BRIEF REPORT

TIMELINE IN PICTURES OF ORAL APHTHAE AS A PRESENTING SYMPTOM FOR BEHCET'S DISEASE

Adam Ramsey, MD[1]; Abdelkader Mallouk, MD[1]; Amit Sharma, DO[1]; Timothy Baxter, MD[1]

[1] Family Medicine Residency Program, Arnot Ogden Medical Center, Elmira, NY

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Behcet's Disease

Inflammatory Disorder

Oral Aphthae

Oro-Genital Ulcers

ABSTRACT: Behcet's Disease (BD) is a chronic relapsing and remitting vasculitis with an unknown cause. With its propensity to involve all size arteries and veins and the ability to affect all organ systems, BD can result in significant mortality. BD is commonly referred to as the "silk road" due to the high incidence of BD in the ancient Mediterranean trading route known as "Old Silk Road." A timeline in pictures of oral aphthae is presented to emphasize the need for increased awareness among clinicians to recognize the various manifestations of BD to diagnose and offer prompt, timely treatment. The evidence base for treatment is limited and further studies are needed to ascertain the prevalence and distribution as well as associated genetic factors of BD in the U.S.

INTRODUCTION

Behcet's disease (BD) is a chronic, relapsing, inflammatory vascular disease with no pathognomonic test. It is named after the Turkish dermatologist, Hulusi Behcet, who described the disease in 1937, characterized by recurrent oral ulcers and several other systemic manifestations.[1] It is believed to exist in many parts of the world with high incidences in the Middle East, Far East, Mediterranean region and an area of the ancient trading route known as "Old Silk Road" between latitudes 30° and 45° north in Asia and Europe.[2,3]

BD is thought to be an autoimmune over-reaction to either an infectious or environmental insult in a subset of patients genetically predisposed with an HLA-B51 genetic risk factor. BD typically presents in the third and fourth decade of life with no specific sex predilection.[1,3,4] The diagnosis relies on the clinical criteria according to the International Study Group for Behcet's Disease (ISGBD).[5] (Table 1)

Low sensitivity of the currently applied International Study Group (ISG) clinical diagnostic criteria led to their reassessment. An international team for the revision of the international criteria for BD (from 27 countries) submitted data from 2,556 clinically diagnosed BD patients and 1,163 controls with BD-mimicking diseases or presenting at least one major BD sign. For the International Criteria of Behcet's Disease (ICBD) ocular lesions, oral aphthosis and genital aphthosis are each assigned two points. In contrast, skin lesions, central nervous system involvement and vascular manifestations are one point each. The pathergy test, when used, is assigned one point. A patient scoring ≥4 points is classified as having BD.[6] Corticosteroids, immunosuppressant drugs, tumor necrosis factor-inhibitors and other symptomatic treatments are commonly used in the management of BD.[7] We report a case of a 60-year-old Turkish male who presented with a history of the recurrent oral ulcers of BD with the chronological order of oral aphthae development documented in illustrated pictures.

TABLE 1:

ISGBD clinical Behcet's Disease diagnosis criteria[5]

ISGBD REQUIRES THE PRESENCE OF RECURRENT ORAL APHTHAE (THREE TIMES IN ONE YEAR) WITH AT LEAST TWO OF THE FOLLOWING:
• Recurrent genital aphthae (aphthous ulceration or scarring)
• Eye lesions (retinal vasculitis, cells in vitreous or uveitis)
• Skin lesions (papulo-pustular lesions, pseudo-vasculitis, acneiform nodules or erythema nodosum)
• Positive pathergy test

EPIDEMIOLOGY

A high prevalence of BD (420 per 100,000) has been reported in Turkey[1] with the lowest prevalence of 0.38 per 100,000 being reported in North America.[2] In sub-Saharan Africa, the prevalence of BD is not known as few cases have been reported,[8–10] with only one case being reported from Tanzania over 40 years ago.[11]

CORRESPONDENCE:
Adam Ramsey, MD | aramsey@arnothealth.org

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CASE REPORT

A 60-year-old Turkish male presented to our outpatient clinic frustrated by multiple visits to many doctors and clinics for his seemingly puzzling symptoms. The patient initially had blisters over his lips and tongue. He went to a walk-in clinic and was told he had HSV-1. The patient also had a blister on his penis. However, he denied being sexually active for years, which prompted us to conduct further investigation and search the literature. Two weeks later, the patient stated that the ulcers were healing and everything was beginning to look normal again. The next day the patient woke up with back pain radiating to the right side of the mid-thoracic area for which he went to the emergency department (ED). He was diagnosed with a kidney stone on CT imaging. The patient described two similar episodes of oral ulcerations over the previous year. The mouth ulcers started gradually in the buccal cavity, tongue and lips. There have been periods of complete healing and recurrences. His recurrences were neither bleeding nor discharging. The patient also had recurrent genital ulcers. He denied a history of epigastric pain, painful defecation, painful micturition, hematuria, reduced amount of urine or any history suggestive of sexually transmitted diseases in the past. There was no blurred vision or photophobia. Throughout his illness, the patient had neither fever nor weight loss.

