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Oral involvement of cutaneous T-cell lymphoma

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Abstract

Mycosis fungoides (MF) is the most common cutaneous T cell lymphoma that involves the oral mucosal. The manifestation of lesions within the oral cavity generally correlates with a poor prognosis. Management of MF includes skin directed therapies and localized radiation treatment, with systemic biologic therapies and chemotherapy used for more advanced stages. The clinical and histologic features of MF in a patient with oral disease are reviewed.

Keywords: mycosis fungoides, T cell lymphoma, oral cavity

Introduction

Mycosis fungoides (MF) is the most common form of cutaneous-T cell lymphoma. Oral cavity involvement, however, is uncommon and reported only in a minority of patients. When oral involvement does occur, the most commonly involved sites are the tongue, palate, gingiva, buccal mucosa, lips, and oropharynx. Oral lesions typically present after cutaneous findings are identified [1]. MF may develop internally over time to include lymph nodes and other organs, but involvement in the oral cavity indicates systemic progression and a poor prognosis.

Case Synopsis

An 85-year-old man presented to the clinic with painful lesions on his tongue and a lump in his neck over the past 6 weeks. On examination, there were multiple pink and white papules on the tongue and a 6 cm pink tumor on the anterior neck (Figures 1, 2). A positron-emission tomography-computed

tomography scan demonstrated abnormal enhancement at both sites. Skin and tongue biopsies were then performed, which revealed cutaneous T-cell lymphoma (Figures 3 and 4). On systemic workup, there was evidence of right axillary lymph node involvement. The patient received a course of palliative radiotherapy, with complete resolution of his mucosal findings. Unfortunately, our patient died 6 months after initial presentation from progressive disease.

Case Discussion

Oral involvement of MF is an uncommonly reported manifestation. One study by Sirois et al. reported a lower than 1% incidence of oral lesions in 824 patients with MF over a 25-year period [2]. However, as an incidental finding observed in autopsy studies, 7% to 18% of patients with MF have oral involvement [3].



Figure 1. Neoplastic changes resulting in hypertrophy of lingual tissue.



Figure 2. Mycosis fungoides plaque of the neck.

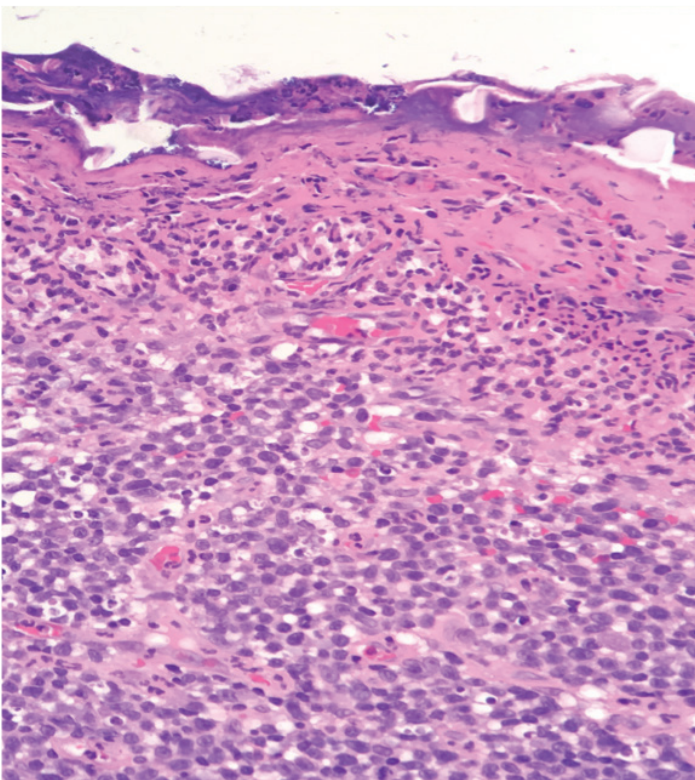


Figure 3. Ulceration at the base of the tongue is associated with an infiltrate of atypical lymphocytes with hyperchromatic nuclei, H&E, 200x.

The disparity between clinical and autopsy findings suggests that oral MF may be frequently missed on examination. Although the oral mucosa is frequently involved in hematologic disorders that affect the skin, in certain conditions such as MF, intraoral lesions may indicate progression to a more severe, systemic disease.

