Case Report

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Oral Mucosal Morphea: A New Variant

M.M. Tang[a] M.M. Bornstein[b] N. Irla[a] H. Beltraminelli[a] T. Lombardi[c] L. Borradori[a]

[a]Department of Dermatology, Inselspital – Bern University Hospital, [b]Department of Oral Surgery and Stomatology, Bern University, Bern, and [c]Division of Stomatology and Oral Surgery, Geneva University, Geneva, Switzerland

Key Words
Morphea · Localized scleroderma · Gingival recession

Abstract
Morphea is a cutaneous disorder characterized by an excessive collagen deposition. While in almost all cases the sclerosing process exclusively affects the skin, there are anecdotal cases in which associated mucosal involvement has been described. We here report the case of a woman developing a whitish indurated plaque over the left upper vestibular mucosa and hard palate leading to dental mobility and exposure of the roots of several teeth. Cone beam computed tomography of the left maxilla showed bone resorption involving the upper cuspid to the second molar region with widened periodontal ligament spaces, while light microscopy studies demonstrated epithelial atrophy and fibrosis of the dermis extending into the submucosa with hyalinization of subepithelial collagen. Our observation expands the spectrum of clinical presentations of morphea and provides the first example of isolated oral morphea. Its recognition is important to avoid significant local complications.

Introduction

Morphea, also known as localized scleroderma, is a clinically distinct inflammatory disease, primarily affecting the dermis and subcutaneous fat. It is characterized by excessive collagen deposition leading to thickening of the dermis and/or subcutaneous tissues, ultimately leading to a scar-like sclerosis [1, 2]. The etiology of morphea remains elusive. Multiple mechanisms have been implicated, such as autoimmunity, infection, drugs, radiation and microchimerism [3, 4]. Clinically, various types of morphea are distinguished, such as plaque morphea, bullous morphea, deep morphea and linear morphea. The latter also encompasses morphea en coup de sabre and Parry-Romberg syndrome affecting the scalp and face with potentially devastating complications [3, 5–11]. Whilst in almost all cases the sclerosing process exclusively affects the skin, there are anecdotal cases in which associated mucosal involvement has been described [12–19].

We here describe a unique variant of morphea affecting only the oral mucosa with significant local tissue damage and no evidence of cutaneous sclerosis.

Case Report

A 20-year-old female presented with a 12-month history of progressive recession of the gingiva around the premolars and molars in the left maxilla. This was associated with a slowly enlarging painless whitish plaque over the left upper vestibular mucosa and hard palate. There was no history of preceding trauma or spontaneous gum bleeding, and her affected teeth were not mobile. Her personal and family history was otherwise unremarkable. She had no history of photosensitivity, Raynaud's phenomenon, arthralgias, dry eyes, fever or exposure to irradiation. Her only regular medication was an oral contraceptive (drospirenone and ethinyl estradiol, Yasmin®). She was a smoker (2 pack-years), but had quit smoking prior to the lesions' development.

Examination of the oral mucosa revealed a pronounced recession of the gingiva involving the upper left premolars extending to the first molar with a white, mildly indurated, nontender plaque located on the keratinized mucosa of the gingival region of the cuspid to the second molar, and further extending to the left maxillary vestibular mucosa and left upper buccal mucosa (fig. 1a–c). External resorption was prominent over the palatal root of

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Min Moon Tang
Department of Dermatology
Inselspital – Bern University Hospital
CH–3010 Bern (Switzerland)
Tel. +41 31 632 2288, E-Mail minmoon2005@yahoo.com

