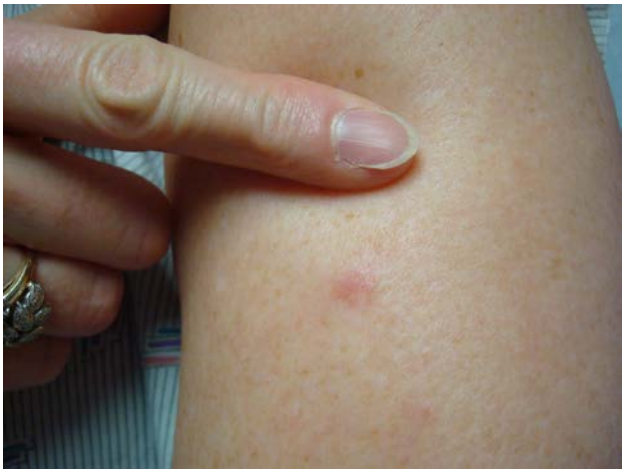
Case synopsis

A 60-year-old woman presented to the dermatology clinic with a papule on her right eyebrow. The patient denied any fevers, chills, sweats, or weight loss; there was no edema, cyanosis, or clubbing of the extremities. The patient had a past medical history of hypercholesterolemia, osteoporosis, seasonal allergies, and premature ventricular contractions. Her medications included metoprolol, atorvastatin, ezetemide, cholesevelam, fish oil, calcium, vitamin D, ceterizine, and alendronate. On physical exam, a light gray papule was observed on the medial edge of the right eyebrow. No cervical, supraclavicular, or axillary adenopathy was present. No systemic disease was noted.

Biopsy of the lesion demonstrated a sheet-like proliferation of monocytoid lymphocytes with focal germinal center formation composed of tingible body macrophages. Immunohistochemical stains found nodules of CD20 positive B cells, small CD3 positive T cells, and sheets of CD79a positive B cells. The CD21 stain demonstrated germinal centers with some intact follicular dendritic networks. Bcl-2 testing was not done because the diagnosis of PCMZL was strongly supported by the monoclonal population of plasma cells within the lesion. Lymphoid hyperplasia does not have this monoclonal population, and as a result, running the Bcl-2 was believed to be non-contributory. The plasma cells were kappa light chain restricted and the kappa:lambda ratio was determined to be greater than 10:1. The biopsy was consistent with marginal zone lymphoma. A positron emission tomography and computed tomography scans showed only a reactive lymph node in the left inguinal region. No hypermetabolic lymphadenopathy was noted in the neck, chest, abdomen, or bilateral lower extremities. A bone marrow biopsy was negative for lymphoma.

Because the biopsy was consistent with PCMZL, the patient received local radiation, which led to resolution. However, the lymphoma recurred lateral to the radiation field one year later. Repeat biopsy was similar to the first and she had a second course of radiation with resolution.

Three years later, the patient returned with lesions on both arms. On her left upper arm, she had a one-centimeter erythematous nodule (Figure 1). There was also a one-centimeter erythematous plaque just inferior to the first lesion and a 0.5-centimeter erythematous plaque on the right extensor forearm (Figure 2). No lesions were noted at the right eyebrow nor was there any lymphadenopathy. Biopsies of the lesions were similar to the previous biopsies of the eyebrow lymphomas, essentially consistent with cutaneous marginal zone lymphoma (Figure 3-5). A positron emission tomography, computed tomography, and bone marrow biopsy were once again unremarkable.



