THERAPEUTIC ADVANCES in
Rare Disease

Case Report

Behcet's disease in a tertiary eye hospital in Pakistan

Ther Adv Rare Dis

2025, Vol. 6: 1–5

DOI: 10.1177/
26330040251314126

Tabish Ali Shalwani, **Alishan Khowaja** and **Narmeen Punjwani**

***Abstract*:** Behçet's disease is a kind of variable vessel vasculitis (VVV) and inflammatory systematic disease affecting various organs of the body. The cause of the disease is idiopathic but is most commonly genetic in origin. A positive skin prick test (dermatographia), genital sores, eye irritation, skin sores, and at least three episodes of mouth sores in a year confirm the diagnosis. Treatment may include immunosuppressive agents, immune modulators, and biological markers such as corticosteroids, immunosuppressants, and antibodies. We report a case of a 23-year-old male patient, presented in an outpatient clinic in a tertiary care eye hospital located in Pakistan. The patient reported sudden loss of vision in one eye and graduate loss of vision in the other eye. Ocular and systemic investigations were performed to correlate with clinical findings to reach a diagnosis. The patient was managed symptomatically and was put on corticosteroids. Our hospital is a research and postgraduate educational institution that deals with complex eye diseases. The range of investigations and clinical exams helped clinical decision-makers in evaluating the patient's diagnosis.

Plain language summary

A rare eye disease in Pakistan

A 23-year-old male patient consulted an outpatient clinic in a tertiary care ophthalmology hospital with sudden loss of vision. On detailed head-to-toe assessment and laboratory investigations, the patient was diagnosed with Behcet's disease. This is a rare disease that is more common in the Middle East and some parts of Asia. The disease has no specific treatment and is managed for symptoms mainly. The patient was treated for his symptoms including ulcers and rashes on different parts of the body and underwent a surgical eye procedure to improve his vision. The patient was under the care of a medical physician as well as an ophthalmologist for treatment of his disease. This case presented that rare diseases such as Behcet's that affect multiple parts of the body can be treated using a multidisciplinary approach.

Keywords: Behchet's disease, hypopyon, Pakistan, uveitis

Received: 24 April 2024; revised manuscript accepted: 23 December 2024.

Correspondence to:
Tabish Ali Shalwani
Liaquat University of Medical and Health Sciences, Jamshoro 74800, Pakistan
tabish.shalwani@hotmail.com

Alishan Khowaja
Liaquat College of Medicine and Dentistry, Karachi, Pakistan

Narmeen Punjwani
Jinnah Medical and Dental College, Karachi, Pakistan

Introduction

Behçet's disease (BD) is a kind of inflammatory disorder that affects several body organ systems.[1] Inflammation of the eye tissues, arthritis, and painful sores in the mouth along with mucous membranes of various organs are the most typical symptoms.[1] The sores may persist for a few days, a week, or longer. Moreover, blood clots, aneurysms, inflammation of the brain or spinal cord, and blindness are typical features of Bechet's disease.[1] The symptoms are frequently recurrent with remittances and relapses.

The cause of the disease is idiopathic[2] but is assumed to be associated with genetic causes.[2] BD is noncontagious, and the triad of signs including genital ulcers, eye inflammation, and at least three episodes of mouth ulcers in a year along with a positive skin prick test/dermatographia, pathergy test, and human leukocyte antigen (HLA) complex B51 confirm the diagnosis.[2]

There is no specific treatment for BD. Treatments may include immunosuppressive medication such as corticosteroids and immunomodulators with lifestyle changes being the mainstay of treatment. Mouthwash containing lidocaine could ease the pain. Colchicine may lessen frequent attacks.[3]

While rare in the United States and Europe, it is more common in the Middle East and some parts of Asia.[4] The onset of Behcet's disease is usually in the second or third decade with male predominance.[1] The disease was initially described by Turkish dermatologist Hulusi Behcet in 1937.[5]

Case report

We report a case of a 23-year-old male patient who presented in an outpatient clinic in a tertiary care eye hospital located in Pakistan. The case report has been stated as per the guidelines stated in the CARE checklist (Supplemental File 1).[6]

The patient complained of a sudden loss of vision in the right eye that happened 2 months ago which had a visual acuity of 3/60 counting fingers. On inquiring, he also complained of gradual loss of vision in his left eye for 1 year which was 6/36 on Snellen's chart. There was no significant past medical history of systemic illnesses, infections, or inflammatory disorders. However, the patient experienced an episode of vision loss in one eye a year back which resolved on its own without any intervention. There was no history of any surgical procedures/interventions. Upon examination, the patient was well oriented to time person, and place.

Clinical findings

On slit lamp examination, in the right eye (OD), there was a white mobile hypopyon in the anterior chamber that on head movement moved with gravity. Grade II nuclear sclerosis mixed cataracts with grade IV vitritis and preretinal vitreous hemorrhage was observed in the right eye. On fundus examination with the 90-dioptre lens, a superior ghost vessel was seen as indicative of old branch retinal vein occlusion along with vascular sheathing and pigmentation.