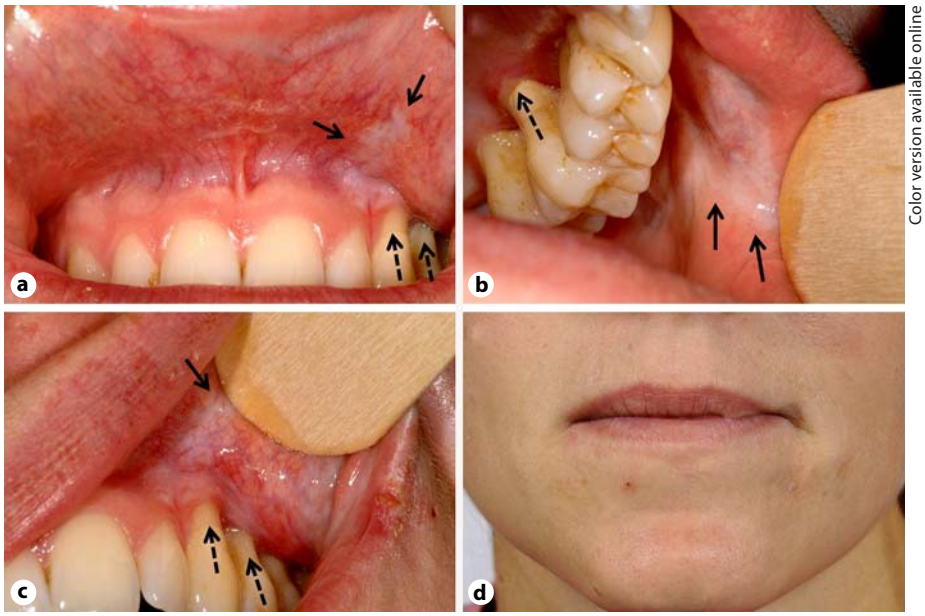

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Fig. 1. a–**c** Clinical feature of gingival recession affecting the upper left cuspid to the first molar (black dashed arrows) with a well-demarcated white plaque at the maxillary vestibule (black solid arrows). **d** Photo of the patient to demonstrate that there is no sclerotic skin lesion on the face apart from acne scars.

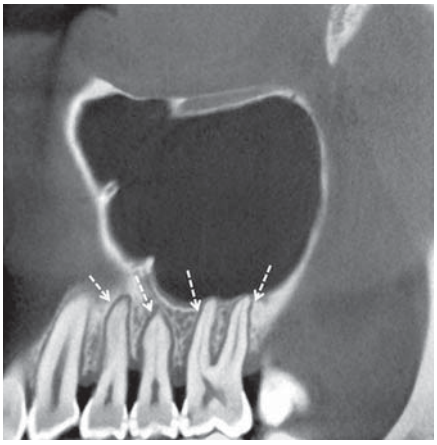

Fig. 2. Sagittal view of the cone beam computed tomography scan of the left maxilla shows widened periodontal ligament spaces and bony resorption around the first and second premolars and the first molar (white dashed arrows).

the left second molar. The oral hygiene was good. There were no other lesions of the rest of the oral mucosa. Examination of the rest of the skin was otherwise unremarkable (fig. 1d). The sagittal cut in the cone beam computed tomography of the left maxilla showed bone resorption involving the upper cuspid to the second molar region with widened periodontal ligament spaces (fig. 2). Light microscopy studies of a biopsy specimen obtained from the left oral vestibule demonstrated a mild hyperkeratotic parakeratinized squamous epithelium with epithelial atrophy, fibrosis of the dermis that extended into the submucosa and hyalinization of subepithelial collagen (fig. 3a). There was a lymphocytic inflammatory infiltrate around the vessels (fig. 3b). Special staining showed the presence of elastic fibers in the dermis. Direct immunofluorescence microscopy study of a mucosal biopsy sample was negative.

The full blood count, thyroid function test, renal and liver tests were normal. Search for autoantibodies, such as anti-dsDNA, antinuclear, anti-SS-A, anti-SS-B, anti-RNP, anticentromere, anti-Scl-70 (topoisomerase I) and anti-Jo-1 antibodies, was negative. Serologies for syphilis, *Borrelia burgdorferi* infection, hepatitis B and C viruses and HIV were negative.

A course of doxycycline 100 mg daily was initiated together with regular professional dental hygiene procedures for more than 1 year with little effect. A regular follow-up of the patient was difficult, making management even more difficult. She developed significant mobility of the affected 4 teeth later, making their cleaning difficult. The white plaque extended further involving the left soft palate and the posterior aspect of the left hard palate (fig. 4). The roots of teeth 23–26 were almost completely exposed. The patient is now being given intralesional triamcinolone acetate injections.

