Severely Crusted Cheilitis as an Initial Presentation of Systemic Lupus Erythematosus

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Abstract

Lupus erythematosus (LE) is an autoimmune disease which may initially present solely with lip lesions. Due to a wide spectrum of presentation, these features may initially be misdiagnosed as other oral diseases such as lichen planus, erythema multiforme (EM), and actinic cheilitis, leading to a delay in diagnosis and treatment. We discuss a case of severely crusted cheilitis which was initially diagnosed as EM, with subsequent development of subacute cutaneous LE, and progression to systemic LE. We will discuss the clinical and histological features of lupus cheilitis.

Key Words: Cheilitis, crusted, systemic lupus erythematosus

Introduction

A 54-year-old Chinese woman with a past medical history of hypertension on lisinopril for more than 5 years presented to a tertiary dermatology center with a 2-month history of abrupt onset, painless severe crusting of the lower lip [Figure 1a]. Apart from her antihypertensive, she was not on any other medications or supplements. Examination revealed extensive hyperkeratotic crust over the lower lip, sparing the upper lip. The rest of the oral cavity and skin was normal. She had no photosensitivity, oral ulcers, joint pains, dry eyes, and absence of family history of autoimmune disease. Based on clinical examination, an initial diagnosis of erythema multiforme (EM) was entertained with differentials including actinic cheilitis, lichen planus (LP), lupus cheilitis, and paraneoplastic pemphigus. She was referred to a tertiary dental center for a biopsy for which she declined and was subsequently given prednisolone 30 mg daily for 2 weeks for the treatment of presumed EM and topical clobetasol ointment.

On review 2 weeks later, the patient’s severe crusted cheilitis improved [Figures 1b and 2]. We encouraged a biopsy, and further laboratory investigations revealed a normal full blood count and high antinuclear antibodies (ANAs) of >1/800, nucleolar and speckled pattern. However, the patient subsequently defaulted further follow-up.

Six months later, the patient presented with widespread photodistributed erythematous scaly patches and plaques over the face, chest, and extensors of the forearms [Figure 3a and b]. A skin biopsy from the left forearm revealed interface vacuolar dermatitis, lymphocytic inflammation, lower epidermal apoptosis, hyperkeratosis, and hypergranulosis. There was no follicular plugging or vasculitis. Periodic acid–Schiff (PAS) stain demonstrated basement membrane thickening [Figure 4a]. Direct immunofluorescence (DIF) showed IgM and C3 immunodeposits along the basement membrane zone [Figure 4b].

Based on clinicopathological correlation, a diagnosis of subacute cutaneous lupus erythematosus (SCLE) was made, and the patient was started on hydroxychloroquine 300 mg daily, and subsequently increased to 400 mg daily, topical corticosteroids, and photoprotection. She was reviewed by a rheumatologist, and investigations did not fulfill the American College of Rheumatology (ACR) criteria for systemic lupus erythematosus.

What was known?

• Oral manifestation of lupus can have a wide spectrum of clinical presentation
• This patient presented with lower lip involvement suggesting that this was a photoaggravated or induced condition

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Two months later, her rashes progressed despite treatment. Further investigations revealed urine proteinuria (urine PCR, 0.28 mg/mmol), autoimmune hemolytic anemia (elevated lactate dehydrogenase, 529 U/L; low haptoglobin, 0.35 G/L; positive direct anti-human globulin test 1+), and positive autoantibodies to extractable nuclear antigens, including ribonucleoprotein, and Ro (SSA) antibody were detected. Rheumatological evaluation fulfilled the ACR criteria for SLE. Prednisolone 40 mg daily was added with control of disease activity.

**Discussion**

The frequency of which SLE presents with oral lesions ranges from 9% to 45%.\(^1\) Mucocutaneous manifestations of lupus erythematosus (LE) can present in a variety of ways, which can cause difficulties in early diagnosis as demonstrated in our case.\(^2\)

Lupus cheilitis has various clinical presentations ranging from atrophic plaques to white/keratotic, purpuric, bullous, and verrucous lesions. Concomitant oral and cutaneous discoid lesions with high titers of circulating ANA may be at particular risk of developing systemic disease and warrant careful monitoring.\(^1\) The most common areas for lesions are buccal mucosa followed by hard palate and lower lip.\(^4\) Histopathological features of oral LE lesions are mainly lichenoid mucositis, with perivascular inflammatory infiltrate, atrophy of rete pegs, liquefactive degeneration of basal epithelial cells, and patchy PAS-positive subepithelial deposits.\(^2\) Histopathological diagnosis of LE should be confirmed with DIF to rule out other lesions such as LP; DIF in oral LE lesions is frequently positive, and the most common immunoreactants identified are IgM and C3.\(^5\) There are no large or controlled studies in literature on the management of oral LE. General measures include smoking cessation and topical and oral corticosteroids. Antimalarials, azathioprine,\(^6\) methotrexate,\(^7\) dapsone,\(^8\) and gold\(^9\) have been described in isolated and small case series. Our patient's severe cheilitis responded well to oral prednisolone with no recurrence of lesions. She was subsequently treated with oral hydroxychloroquine and prednisolone for progression from SCLE to LE with good response.

Oral manifestations of EM can range from tender superficial erythematous and hyperkeratotic plaques to painful hemorrhagic bullae and erosions.\(^10\) Most patients have chronic or recurrent oral lesions, which can occur with typical and atypical target lesions. The exact pathogenesis is unknown but has been suggested
to be an immunologically mediated reaction to infectious agent (such as herpes simplex virus) or drugs. Our patient’s severe crusting was painless, with no bullae or erosions. Her episodes were not recurrent and targetoid lesions were absent.

This case demonstrates an unusual presentation of severe crusting cheilitis as a presenting feature of SLE. Our patient presented with lower lip involvement only, suggesting that this was a photoaggravated or induced condition. High ANA titers should have prompted a high suspicion for lupus cheilitis instead of EM. Close follow-up is needed, and repeat biopsies and laboratory investigations should be done as patients can progress to SLE.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.