



# Asian Journal of Hospital Pharmacy

Available at [www.ajhp.online](http://www.ajhp.online)

ISSN: 2583-0724



## Behcet's disease (silk road disease): a rare inflammatory disease

Leya P. Babu<sup>1</sup>, Shaji George<sup>2</sup>, Johnson V. Babu<sup>1</sup>, Nimmy Robin<sup>1</sup>, Joicy Jose<sup>1</sup><sup>1</sup>Pharm.D Interns, Nirmala College of Pharmacy, Muvattupuzha. Kerala-686661<sup>2</sup>Professor and Head, Department of Pharmacy Practice, Nirmala College of Pharmacy, Muvattupuzha. Kerala-686661*Received: 03 Oct 2021 Revised: 18 Oct 2021 Accepted: 22 Dec 2021*

### Abstract

**Background:** Behcet's disease (Silk Road disease), a rare immune-mediated multisystem inflammatory disorder described by intermittent oralaphthae and genital ulcer, backsliding uveitis, mucocutaneous, articular, gastrointestinal, neurological and vascular manifestations, with no cure. It is brought about by changes in the: a) arteries that flexibly blood to the body tissues b) veins that return the blood to the lungs, the rear of the eyes retina, brain, joints, skin and bowels.

**Case Presentation:** A 55-year-old male patient was sensed with c/o joint agony in lower appendages, oral ulcer and scrotal ulcer. On physical assessment the patient was cognizant and oriented with B/L lower leg joint emanation. All lab examination including RA factor was within normal limits, with diminished Serum Vitamin D. HLA B51, ANA were checked and oral mucosal biopsy was done. The most punctual sign exhibited was oral disintegration, various shallow ulcer and scarcely any dissolved knobs in the scrotum. At that point the patient gave joint pain and numbness on right leg. On neurological assessment, a strange motor nerve conduction saw with right tibial neuropathy. At first, doubt with syphilis and tarsal tunnel disorder and following 7-8 days of affirmation, analyzed as Behcet's illness dependent on dermatological, rheumatologic and neurological signs. Treatment given was symptomatic and supportive with pain relievers, corticosteroid, antibiotics, IV fluids, PPI, vitamin supplement, laxative and local anaesthetic.

**Discussion:** Without adequate data it's difficult to examine, in light of anomaly and standardized treatment are questionable at present. New information with respect to its immunopathogenesis, genetics will significantly help in the advancement of research center tests, diagnostic criteria and particularly in the decision of the best treatment

**Keywords:** Behcet's disease, Silk Road disease, Ulcers, Triad, Multisystem inflammation.

This article is licensed under a Creative Commons Attribution-Non Commercial 4.0 International License. Copyright © 2021 Author(s) retain the copyright of this article.



### \*Corresponding Author

Shaji George

Email: [shajige@gmail.com](mailto:shajige@gmail.com)DOI: <https://doi.org/10.38022/ajhp.v1i2.33>

### Introduction

Behcet's disease was portrayed by Hulusi Behcet in 1937 as a rare chronic relapsing inflammatory process of obscure aetiology, characterised by an atopic artery in the mouth mucus, genital mucus ulcerations and eye changes. These are significant manifestations, joined by different symptoms in the skin as erythematosus, disseminated papillomas, pustules and purple and no dose shifts. Notwithstanding the skin, different organs, for example, rheumatoid arthritis, thrombophlebitis,

Gastrointestinal issues, kidney, respiratory framework, CNS changes with symptoms such as psychiatric meningoencephalitis and psychogenic alterations [1].

Behcet's disease is most common along the "Old Silk Route," which traverses the region from Japan and China in the Far East to the Mediterranean Sea, including nations such as Turkey and Iran. Despite the fact that the disease is uncommon in the United States, irregular cases do happen in patients who might not seem, by all accounts, to be in danger in view of their ethnic background (e.g., in Caucasians or African-Americans). The infection isn't uncommon in locales along the Old Silk Route, yet the sickness' the disease's epidemiology isn't surely known. In Japan, Behcet's

sickness positions as a main source of visual deficiency [3].

The aetiology is not known, however in the viral or autoimmune genesis, but isn't yet affirmed by significant examination. Behcet disorder is believed to be brought about by a combination of genetic and environment factors. The presence of HLA-B51 allele is a predominant factor for the development of BD [1].

Mucocutaneous manifestations are markers of BD. The primary symptom perceived in between 25 and 75% of cases are the oral injuries, which are showed in a same region during the whole course of disease in practically all patients. Such lesions are like the typical aphthous ulcers that are naturally introduced in an incredible number of sores (at least six), occurring mainly on the soft palate, lips, tongue, gingiva, oral mucosa and oropharynx, while the larynx and the nasal mucosa are seldom influenced [2].

The genital ulcers seem, by all accounts, to be the second incessant sign in the disease's cases and are found in a vast dominant part of patients. The perianal lesions are found in both gender and their development is like what occurs in the mouth. In women, sores happen for the most part on the lips, vulva and vaginal wall, and may have painless signs that go unobserved, and in these cases, the conclusion is made through pelvic examination [2].

The most frequent ocular manifestations are posterior uveitis, retinal vasculitis, conjunctivitis, optic neuritis and retinal joint inflammation, yet anterior uveitis with hypopyon (presence of discharge in the anterior chamber of the eye) is the classical sign in the start of the Behcet's disorder. These ocular sores might be reversible.

Over half of the patients may present with articular manifestations such as monoarthritis or polyarthritis and may precede, accompany or follow other manifestations of the Behcet's disorder. Due to inflammation of the central nervous system neurological manifestations occur and are not very frequent, because of vascular involvement at the area or as a result of fringe polyneuropathy.

