



## Case Report

## Behcet's disease: Diagnosed as isolated recurrent oral aphthae; a case report

Maysoun Kudsi<sup>a,b</sup>, Naram Khalayli<sup>a</sup>, Amr Allahham<sup>b,\*</sup><sup>a</sup> Faculty of Medicine, Damascus University, Damascus, Syria<sup>b</sup> Faculty of Medicine, Syrian Private University, Damascus, Syria

## ARTICLE INFO

## Keywords:

Behcet's disease  
 Oral aphthae  
 Dapsone treatment

## ABSTRACT

**Background:** Behçet's disease is a multisystem autoimmune disorder of unknown origin, characterized by frequent oral and genital ulcerations, ocular and cutaneous lesions, arthritis, and it may also involve the gastrointestinal, and central nervous system. In this paper we discuss a case of Behcet syndrome presenting with isolated oral aphthous initially and spotlight on the clinical features and treatment.

**Case report:** A case of a 32-year-old male with repetitive oral aphthous on the tongue and the inner cheeks for four days with a history of frequent recurrence. The patient responded to topical triamcinolone acetonide and 5–10 mg/day prednisone. Three weeks later, there was a recurrence of oral aphthous, genitalia lesions, and blurred vision with left eye redness. A uveitis diagnosis was made, and the patient was placed on 75mg (1mg/kg) prednisone with a resultant response. There was another recurrence with all the previous symptoms, so the decision with Dapsone treatment was made. The patient was placed on Dapsone, resulting in complete resolution of the lesions with no reproduction within and after six months of several recall visits. The diagnosis of Behcet syndrome was made.

**Discussion:** Aphthous ulcers may be seen in a variety of disorders, while recurrent and persistent >6 weeks, individual non-healing ulcers require a review of the diagnosis. Behcet disease usually appears 2–3 years after oral and genital ulcers, but oral aphthae may be its first manifestation in 10–20% of cases. AS oral ulcers are among the common complaints of the polyclinics-patients, it is recommended to educate the health professionals to diagnose these unusual cases in the first instance, thus avoiding underdiagnosed and the time management to reduce complications and improve the quality of life in these patients.

**Conclusion:** A case of Behçet's disease in a 32-year-old male who presented initially with oral mucosa and later caused uveitis and genitalia aphthae.

## 1. Introduction

Behcet disease, also known as Silk Road disease, is a chronic multi-system autoimmune syndrome of unknown origin [1]. Its prevalence ranges between 0.1/1000 and 1/10,000, with a higher incidence in Asia, Mediterranean countries, and Japan; it remains rare in other countries such as the Americas [2]. Its peak incidence in the third decade, with women predominance [3]. It represents an autoimmune reaction triggered by an infectious agent in a genetically predisposed person [4]. Genetic studies should that HLA-B\*51 is a significant genetic risk factor [5]. It affects all organs such as the skin, the eye, the blood vessels, gastrointestinal tract, lungs, central nervous system, heart, and others [6]. Painful recurrent mouth ulcers may occur at some time in all patients. They begin as raised, round lesions that quickly turn into painful ulcers. They're often the first noticed symptom and may occur 2–3years

before any other symptoms of Behcet disease appear. In addition, they may be the first manifestation of the disease in 10–20% of cases [7,8]. Multiple criteria have been suggested as guides for diagnosis, although some cases remain undiagnosed [9]. The American Criteria of Rheumatology (ACR) put five criteria for diagnosis: oral aphthae, genital aphthae, skin features, eye involvement, and positivity of pathergy test [10].

We present a case of Behcet disease in a male patient who presented initially with the frequent, persistent isolated occurrence of oral aphthae.

This case series has been reported in line with the SCARE Criteria [23].

Research Registry unique identifying number (UIN) 7991 [24].

\* Corresponding author. Sharqi Rkn Aldeen, Damasus, Syria.

E-mail address: [Amroapple98@gmail.com](mailto:Amroapple98@gmail.com) (A. Allahham).

<https://doi.org/10.1016/j.amsu.2022.104327>

Received 15 June 2022; Received in revised form 30 July 2022; Accepted 31 July 2022

Available online 24 August 2022

2049-0801/© 2022 The Authors. Published by Elsevier Ltd on behalf of IJS Publishing Group Ltd. This is an open access article under the CC BY license (<http://creativecommons.org/licenses/by/4.0/>).

