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Clinical Challenges Q&A

13. Painful Lip and Mouth Ulceration

A 21-year-old woman presented with multiple oral ulcerations and bloody crusts on the ulcerated vermillion of both her lips. She also noted a rash, together with eye soreness and general myalgia. Her lesions began to appear 10 days after an episode of herpes labialis. She had had similar episodes of oral and lip ulcerations many times since the age of 18 and these often followed a respiratory or herpetic infection. Apart from a mild iron deficiency anaemia related to menorrhagia, for which she was treated with iron tablets, her medical history was otherwise clear.

Extra-oral examination showed lip lesions covered by bloody crusts (Figure 1). Reddish macules were also developing on the skin of her hands (Figure 2).

Oral examination showed painful ulcerations of various shapes and sizes on a diffuse erythematous background, mainly on the tongue, buccal mucosa and lips. The oral lesions were

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covered by whitish pseudo-membranes.

Q1. Which is the probable diagnosis?

- (a) Mucous membrane pemphigoid;
- (b) Herpetic stomatitis (primary);
- (c) Oral burns;
- (d) Erythema multiforme;
- (e) Behçet's syndrome.

A1: The answer to which is the probable diagnosis?

(a) *Mucous membrane pemphigoid* is a chronic autoimmune bullous disease affecting mainly the oral mucosae (95%), sometimes the eyes and rarely other mucous membranes, such as genitals, anus, pharynx and larynx and rarely the skin of the scalp, face and neck. This disease occurs more frequently in middle-aged women and presents as multiple vesicles or bullae that break leaving painful erosions which are covered by whitish pseudo-membranes. Gingival lesions present as desquamative gingivitis, while eye lesions can be conjunctivitis and symblepharon, leading sometimes to blindness. Our patient, however, had long periods free of oral lesions, while in pemphigoid such remissions are rare.

(b) *Herpetic stomatitis (primary)* is an acute HSV-1 or HSV-2 virus infection characterized by multiple small vesicles which break within 24 hours leaving painful, scattered ulcers over the oral mucosa. Lesions are always present on the gingivae which become swollen, red and tender with only a few ulcers, but associated with high fever,

malaise, fatigue and bilateral painful cervical lymphadenopathy. Secondary oral herpetic stomatitis is usually characterized by a few vesicles but without general symptomatology. Our patient had many episodes of labial and oral lesions with similar severity, suggesting that herpetic infection was not the cause.

(c) *Oral burns* can be caused by various chemical, thermal or electrical irritants when these contact the oral mucosa. Our patient denied being exposed to chemical, thermal or electrical irritants.



Figure 1. Oral ulcerations.



Figure 2. Skin macules.

(d) *Erythema multiforme* is the likely answer. It is a chronic, recurrent mucocutaneous disease possibly caused by a hypersensitivity reaction which may be triggered by infections (herpes mainly), drugs such as non-steroidal anti-inflammatory agents, or other factors. This disease is characterized by: i) large variations of skin and oral lesions regarding their type, severity and distribution; ii) the early peak of onset (at the second and third decade) and iii) a low recurrence rate (1–3 episodes/year). The initial typical skin lesion is a red purpuric macule or plaque with a clear centre surrounded by one or two pale cyanotic rings, giving the impression of an archery target or iris. These target

lesions are mainly seen on the extensor surfaces of extremities, palms, neck and face. They are often present 2–5 days before the onset of mouth, eye or genital lesions. Oral lesions are seen usually on the lips, tongue and palate as symmetrical painful erosions or ulcerations, while the eyes can become red and dry with itching, pain or discharge. Erythema multiforme can be characterized as a mild type (with only skin lesions and no symptomatology) or major type (with one or more mucosal involvements). Blood crusted lips are typical of erythema multiforme major but target skin lesions are a common finding in both types. Our patient had all the characteristics of erythema multiforme, such as the young age of onset, oral, skin or eye involvement,

the absence of severe fever or general symptomatology, the frequency of recurrence and the association with other viral infections.

(e) *Behçet's syndrome* is a rare disease characterized by an immune-mediated vasculitis of the small vessels. It presents with aphthous-like mouth ulcerations, sometimes accompanied by genital ulceration, skin lesions, like erythema nodosum and folliculitis, and eye problems, such as iritis, uveitis and optic neuritis. Other problems, such as diarrhoea and abdominal pain, pleuritis, pulmonary artery thrombosis, epididymitis, pericarditis and chronic meningo-encephalitis, are occasionally experienced.

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