

A case of linear morphea involving the oral cavity



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INTRODUCTION

Morphea en coup de sabre is a form of linear scleroderma that primarily affects the head and neck and is named from its linear shape and characteristic lesions that resemble the wound from a saber. It is an inflammatory disease that results in excessive collagen deposition, sclerosis of involved skin and tissues, and progressive atrophy of involved areas. It may present as a hypopigmented or hyperpigmented linear, indurated plaque that progresses to assume an atrophic, depressed, ivory appearance.¹⁻⁴ The disease can lead to alopecia of the scalp and eyelids and may progress into deeper tissues and cause muscle atrophy, damage to nerves, and demineralization of bone and teeth.¹⁻⁶ In addition, neurologic involvement in patients with localized scleroderma has been increasingly reported, with seizures and headaches being the most common manifestations.⁷ Histopathologic findings typically show an atrophic epidermis with effacement of the epidermal rete ridges as well as homogenization and thickening of the dermal collagen.^{2,8} In the earlier or active stages of morphea, a primarily lymphoplasmacytic inflammatory cell infiltrate is more pronounced as well as tissue edema and enlarged, tortuous vessels.^{2,8} Later stages show loss of adnexal structures, decrease in blood vessel size and number, and disappearance of the inflammatory infiltrate.^{2,8} Typically, this form of localized scleroderma has no systemic involvement and most commonly affects the head and neck.

Cases of oral involvement are rare, with less than 20 reports noted in the literature. Although the etiology of this disease remains unknown, it is thought to be secondary to abnormalities in the

immune system. An example would be as a response to environmental triggers such as trauma in the context of genetic predisposition.^{5,9} Therefore, treatment relies on immunosuppressive measures such as methotrexate and corticosteroids.

CASE REPORT

We report on a 33-year-old African-American woman who presented to our clinic with a 1-year history of white discoloration of her superior gingiva and lip. At the time of evaluation, she described a 1-year history of a linear white patch that started along the gingiva and progressed to involve the mucosal and cutaneous lip as well as the columella and tip of the nose. Associated with this were loosening of the central and lateral incisors and mild soft tissue edema. When she first noticed the white discoloration, she was receiving a routine dental examination. The dentist presumed it to be fungal and proceeded to treat with nystatin solution. As the discoloration continued to progress, a biopsy was performed at an outside facility, which was reportedly read as premalignant. She was treated with a laser, yet the white patch continued to progress. She was referred to the oral and maxillofacial surgery department where she had several biopsies and teeth extractions before evaluation by the dermatology department. Initial biopsy of the anterior gingiva showed acute and chronic inflammatory changes without evidence of dysplasia. Biopsy of the anterior maxillary bone was unremarkable. Upper lip biopsy found squamous epithelium with spongiosis and acute chronic inflammation without dysplasia. She was then referred to the infectious disease

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Fig 1. **A**, A 0.5-cm (1.0 cm at Cupid's bow) $\times$ 5.5-cm (from mucosal lip to tip of nose with mouth closed) linear hypopigmented patch. **B**, Linear hypopigmented patch involving the upper gingiva, upper mucosal, and cutaneous lip extending onto columella and the tip of the nose.

department, where a culture was performed and grew *Streptococcus gordonii*. She was treated with oral clindamycin. However, the white discoloration continued to progress and involve the nasal tip. Imaging was performed to rule out osteomyelitis, and the results showed mild soft tissue edema. A subsequent culture grew *Streptococcus anginosus* and she was treated with oral metronidazole and levofloxacin without improvement. She was referred to us for further evaluation. On physical examination, she had a 0.5-cm (1.0 cm at Cupid's bow) $\times$ 5.5-cm (from mucosal lip to tip of nose with mouth closed) linear hypopigmented patch involving the upper gingiva, upper mucosal/cutaneous lip, and extending onto columella and tip of the nose (Fig 1, A and B).

The unusual location of these findings, posed a diagnostic challenge. Although morphea was clinically suspected, a repeat biopsy of the upper mucosal lip was performed. Histopathologic findings showed an atrophic epidermis with effacement of the epidermal rete ridges. The papillary and early reticular dermis consisted of homogenization of the dermal collagen with a sparse perivascular and somewhat lichenoid lymphoplasmacytic infiltrate with a few isolated eosinophils (Fig 2, A and B).

The diagnosis of linear morphea was confirmed, and the patient was started on methotrexate and high-dose prednisone with rapid improvement.

DISCUSSION

On review of additional cases reported in the literature, linear morphea of the oral cavity typically involves the mucosal lip and upper anterior teeth, such as in our case.^{1,2,5} Depending on the stage of

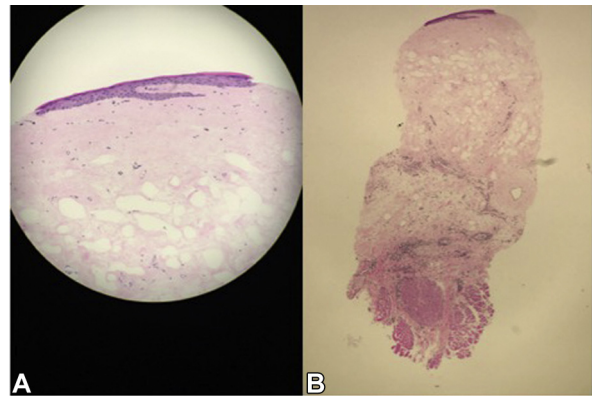


Fig 2. Histologic Images show a punch biopsy from the upper lip. **A**, Remarkable findings are an atrophic epidermis, effacement of the epidermal rete, and homogenization of dermal collagen. **B**, Sparse perivascular and somewhat lichenoid lymphoplasmacytic infiltrate. (Original magnifications: A, $\times 10$; B, $\times 40$.)

progression at time of diagnosis, each patient had varying degrees of gingival recession, alveolar bone loss, atrophy of the tongue, and loosening of the teeth.^{1,2,5} Most of these patients have been treated successfully with an initial combination of oral or intramuscular methotrexate at doses of 7.5 mg/wk to 25 mg/wk as well as systemic corticosteroids at doses of 15 mg/d to 60 mg/d for at least 1 year before a subsequent taper.²⁻⁵ Other potential therapeutic options have been discussed including topical tacrolimus, topical calcipotriene, Chinese herbal medicine *Salvia miltiorrhiza*, mycophenolate mofetil, and photochemotherapy.¹⁰ It is important for the clinician to keep morphea in the differential diagnosis when evaluating the oral cavity, as untreated linear morphea may lead to atrophy of the tongue papillae, demineralization and loosening of teeth, and recession of oral gingiva. Prompt treatment of this inflammatory disease with immunosuppressive medications allows for prevention of these possible complications and need for further trauma to the affected area with maxillofacial reconstructive efforts.

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