## 2. Case report

A 32-year-old married male hand worker presented to the Rheumatology clinic as consultation from the Dental clinic, Damascus University, with a two-year history of frequent recurring oral aphthae. No considerable family history nor psychosocial history has been reported by the patient. He was on topical steroid-triamcinolone acetonide 0.1%, colchicine 1mg daily for four weeks. Following several follow-up visits, the ulcers healed. After six months, the patient presented with recurrent aphthous ulcers on the tongue and inner cheeks for four days.

On examination, a pale oval floor with erythematous margins was found. He was placed on topical triamcinolone acetonide twice daily for seven days, Prednisolone 10mg daily for seven days followed by 5 mg for the next five days, and tetracycline mouth washes four times daily for seven days. The patient responded well with the healing of the ulcers after the steroid therapy. However, just after two weeks, there was a recurrence of the oral aphthous ulcers with erosions, genitalial lesions, and blurred vision with the red left eye (figure-1,2).

According to the ophthalmology consultation, acute anterior uveitis was seen. Our patient was placed on 75mg/OP/day (1mg/kg) Prednisone for seven days, tapering down gradually by 5 mg after every seven days; continue the topical triamcinolone acetonide. There was magnificent healing of the ulcers and erosions, uveitis, and genital ulcers after two weeks of treatment. Again, after four weeks, there was a relapse with manifestations of sore throat, genitalia ulcerations, redness, left eye pain and poor vision, and associated abdominal pain. Based on these findings, the diagnosis of Behcet's syndrome was made according to the ACR criteria. To avoid the side effects of using systemic steroids, the patient was placed on Dapsone 25 mg twice daily for one week and continued with the topical triamcinolone acetonide. The patient was also referred to the ophthalmology clinic, where he was managed for uveitis. Complete resolution of all clinical manifestations happened, with no recurrence within six months of several visits.

## 3. Discussion

Behcet's disease is a multiorgan disease that affects men more than women [11], with a peak incidence in the third decade [12]. Our patient is a male presented in his fourth decade. As there were no previous studies about age and gender in Syria, more extensive studies would help determine the concordance with the international studies.

The American Criteria of Rheumatology (ACR) put five criteria for diagnosis of Behcet disease, which include the presence of recurrent oral ulceration along with any two of the following: recurrent genital ulceration, ocular lesions, and cutaneous lesions, or positive pathergy test results [10]. Almost everyone with Behcet's disease develops mouth ulcers. Thus, these ulcers look the same as typical mouth ulcers, but can be more painful, more frequent, multiple, and larger, found on the lips, tongue, and inside of the cheek, and usually appears 2–3 years before the onset of Behcet disease, as its first manifestation in 10–20% of cases [6, 7].

Our patient had an oral aphtha which was the first presentation, then he had genital ulcer and uveitis, which made the diagnosis of Behcet's disease.

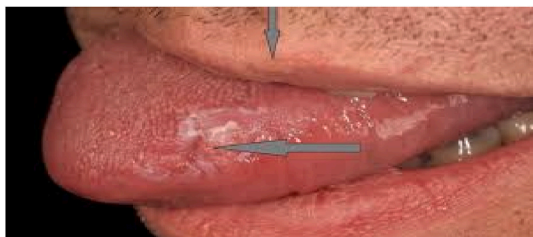


Fig. 1. Major Aphthous ulcer on the tongue.



Figure-2. genitalial aphthae.

Many treatments have been used to decrease morbidity and mortality [13]. The treatment includes steroids, immunosuppressive, and biological therapies [14].

Corticosteroids can help reduce the inflammation associated with Behcet's disease. Depending on the specific symptoms being treated, corticosteroids are used as topical, oral, and injections [1,6]. Topical corticosteroid side effects are uncommon, but long-term use may lead to thinning of the skin. The long-term use of oral corticosteroids is associated with more side effects; some of them are serious [15].

To avoid our patient steroids side effects, we used Dapsone. Dapsone has been reported to treat mucocutaneous manifestations [6], reducing the frequency and duration of oral and genital ulcers [16]. Although it has side effects [17], our patient had no adverse effects.