Discussion

We describe a unique case, the clinical, histopathological and radiological features of which were consistent with the diagnosis of isolated morphea of the oral cavity. Morphea has a wide range of clinical presentations and courses [3]. Although in the vast majority of cases there is no internal organ involvement, the fibrotic process may extend into and affect deep structures such as the hypodermis, muscles, joints and bones, causing severe disfigurement and morbidity [3]. Furthermore, morphea of the head and face (especially the linear variant) may also be associated with ocular as well as central nervous system anomalies [5–12].

Oral mucosa and dental involvement have only been rarely described in morphea. In virtually all previously described cases, as summarized in table 1 [13–27], oral involvement was secondary to a linear form of morphea, either in the en coup de sabre variant or in the Parry-Romberg syndrome. Besides the typical cutaneous features, the patients presented with scar-like or white indurated oral plaques, unerupted teeth, root development defects, root atrophy, localized gingival recession and alveolar bone resorption. Barber [28] described a middle-aged patient with a white patch on the mucous membrane of the left lower lip extending to the alveolar sulcus which was diagnosed as circum-

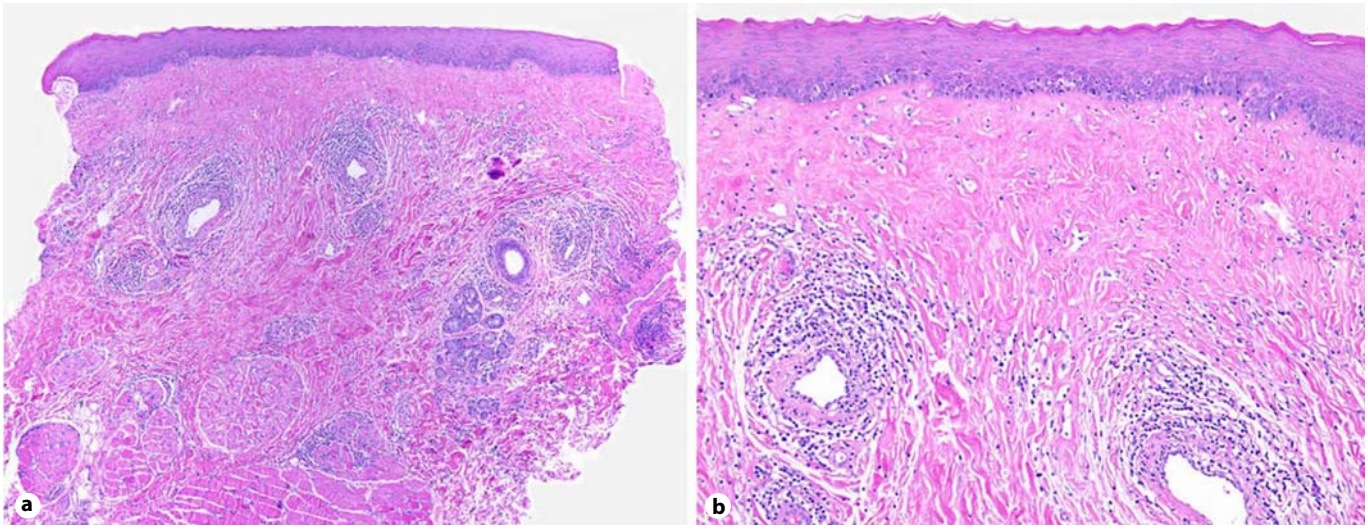

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Fig. 3. a Light microscopy of a biopsy specimen from the left oral vestibule showing mild hyperkeratotic parakeratinized squamous epithelium with epithelial atrophy, fibrosis of the dermis and hyalinization of subepithelial collagen. ×40. **b** There was presence of perivascular lymphocytic infiltrates. Hematoxylin-eosin stain. ×100.

scribed scleroderma of the buccal mucous membrane. However, the diagnosis was not histologically confirmed.

