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Letter: Primary cutaneous marginal zone B cell lymphoma of the face: A challenging diagnosis

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Abstract

The primary cutaneous marginal zone B cell lymphoma is a small B cell lymphoma, including cells of the marginal zone, lymphoplasmacytic cells, and plasma cells. Clinically it manifests as erythematous or erythematous-violaceous papules, plaques, or nodules, single or multiple, most often located to the extremities. Its course is usually indolent, with a survival at 5 years of approximately 97 percent. The tumor exhibits a tendency towards local recurrence, but spread to locations outside the skin is extremely rare. We present a case report of a man, 80 years of age, with a primary cutaneous marginal B cell lymphoma of the chin, an atypical location.

Primary cutaneous B cell lymphomas represent a proliferation of aberrant immunophenotype B cells localized to the skin, without evidence of extracutaneous disease at the time of diagnosis. They must be distinguished from systemic lymphomas, which may involve the skin secondarily and have a completely different clinical behavior and prognosis.

The World Health Organization/European Organization for Research and Treatment of Cancer (WHO/EORTC) classification divides cutaneous B cell lymphomas (CBCL) into two major groups: lymphomas with indolent clinical behavior and lymphomas with intermediate clinical behavior [**1**]. Primary cutaneous marginal zone B cell lymphomas (PCMBCL) are included in the first category.

An 80-year-old male was referred to our department in 2009 because of an asymptomatic growth on his chin. The nodule appeared one year before and had gradually enlarged over time. An ultrasound performed two months before suggested the diagnosis of a vascular tumor. The patient was otherwise asymptomatic and denied any constitutional symptoms. The medical history was positive for hypertension and benign prostatic hyperplasia, for which he was medicated with enalapril and finasteride, respectively.

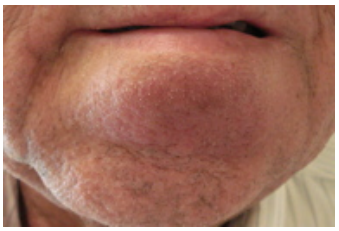


Figure 1

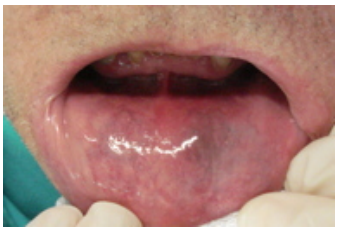
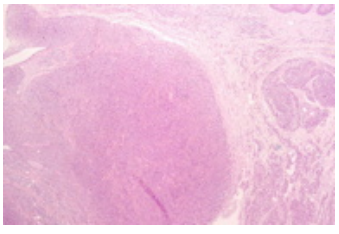


Figure 2

Figure 1. Tumor on patient's chin

Figure 2. Tumor on patient's oral mucosa

On physical examination, an erythematous-violaceous, slightly quadrangular, infiltrated and non-pulsatile nodule of 6 cm x 5 cm appeared localized to the chin (Figure 1) and corresponding oral mucosa (Figure 2) was noted. No lymphadenopathy, abdominal masses, or organomegaly were detected.



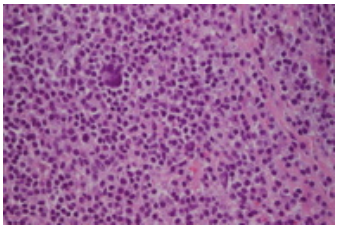
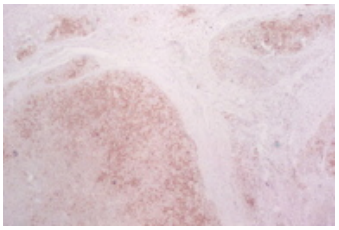


Figure 3	Figure 4

Figure 3. Unremarkable epidermis overlying a dermis with a nodular infiltration (H&E x4)

Figure 4. Lymphocytic infiltration (H&E x40)

A punch biopsy revealed an unremarkable epidermis overlying a dermis with a nodular infiltration of atypical lymphocytes, histiocytes, and eosinophils (Figures 3 and 4). The immunohistochemistry showed that the lymphocytic infiltrate was positive for CD20 (Figure 5) and BCL-2 (Figure 6) and negative for CD10, CD5, and BCL-6. It also demonstrated a kappa light chain restriction. A diagnosis of cutaneous marginal zone B cell lymphoma was made.



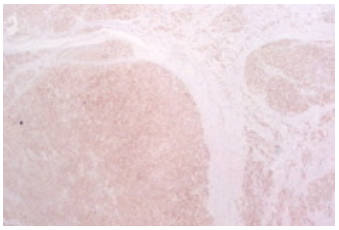


Figure 5	Figure 6

Figure 5. CD20 positive (x4)

Figure 6. BCL-2 positive (x4)

Borrelia burgdorferi serology and PCR on the tissue sample were negative. Laboratory tests were within normal ranges. CT scan of the neck, chest, abdomen, and pelvis showed no lymphadenopathy, hepatomegaly, or splenomegaly and the flow cytometry immunophenotyping of peripheral blood lymphocytes demonstrated no B cell monoclonality, establishing a definitive diagnosis of primary cutaneous marginal zone B cell lymphoma.

The patient was treated with 3 million units of subcutaneous interferon α2a three times a week during six months.

Twelve months after the beginning of the treatment, there is no evidence of local recurrence or extracutaneous disease.

PCMBCL accounts for approximately 8 percent of all non-Hodgkin lymphomas [**2**] and is one of the most frequent types of CBCL, representing, along with follicle center cell lymphoma, 75 percent of the cases [**3**].

It usually presents in the fifth and sixth decades as red to violaceous papules, plaques or nodules preferentially located to the trunk (46%) and arms (17%) [**2**]. In this case, the atypical location on the face and the presentation as a subcutaneous nodule probably led to an incorrect clinical suspicion of a vascular tumor. This hypothesis was also supported by the ultrasound, an exam that can be subjective. The correct diagnosis was made on the basis of the clinical history, physical examination, and histological and immuno-histochemical analysis.

In 2008, a consensus recommendation for treatment of CBCL was published by the EORTC and the International Society for Cutaneous Lymphomas [**4**]. According to these, patients presenting with solitary lesions can be treated conveniently by surgical excision or radiation therapy. These options may not be the best for young patients or in lesions, as in the present report, located on the face, where these treatments may cause disfiguration [**5**]. In these conditions, the alternative therapies are subcutaneous or intralesional interferon alpha or rituximab. Given the extent of the tumor, intralesional injection would have been a painful and lengthy procedure. Therefore, we performed subcutaneous instead of intralesional interferon α2a and this produced an excellent response.

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