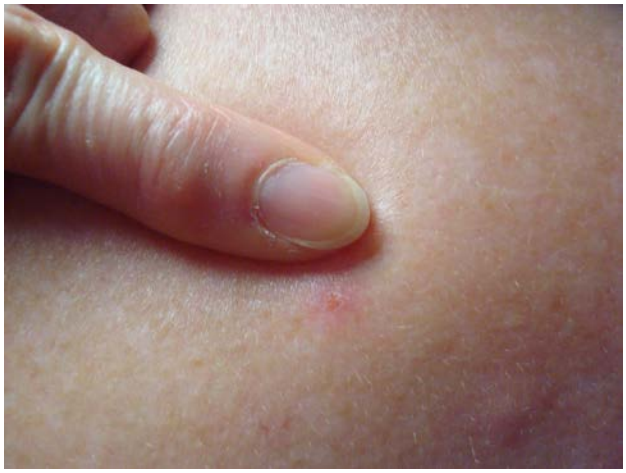
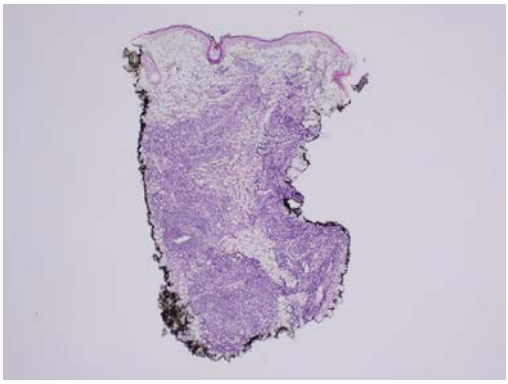
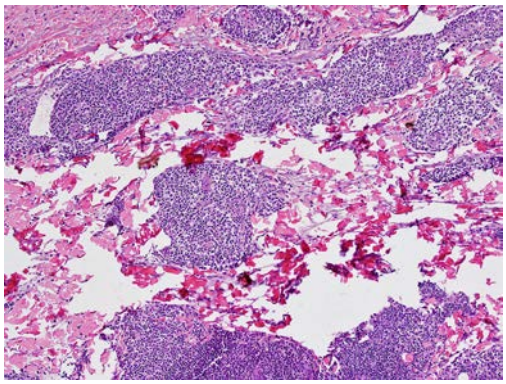


Figure 1. Left upper arm nodule prior to intralesional steroid treatment. **Figure 2.** Right extensor forearm nodule prior to intralesional steroid treatment





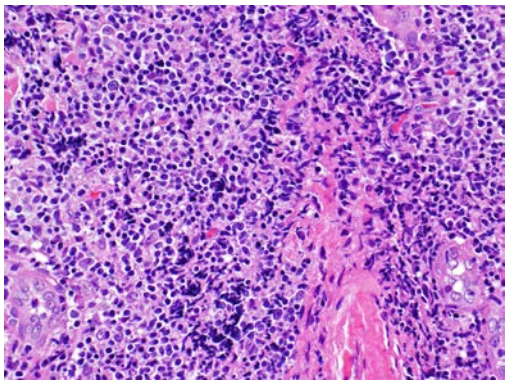
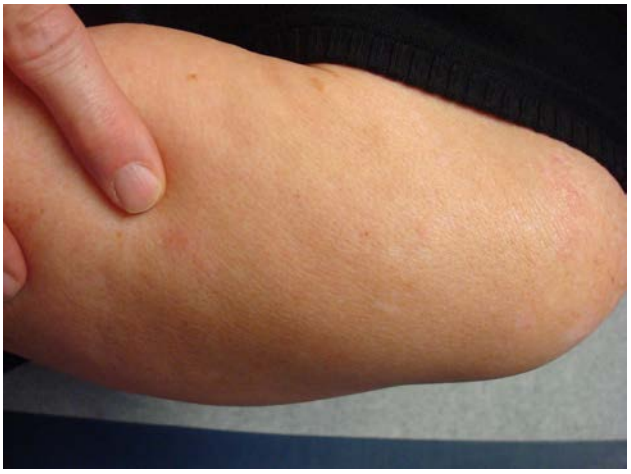


Figure 3: Biopsy of left upper arm erythematous nodule with features of cutaneous marginal zone lymphoma: sheet-like proliferation of monocytoid lymphocytes (scanning magnification image) **Figure 4:** Biopsy of left upper arm erythematous nodule with features of cutaneous marginal zone lymphoma: sheet-like proliferation of monocytoid lymphocytes with focal germinal center formation composed of tingible body macrophages (medium power image). **Figure 5:** Biopsy of left upper arm erythematous nodule with features of cutaneous marginal zone lymphoma: sheet-like proliferation of monocytoid lymphocytes with focal germinal center formation composed of tingible body macrophages (high power image).

Because the patient had had relapses treated with radiation and wanted noninvasive therapy, the decision to try intralesional corticosteroids at each of her seven lesions was made. She received five milliliters of triamcinolone to each. In total, she received nine courses of five milliliters of triamcinolone to each lesion over a course of 25 weeks. The patient demonstrated gradual improvement after each injection with eventual resolution (Figures 6-7).