Our patient is therefore peculiar and striking for many reasons. First, the morphea was limited to the buccal, vestibular, palatal and gingival mucosa without any evidence of sclerosis of the adjacent skin. Light microscopy studies confirmed the presence of subepithelial fibrosis that extended into the submucosa with hyalinization of subepithelial collagen. Second, the involvement was severe leading to significant gingival recession, bone resorption and external tooth resorption. Third, no potential triggering factor, such as trauma, radiation or *B. burgdorferi* infection [29] was identified. Multiple drugs have been implicated in the development of morphea-like lesions [30]. Our patient took only oral contraceptive pills on a regular basis, which has not been previously linked to morphea. In the differential diagnosis we also considered lichen sclerosus et atrophicus (LSA) that may also lead to well-demarcated whitish plaques of the oral mucosa with dental problems. Oral LSA is also a very rare condition that may lead to gingival recession, widening of the periodontal space, loss of periodontal attachment and subsequently tooth loss [31–33]. Although in the oral mucosa it is clinically difficult to differentiate LSA from morphea, the histopathological findings in LSA are distinctive enough to allow its differentiation from morphea. There are epithelial hyperplasia with hyperkeratosis, focal hydropic degeneration of basal cells, subepithelial hyalinization and a band-like lymphocytic infiltrate beneath the hyalinized area [34]. The subepithelial elastic fibers are decreased in LSA [34]. In our case, there was no band-like infiltrate, while a prominent perivascular lymphocytic infil-

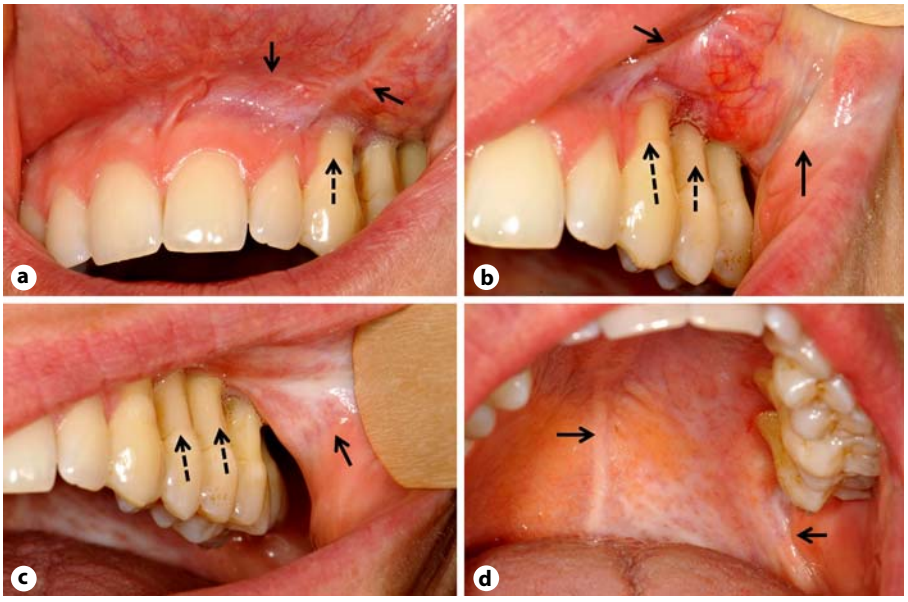

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Fig. 4. Clinical features at the 2-year follow-up visit. There is almost complete root exposure of teeth (black dashed arrows) and a progressively enlarging whitish plaque at the mucosa of the soft and hard palate (black solid arrows).

Table 1. Survey of the reported cases of morphea associated with oral and dental manifestations