Some diseases such as erythema multiforme, Steven-Johnson syndrome, recurrent aphthous stomatitis, herpetic stomatitis, and complex aphthosis may mimic Behcet disease [6].

Erythema multiform shows hemorrhagic and crusting ulceration with multiple erythematous patches on the skin and mucosa [18]. Steven Johnson's syndrome is characterized by polymorphic skin lesions, acute blisters, and mucous membrane erosions [19]. Complex aphthosis is an uncommon, persistent type of recurrent aphthous stomatitis which may also be associated with systemic diseases [20].

In medical literature, limited case reports have been published about oral ulcers as a first manifestation of Behcet disease, which was first described in 1981 (21). In our case, the oral ulcers were presented 2–3 years before the diagnosis of Behcet disease. This extended time of misdiagnosis may lead to inappropriate treatment, and thus undesired complications and decreasing the quality of life in these patients. Within two weeks, he had a genital ulcer, uveitis, which may cause blindness, and gastrointestinal involvement, which is considered an essential manifestation of active disease; meanwhile, the previous case reports, the duration was about one year till the other symptoms like genital ulcers, conjunctiva inflammation, erythema nodosum, and skin ulcers appeared [6,21,22]. No data are available about the economic impact of this condition, but the high rate of morbidity and mortality of the disease, due to the major organ involvement, should be considered.

## 4. Conclusion

We report a case of Behcet's syndrome presented initially with just recurrent oral aphthae. Awareness of this rare condition should be thought of by Healthcare professionals.

## Ethical approval

This study was ethical approved by faculty of medicine, Damascus University.

## Sources of funding

The study has no funding of any sources.

## Author contribution

All the authors of this paper have contributed to the study concept, data collection, data analysis, and writing the paper.

## Conflicts of interest

No conflicts of interest.

## Registration of research studies

1. Name of the registry: Behçet's Disease: Diagnosed as Isolated recurrent oral aphthae; A case report
2. Unique Identifying number or registration ID: researchregistry7991
3. Hyperlink to your specific registration (must be publicly accessible and will be checked): [Browse the Registry - Research Registry](#)

## Guarantor

Amr Allahham, [amroapple98@gmail.com](mailto:amroapple98@gmail.com).  
Maysoun Kudsı, [MayKuds@gmail.com](mailto:MayKuds@gmail.com).

## Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

## Annals of medicine and surgery

The following information is required for submission. Please note that failure to respond to these questions/statements will mean your submission will be returned. If you have nothing to declare in any of these categories then this should be stated.

## Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.amsu.2022.104327>.