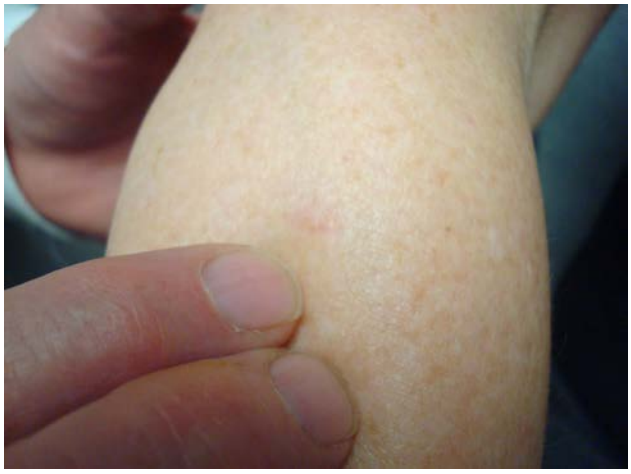


Figure 6. Left upper arm nodule after intralesional steroid treatment. **Figure 7.** Right extensor forearm nodule after intralesional steroid treatment.

Discussion

This case reminds practitioners that intralesional corticosteroids can be effectively and safely used to treat PCMZL. Despite safety and efficacy, there is a dearth of literature regarding intralesonal corticosteroids, indicating that this is not a commonly favored treatment option for PCMZL.

PCMZL has been found to primarily affect middle-aged individuals with a male predominance [3]. On clinical presentation, early cutaneous lesions can present as a nonspecific erythematous eruption, with subsequent progression to red-brown papules, plaques, and nodules localized particularly to the trunk, extremities, or head and neck [2]. The majority of patients present with multiple lesions [4]. Extracutaneous dissemination is rare [5]. An association with *Borrelia burgdorferi* has been reported in European countries but no such link has been found in the USA or Asia [6].

PCMZL is characterized by a polymorphous infiltrate that includes centrocyte-like, monocytoid, and lymphoplasmacytoid lymphocytes and plasma cells, all of which show monotypic cytoplasmic immunoglobulin light chain expression on paraffin sections [1,2]. The neoplastic cells in PCMZL express B-cell markers (CD20 and CD79a) and bcl-2, but are negative for CD5, CD10, and bcl-6 [2]. PCMZL tends to exhibit specific genetic aberrations, including the chromosomal translocation t(14;18)(q32,q21) involving the immunoglobulin heavy-chain locus and a gene for mucosa-associated lymphoid tissue 1 (MALT 1), as well as trisomy 18 [1].

Although PCMZL runs an indolent course, the prognosis is excellent. The 5-year survival rate is over 95%. Recurrences do occur in over half of the patients but do not confer a poor outcome [7]. Management of PCMZL is typically determined by the extent of disease. For localized disease with solitary or few skin lesions, first line treatment is complete surgical excision or radiotherapy [8]. Radiation doses of 35-40 Gy are commonly used [9]. For disseminated disease, multiple treatment options are available. A wait and see strategy has been described in some literature. Patients are carefully monitored and only symptomatic lesions are treated with either excision, topical or intralesional corticosteroids or low dose radiation [5]. Alternatively, another treatment option to treat with intravenous or intralesional therapy with rituximab (anti-CD20 chimeric monoclonal antibody) at diagnosis [7]. Finally, in many European centers chlorambucil is used in older patients with PCZML [5]. Chemotherapy (typically a cyclophosphamide, adriamycin, oncovin, and prednisone regimen) is only used when there is extracutaneous involvement or with aggressive disease depicting histological progression/transformation, B symptoms, or high LDH level [7,9].

Although intralesional corticosteroids are mentioned as second line therapy therapy for PCMZL, specific data regarding use is lacking [7]. Of the few cases in the literature, intralesional corticosteroids have been used as solo therapy for symptomatic lesions in disseminated disease or in conjunction with radiotherapy for localized disease [5,10]. One retrospective chart review found improvement in induration, palpability, and size of nodules in all PCMZL patients treated with solo intralesional steroid therapy. All patients had localized disease without any leg involvement. Between 0.5 to 12 milligrams was injected into each lesion. Complete resolution occurred in 4 out of the 9 patients. Overall, response was noted to be long-lasting with a median of 47 months [11]. Although some data exists for corticosteroids in localized disease, studies need to be done to determine use in disseminated and indolent PCMZL.

Intralesional corticosteroids are less invasive, cheaper, and easier to administer than the majority of other recommended therapies for PCMZL [11]. Furthermore, they have been shown to be efficacious in the few studies existing in the literature, as well as in our case. Intralesional corticosteroids also tend to have fewer adverse effects compared to other common therapies for PCMZL, including radiation, rituximab, and excision. Considering these advantages, intralesional corticosteroids injection is a therapy that can be used more often in treating PCMZL, especially in the setting of localized disease as in our case.

This case reminds practitioners that intralesional corticosteroids can be effectively and safely used to treat localized PCMZL. Although often overlooked, intralesional corticosteroids should be more often considered when developing a treatment plan for localized PCMZL.

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