Authors, year	Case	Duration of lesions	Cutaneous lesions	Oral or dental manifestations	Biopsy of oral lesions	Treatment
Looby and Burket [13], 1942	8-year-old	3 years	Linear morphea, left face	Scar-like appearance of oral mucosa of the maxillary left first incisor, depression in the alveolar process, unerupted maxillary left first incisor	NA	NA
Foster and Fairburn [14], 1968	6-year-old male	NA	Morphea of the face	Short roots of mandibular central and lateral incisors with short roots	NA	NA
Hoggins and Hamilton [15], 1969	4-year-old female	NA	Localized morphea	Lack of eruption of primary teeth	NA	NA
Foster [16], 1979	12-year-old male	7 years	Parry-Romberg syndrome, right face	Grooved right side of the tongue; shorter roots of the right lower teeth	NA	NA
Billet et al. [17], 1983	11-year-old	not mentioned	Morphea en coup de sabre, left nasolabial region	Gingival retraction of 2 left upper incisors with ulcer at the affected gingiva	Histological image of morphea	Excision of lesion with reconstruction; extraction of 2 incisors, prosthesis
Barton and Handerson [18], 1993	10-year-old female	5 years	Morphea en coup de sabre, left face	Root development defects of the left maxillary central and lateral incisors	NA	NA
Mazzeo et al. [19], 1995	10-year-old female	9 years	Parry-Romberg syndrome, left face	Roots of the left mandibular first and second molars and the maxillary left canine were atrophic; involved cuspid and second molar unerupted	NA	NA
Baxter et al. [20], 2001	12-year-old female	1 year	Morphea en coup de sabre, right face	Right upper labial gingival recession, mobility of right maxillary incisors; X-ray: bone loss over the right maxillary incisors	NA	Extraction of affected mobile teeth, replaced with upper removable prosthesis
Marzano et al. [21], 2003	7 cases	NA	Morphea en coup de sabre or Parry-Romberg syndrome	Dental abnormalities, malocclusion and tongue changes	NA	NA
Kurian et al. [22], 2003	15-year-old male 14-year-old female	2 years 8 years	Parry-Romberg syndrome of left face in both cases	Atrophy of filiform papillae of left side of the tongue, palatal constriction, disturbed eruption of second left bicuspid and second molar	NA	NA
Pekiner et al. [23], 2006	4-year-old female	2 years	Morphea en coup de sabre, left face	Deviation of maxillary midline to the left side, atrophy of the left side of the tongue	NA	NA
Virdi and Kanwar [24], 2009	39-year-old male	5 years	Generalized morphea	Fibrosis of soft palate and tongue	Inflammatory infiltrate in the subepithelium with submucosal fibrosis of the tongue	Pulse dexamethasone therapy

Table 1 (continued)

Authors, year	Case	Duration of lesions	Cutaneous lesions	Oral or dental manifestations	Biopsy of oral lesions	Treatment
Pace et al. [25], 2010	36-year-old female	NA	Morphea en coup de sabre, right face	Scar tissue extending to the attached gingivae, mobile upper right central incisor and gingival recession on the mesial aspect of 12	Atrophic stratified squamous epithelium with chronically inflamed hyalinized collagen	Extraction of upper central incisor and prosthesis
Liu et al. [26], 2010	59-year-old female	4 months	Morphea, right chin	White patch over the right lower labial mucosa, gingiva attached to the right mandibular canine and lateral incisor; radiologically, alveolar bone resorption	Mucosal biopsy consistent with morphea	Traditional Chinese medicines, asiaticoside ointment
	12-year-old female	12 months	Morphea, right lower lip	White patch over the right lower labial mucosa		
Herrick et al. [27], 2011	3 cases	NA	Linear morphea, Parry-Romberg syndrome, mixed scleroderma	Involvement of mouth, teeth and gums	NA	NA

NA = Not available or not specified.

trate was observed. In addition, there was no decrease in dermal elastic fibers. Thus, the diagnosis of morphea was retained.

Evidence-based therapy for morphea is almost entirely lacking because of the rarity of the disease and the lack of accepted validated outcome measures [35, 36]. Management of the mucosal lesion in our patient was challenging, not only because of the patient's poor compliance, but also because of the location of the lesion, which is a limiting factor for all topical treatments such as corticosteroids and tacrolimus. Oral phototherapy, including PUVA, narrow-band UVB and excimer laser have all been tried with variable success in oral lichen planus and oral manifestations of chronic graft-versus-host disease [37–40]. Doxycycline was given to this patient based on its anti-inflammatory and immune-modulating properties which were found useful in anecdotal cases of scleroderma and bullous dermatoses [41], as well as for its effectiveness in chronic periodontitis [41, 42]. Since there was a lack of response to doxycycline on top of the poor compliance, the patient was started on intralesional corticosteroids. In case of further progression of the disease, either oral methotrexate [35, 36, 43, 44] or cyclosporine [45, 46] will be tried.

In conclusion, we present a unique case of morphea limited to the oral mucosa in a young woman. Clinicians should be familiar with the wide spectrum of presentations of morphea, and now further consider this diagnosis in the presence of isolated whitish sclerotic plaques of the oral mucosa. Its prompt recognition and close follow-up are important to avoid significant local complications.

Disclosure Statement

No conflicts of interest.

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