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# Clinical Challenges Q&A

## 18. Oral Ulceration

A 36-year-old married woman presented for evaluation of three painful chronic ulcers (>1 month duration). Similar oral lesions had been recorded many times in the past, with the first episode reported at 3 months of age. These current ulcers presented after she had a staphylococcal skin infection but were not associated with lymphadenopathy, fever or other general symptomatology. Her medical history was otherwise clear apart from occasional skin and respiratory infections since childhood, and she was on no medication. The patient was only an occasional smoker, non-drinker and none of her family has a history of similar oral lesions.

Extra-oral examination revealed no lesions and no cervical lymphadenopathy.

Oral examination revealed chronic periodontitis and caries in most of her teeth, despite her good oral hygiene. She had one superficial ulcer in the upper

right alveolar mucosa, another on the right buccal mucosa and a third ulcer, which was ovoid and deep with well-defined but not erythematous margins, on her tongue dorsum (Figure 1).

Q1. What is the probable diagnosis?

- (a) Aphthous stomatitis;
- (b) HIV infection;
- (c) Syphilis;
- (d) Congenital agranulocytosis;
- (e) Hand, foot and mouth disease.

**A1. The answer to what is the probable diagnosis?**

(a) *Aphthous stomatitis* is a

common condition characterized by recurrent oral painful ulcerations, often from childhood, as in this patient. Three clinical types are well known, according to the ulcer's size, location and duration of healing: minor; major and herpetiform ulcers. The ulcers in our patient have some features in common with major aphthae regarding the symptomatology, the distribution and duration and the frequency of episodes. However, the ulcers in our patient were not surrounded by an erythematous halo, were not resolving in adulthood and were associated with skin and respiratory infections, so are unlikely to be aphthae, rather they are aphthous-like

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**Figure 1.** Ulcer on tongue dorsum.

ulcers. These are seen in various conditions, such as immune defects.

(b) *HIV infection* can result in oral ulceration from a number of causes, eg herpes simplex 1 or 2, zoster and cytomegalovirus infections or drugs. The ulcers in our patient and in HIV+ve patients are both sometimes preceded by a skin staphylococcal infection, but the age of onset in HIV/AIDS is generally older in developed countries and there is general symptomatology and lymphadenopathy which is not seen in our patients.

(c) *Syphilis*, an infection with *Treponema pallidum*, may present in the mouth as primary or secondary painless ulceration with generalized symptomatology. Syphilis is currently epidemic, especially in some prostitutes and men who have sex with men. The primary ulcer or chancre is usually a single

round ulcer with an indurated base and discrete rubbery cervical lymph node enlargement, while ulcers in secondary syphilis are flat, linear (snail track-like) or coalesce to form well-defined round mucous patches, and are usually associated with fever, malaise and generalized lymphadenopathy. The early onset (3 months of age) of lesions could suggest the ulcers might be manifestations of pemphigus syphiliticus (early congenital syphilis), but this is unlikely given the lack of skeletal abnormalities in our patient. The ulcers in this patient are also unlikely to be syphilitic as they are painful and are not associated with cervical lymphadenopathy.

(d) *Congenital agranulocytosis (Kostmann syndrome)* is the diagnosis in our case. Kostmann syndrome is a

rare congenital disease characterized by neutrophil maturation arrest at the myelocyte stage, causing chronic neutropenia and increased susceptibility to infections. This condition presents with mouth and skin infections or aphthous-like ulcerations and an increased tendency to periodontal disease, caries and respiratory and skin infections. Any form of neutrophil defect can cause ulceration.

(e) *Hand, foot and mouth disease* is caused by the Coxsackie A virus and is characterized by a skin rash in the extremities and mouth ulcerations which resolve within days, which is not the case here. This condition is highly infectious among school age children and close contacts, but similar lesions have not been found in friends or family members in this case.



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