## References

- [1] Jagdish R. Nair, Consultant rheumatologist and Robert J Moots. Behçet's disease, *Clin. Med.* 17 (7) (2017 Feb) 71–77.
- [2] Fereydoun Davatchi, Cheyda Chams-Davatchi, Hormoz Shams, Farhad Shahram, Abdolhadi Nadji, Massoomeh Akhlaghi, Tahreh Faezi, Zahra Ghodsi, Bahar Sadeghi Abdollahi, Farimah Ashofteh, Negin Mohtasham, Hoda Kavosi, Mariam Masoumi, Behçet's disease: epidemiology, clinical manifestations, and diagnosis, *Expet Rev. Clin. Immunol.* 13 (1) (2017 Jan) 57–65, <https://doi.org/10.1080/1744666X.2016.1205486>. Epub 2016 Jul 11.
- [3] Didar Ucar-Comlekoglu, Austin Fox, H. Nida Sen, Gender Differences in Behçet's Disease Associated Uveitis. *J Ophthalmol* 2014 (2014), 820710.
- [4] Bainan Tong, Xiaoli Liu, Jun Xiao and Guanfang Su \*mmunopathogenesis of Behçet's Disease. *Front. Immunol.*, 29 March.2019.<https://doi.org/10.3389/fimmu.2019.00665>.
- [5] Mitsuhiko Takeno, The association of Behçet's syndrome with HLA-B51 as understood in 2021, *Curr. Opin. Rheumatol.* 34 (1) (January 2022) 4–9, <https://doi.org/10.1097/BOR.0000000000000846>.
- [6] Okoh Dickson Sopuru, Delta State Asaba, Izevbigie Isaac, Behçet disease presenting initially as isolated recurrent aphthous ulcers only: a case report, *Med. Case Rep. Rev.* (2018), <https://doi.org/10.15761/MCRR.1000119>.
- [7] J.R. Nair, R.J. Moots, Behçet's disease, *Clin. Med.* 17 (1) (2017) 71–77.
- [8] Mehmet Yildiz, Fatih Haslak, Amra Adrovic, Sezgin Sahin, Oya Koker, Kenan Barut, Ozgur Kasapcopur, Pediatric Behçet's disease, *Front. Med.* (03 February 2021), <https://doi.org/10.3389/fmed.2021.627192>.
- [9] Müzeyyen Gönül, Arzu Kılıç, Bilgen Gençler, The history and diagnosis of Behçet's disease, Different Aspects of Behçet's Disease (November 7th, 2019), <https://doi.org/10.5772/intechopen.89927>.
- [10] Hasan Yazici, Yusuf Yazici, Criteria for Behçet's disease with reflections on all disease criteria, *J. Autoimmun.* 48–49 (Feb-Mar 2014) 104–107, <https://doi.org/10.1016/j.jaut.2014.01.014>. Epub 2014 Jan 21.
- [11] Mohamad J. Zeidan, David Saadoun, Patrice Cacoub, Behçet's disease physiopathology: a contemporary review, *Auto Immun Highlights* 7 (1) (2016 Dec) 4.
- [12] F. Ajose, Oderinlo Adelowo, Clinical presentations of Behçet's disease among Nigerians: a 4-year prospective study, *Int. J. Dermatol.* 54 (2015) 889–897.
- [13] Erkan Alpsoy, Pietro Leccese, Giacomo Emmi, Shigeaki Ohno, Treatment of Behçet's disease: an algorithmic multidisciplinary approach. *Front. Med.* <https://doi.org/10.3389/fmed.2021.624795>, 28 April 2021.
- [14] F. Leccese, Y. Ozguler, R. Christensen, et al., Management of skin, mucosa and joint involvement of Behçet's syndrome: a systematic review for update of the EULAR recommendations for the management of Behçet's syndrome, *Semin. Arthritis Rheum.* 48 (2019) 752.
- [15] Nicole Davey-Ranasinghe, Herbert S. Diamond, Behçet disease treatment & management, *Med. Scape Updated* (Dec 21, 2020). <https://emedicine.medscape.com/article/329099-treatment>.
- [16] Burçin Cansu Bozca, Erkan Alpsoy, Experimental therapeutic solutions for Behçet's disease, *J. Exp. Pharmacol.* 2021 (13) (2021) 127–145, <https://doi.org/10.2147/JEP.S265645>.
- [17] Gottfried Wozel, Christian Blasum, Dapsone in dermatology and beyond, *Arch. Dermatol. Res.* 306 (2) (2014) 103–124.
- [18] T. de Risi-Pugliese, E. Sbidian, S. Ingen-Housz-Oro, L. Le Cleach, Interventions for erythema multiforme: a systematic review, *J. Eur. Acad. Dermatol. Venereol.* 33 (5) (2019 May) 842–849 [PubMed].
- [19] J. Auyeung, M. Lee, Successful treatment of stevens-johnson syndrome with cyclosporine and corticosteroid, *Can. J. Hosp. Pharm.* 71 (4) (2018 Jul-Aug) 272–275 [PMC free article] [PubMed].
- [20] J.A. Letsinger, M.A. McCarty, J.L. Jorizzo, Complex aphthosis: a large case series with evaluation algorithm and therapeutic ladder from topicals to thalidomide, *J. Am. Acad. Dermatol.* 52 (3 Pt 1) (2005 Mar) 500–508.
- [21] K v Venkatasubramaniam, Behçet's syndrome: case report and literature review by henry ford hosp, *Med* 29 (3) (1981).
- [22] Kelly Cristine Tarquinio, Clinical reportThe treatment of oral lesions in Behçet's Syndrome: *Rev. Española Cirugía Oral Maxilofac.* 38 (2) (April–June 2016) 101–104.
- [23] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus surgical CAse REport (SCARE) guidelines, *Int. J. Surg.* 84 (2020) 226–230.
- [24] <https://www.researchregistry.com/register-now#user-researchregistry/registersearchdetails/62a1ef894e50b80022ecb547>.