The patient had no history of allergies and had never been transfused with blood or blood products. All of his family members were healthy and none had similar illnesses. He is a former smoker and has no history of alcohol use.

Physical examination revealed an anxious patient with multiple concerns about his health who was fully alert, cooperative and afebrile. He had no oro-genital ulcerations, eye lesions or skin abnormalities at the time of the exam. There was no lymphadenopathy. Eye examination revealed normal visual acuity, normal visual fields, normal optic nerves and no signs of uveitis.

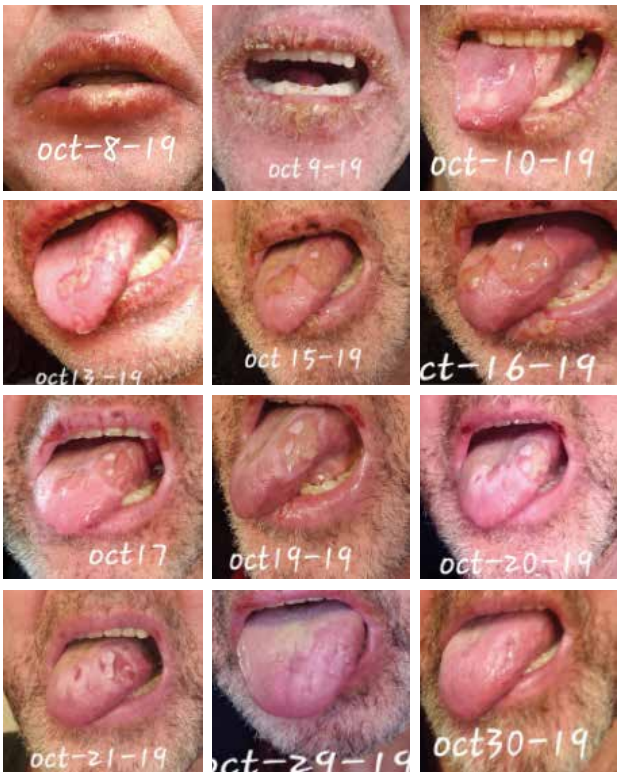
His blood pressure was 122/82 mmHg, pulse rate 68 beats per minute, respiratory rate 17 cycles per minute and oxygen saturation 96% in room air. Urogenital system examination revealed normal male genitalia. The physical examination of the rest of the systems was essentially normal.

The results from the laboratory analysis done were complete blood count (hemoglobin level of 14.6 g/dL and a mean corpuscular volume 93.7 fl., all blood cell counts were within normal ranges), renal function test (normal range), fasting blood glucose (92MG/DL) and pathergy test (negative). HLA-B27 antigen negative. The Erythrocyte Sedimentation Rate (ESR) was 18MM/HR and C-Reactive Protein (CRP) was 1.03mg/L. Herpes simplex Virus (HSV) Type 1 & 2 were undetected in the serum. Varicella-zoster virus (VZV) PCR was negative. The diagnosis of BD was made according to the ISGBD5 based on the presence of recurrent oral aphthae (≥3 times in one year) together with the self-reported genital aphthae and characteristic skin lesions.

The patient required no treatment since the disease course seemed to be in remission. Close monitoring of the various systemic symptoms of BD was advised and appropriate follow-up recommended. Generally, BD responds well to corticosteroids, with the combination of corticosteroids and immunosuppressant drugs being indicated when vital organs are involved.[7]

FIGURE 1:

Timeline in pictures of oral aphthae



CONCLUSION

This case emphasizes the need for increased awareness among clinicians to recognize the various manifestations of BD to diagnose and offer prompt treatment. Further studies are needed to ascertain the prevalence and distribution of BD in the U.S. as well as associated genetic factors.

AUTHOR DISCLOSURE(S):

The author(s) declare no relevant financial affiliations or conflicts of interest.

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