Vascular manifestations are fluctuated and rely upon the sort and area of the vessel in question and they are presented as shallow thrombophlebitis, venous apoplexies or as arteritis. Gastrointestinal manifestations are common and they include abdominal pain, melena and diarrhoea. Patients may also present with pulmonary manifestations such as pulmonary vasculitis and pulmonary arterial aneurysm formation [2].

The specific criteria proposed by the International Study Group for Behcet specifies those recurrent ulcers: smaller naphtha's, larger naphtha's or herpetiform ulcers observed in a minimum of three episodes during a period of 12 months, and two of any of the following manifestations: recurrent genital ulcers (ulcers or genital ulcer scars); joint manifestations; ocular lesions (anterior or posterior uveitis, or presence of cells on the vitreo during ocular exam, or also retinal vasculitis); cutaneous lesions (erythematous nodules, pseudo folliculitis, papular-pustulous lesions or also acne form nodules observed in post-adolescent patients not treated with corticosteroids); analysis of the pathergy signal test.

The primary goal of BD treatment is to induce and maintain remission and improve quality of life, preventing irreversible damage and exacerbation of mucocutaneous and articular disease. Corticosteroids, immunosuppressant drugs, TNF inhibitors, and other supportive medications are normally utilized in the management of BD [1].

### Case Presentation

The case report is of a male patient, 55 year old, who was admitted for the diagnosis and treatment for joint pain in both lower limbs, recurrent oral and scrotal ulcer. The disease started one month ago, first with oral erosion, multiple shallow ulcer and few eroded nodules in the scrotum. At that point following 2 weeks, the patient shows joint pain and numbness on right leg. The changes have stayed for one week and on neurological examination, an abnormal motor nerve conduction observed with right tibial neuropathy. Initially, suspicion with syphilis (because of scrotal ulcer) and tarsal tunnel syndrome (because of neuropathic pain) and after 7-8 days of admission, diagnosed as Behcet's disease based on dermatological, rheumatologic and neurological manifestations.

All vitals were normal.

On CBC examination all parameters were normal (Hb: 12.5 gm. %, WBC: 12080, N: L: E: B: M- 90:24:1:0:3, ESR: 20mm/hr). LFT examination revealed Bilirubin (D): 0.3 mg/dl, Bilirubin (ID):0.8 mg/dl, Bilirubin (D): 1.1 mg/dl, SGOT/PT: 27/38 U/L, S. Albumin: 3.7 mg/dl, Globulin: 3.8 mg/dl, A/G ratio: 1:1.

The parameters observed in RFT were BUN: 39 mg/dl, S. Creatinine: 0.6 mg/dl. Serum electrolytes like Na<sup>+</sup> (133 mmol/l) and K<sup>+</sup> (3.8 mmol/l) were found to be normal. As a result of starting doubt with syphilis VDRL was checked and was seen as negative. The patient shows

joint pain, so RA factor were checked yet it was typical (9.2 u/ml).

Because of the presence of oral ulcer serum vitamin B12 were checked and it was seen as normal (370.9 pg/mL) yet serum nutrient D were decreased (14.6 pg/ml). ANA profile were investigated and the outcome was negative. HLA B51 is firmly connected with Behcet's illness and it was found to be positive.

On dermatological consultation there were multiple shallow ulcer and few eroded nodules in the scrotum and orthodontic consultation bilateral ankle arthritis were observed. Due to numbness on right leg neurological examination was carried out and observed that the right tibial CMAP (Compound muscle action potential) is reduced and the patient have right tibial neuropathy.

The goal of the treatment is to reduce the discomfort and to prevent serious complications. Treatment given was symptomatic and supportive with pain relievers, corticosteroid, antibiotics, IV fluids, PPI, vitamin supplement, laxative and local anaesthetic. The medications administered are Inj. dexona 8mg (dexamethasone) which is the mainstay of BD treatment and other supportive therapy like Tab. Flupirtine 100mg BD which is a pain reliever, Inj. Esomeprazole 40mg OD, Tab. Clonotril 0.25mg stat, Kenacort paste for LA, Lignocaine for LA, Inj. Levofloxacin 500mg OD, Syp. Lactulose 15ml TID, Tab. Riboflavin 10mg 0-0-1 and Metrogyl DG gel for LA. Generally BD responds well to corticosteroids and our patient responded to steroids within 14 days of treatment.

### Discussion and Conclusion

The silk worm disease is one of the rarest type of incendiary infection which mainly affecting blood vessels. It can cause serious complications due to the involvement of vital organs. Many cases of BD remain undiagnosed or unreported due to lack of awareness among clinicians.

Our patient had typical presentation of BD but due to the overlap of signs and symptoms between Behcet's disease and that of syphilis and Tarsal tunnel syndrome is a challenge to specialists for building up a precise finding. In our case based analysis report, based on history and literature consultations, we have inferred that we are dealing with Behcet-syndrome. Without sufficient information it's hard to analyze, because of its rarity and standardized treatment protocol disputable at present. To reduce mortality and morbidity in these

patients there must be acceptable collaboration among specialist and doctor.

### Conflict of Interest

Nil

### References

1. Ferizi M, Gerqari A, Ferizi M. Behçet's Disease- Case Presentation and Review Literature. Open Access Macedonian Journal of Medical Sciences. 2018; 6(10):1871.
2. Marinho KC, Caputo BV, Noro-Filho GA, Giovani EM. Behçet's syndrome: literature review and clinical case report. Revista Española de Cirugía Oral y Maxilofacial. 2016; 38(2):105-10.
3. Meda JR, Seni J, Mpondo B, Peck RN, Jaka H, Kilonzo SB. Behcet's disease presenting with recurrent ocular, oral, and scrotal inflammatory lesions in a young Tanzanian man: a case report. Clinical case reports. 2014 (4):133.