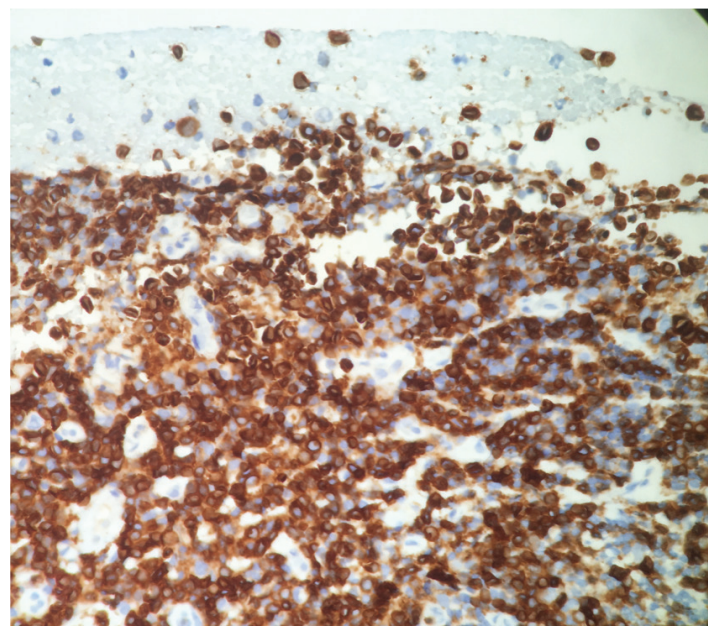


Figure 4. CD3 stain decorates the atypical lymphocytes within the epithelium.

MF can be divided into three progressive stages that includes erythematous patch, infiltrated plaque, and tumor. Overall staging of MF is determined using the TNMB system that assesses the size of lesions and percent skin surface involvement (T), lymph node involvement (N), metastasis (M), and quantitative amount of Sezary cells in the blood (B). Our patient presented in stage IIB with the presence of a tumor on the neck larger than 1 cm with lymphadenopathy.

Clinical presentation of oral MF varies, but most commonly presents as an ulcerated plaque or tumor on the tongue or palate [4]. Other presentations include indurated plaques, papules, leukoplakia-like lesions, or multiple erosions and may present with dysphagia. In a review of 42 reported cases of oral MF, cutaneous lesions typically preceded mucosal lesions in all but two cases [5].

The current management of MF is based on the stage of the disease and includes skin-directed therapies such as topical corticosteroids and topical retinoids, systemic biologic therapy, or chemotherapy for more advanced and aggressive stages. Treating oral lesions poses a greater challenge than treating cutaneous lesions and usually warrants a more systemic approach. Early identification of oral involvement, however, may increase the efficacy of treatment. MF typically responds well to intraoral radiotherapy, with a recent report citing 14/19 patients having a

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complete initial response [4]. Another paper cited systemic chemotherapy, six monthly cycles of CHOEP (cyclophosphamide, hydroxydanorubicin, vincristine, prednisone, and etoposide), combined with alemtuzumab to successfully treat MF with oral involvement [6]. A more recent case reported resolution of disease using 22 Gy electron beam radiation followed by maintenance bexarotene with the patient remaining disease free for seven years since onset of oral lesions [5].

Conclusion

Although oral lesions may precede or follow the appearance of findings at other locations, a careful medical history and physical examination is critical to ensure proper and timely diagnosis and therapy. Even in patients with known cutaneous MF, oral disease can be a diagnostic challenge because the symptoms may be nonspecific and can closely mimic *Candida* infection. Other differential diagnoses include lymphomatoid papulosis, geographic tongue, infection, and other malignancies (such as squamous cell carcinoma). Given the poor prognosis, clinicians should maintain a high level of suspicion to perform a biopsy.

Given that many MF patients with oral involvement go undiagnosed until autopsy, MF patients should have a thorough oral examination and medical history tailored towards oral disease to ensure prompt diagnosis and treatment.

References

1. Goldsmith SM, Seo BL, Kumara de Silva R, Parachuru P, Rich AM, Seymour GJ. Oral mycosis fungoides: report with immune profile. *Oral Surg*. 2014;118(2):e48-e52. [PMID:24405649].
2. Sirois DA, Miller AS, Harwick RD, Vonderheid EC. Oral manifestation of cutaneous T-cell lymphoma: a report of eight cases. *Oral Surg Oral Med Oral Pathol* 1993;75:700-705. [PMID:8515982].
3. Long JC, Mihm MC. Mycosis fungoides with extracutaneous dissemination: a distinct clinicopathologic entity. *Cancer* 1974;34:1745-1755. [PMID:4426033].
4. Wain EM, Setterfield J, Judge MR, Harper JI, Pemberton MN, Russell-Jones R. Mycosis fungoides involving the oral mucosa in a child. *Clin Exp Dermatol*. 2003;28:499-501. [PMID:12950337].
5. Bassuner J, Miranda RN, Emge DA, DiCicco BA, Lewis DJ, Duvic M. Mycosis fungoides of the oral cavity: fungating tumor successfully treated with electron beam radiation and maintenance bexarotene. *Case Rep Dermatol Med*. Epub 2016 Dec 15. [PMID:28074163].
6. Postorino M, Pupo L, Provenzano I, Del Principe MI, Buccisano F, Franceschini L, Rizzo M, Nasso D, Meconi F, Onnis I, Vaccarini S, Zizzari A, Anemona L, Mauramati S, Bruno E, Cantonetti M, Amadori S. A case of oral mycosis fungoides successfully treated by combination of alemtuzumab and chemotherapy. *Ann Hematol*.