In the left eye (OS) on slit lamp examination, grade I nuclear sclerosis mixed cataract, Grades I–II vitritis, and vascular sheathing on fundus examination were observed with three mirror lenses in the extreme periphery. Intraocular pressure was within normal limits in both eyes measured by the applanation tonometer.

On head-to-toe examination, facial symmetry was normal. On inspection of the mouth, oral ulcers were found (Figures 1 and 2). The patient had rashes on the forearm, and when hit with the scale produced the criss-cross pattern (dermatographia; Figure 3). The patient's genitals were examined after obtaining the patient's consent and a genital ulcer was found. The ulcer was close to the testicles onto the perineum region. The presence of the triad of Panuveitis and orogenital ulcers concluded examination to a provisional diagnosis of Bechet's disease that was further correlated with the investigation protocol of uveitis.

Clinical management

On the basis of a provisional diagnosis of active Panuveitis (intermediate and posterior) with orogenital ulcers, a provisional diagnosis of BD was made. Investigations were advised to correlate the clinical findings. B-scan ultrasonography and Optical Coherence Tomography were performed. Systemic investigations including complete blood count (CBC), erythrocyte sedimentation rate (ESR), C-reactive protein, human immunodeficiency virus, urea, creatinine, electrolytes, chest X-ray, and electrocardiogram (ECG) were done. The blood reports for CBC and ESR were deranged with high C-reactive protein results. All other reports were in normal ranges. All tests for uveitis including inflammatory and noninflammatory diseases were done. These tests included serum angiotensin-converting enzyme inhibitors, interferon-gamma release assay, mycobacterium tuberculosis (MTB), venereal disease research laboratory, fluorescent treponemal antibody absorption (FTS-ABS), immunoglobulin G (IGG), and immunoglobulin M (IGM) for toxoplasmosis, antinuclear antibodies (ANA), C-ANCA, and P-ANCA, anti-double-stranded DNA test, and HLA were done.

Following the uveitis protocol, the patient was investigated for underlying infection. Since the patient was noninfectious, the definitive diagnosis of BD was made considering an inflammatory, noninfectious multisystem pathology. The decision to administer a posterior subtenon (local injection) was made in both eyes of the patient accompanied with the aqueous suppressants to prevent the rise of intraocular pressure (IOP) and to prevent intraocular inflammation. The evidence suggests that topical steroids cause an IOP rise and systemic steroids most commonly cause cataracts. Although systemic steroids were not prescribed, and patient was referred to a medical physician for further workup and management of systemic inflammatory signs and symptoms.

Follow-up and outcomes

On a follow-up visit after 2 weeks, the patient's vision was improved in the right eye to 6/60 and left eye to 6/9 on Snellen's chart. The patient was on systemic steroids as prescribed by the medical physician who later referred the patient to a rheumatologist. The patient's oral ulcers were improved and hypopyon disappeared. On the 6-week follow-up, the patient developed cushingoid features due to systemic steroids, and IOP was on the higher side. The patient was advised to continue follow-up with his medical physician and rheumatologist to continue with the medical therapy.

On third monthly follow-up, he returned to his normal state of health; however, his vision was dropping in both eyes due to the progression of cataracts. Therefore, the cataract surgery was performed in both eyes, first the right eye and then the left eye. After cataract surgery, the right eye did not improve and developed serious inflammation causing a drop of vision from 6/60 to 1/60 for which was managed with posterior subtenon (Triamcinolone) and systemic steroids along with immunosuppressants as prescribed by his rheumatologist. The vision in the left eye improved to 6/9 followed by laser to the ischemic areas of the retina to prevent the development of serious complications such as neovascular glaucoma or Rubeosis Iridis. The patient was required to have a 6-month to yearly follow-up for 2–5 years with the eye department.

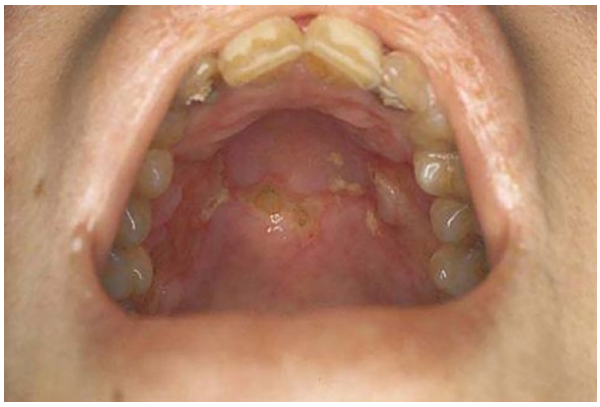

Figure 1. Behcet's disease patient with mouth ulcers.

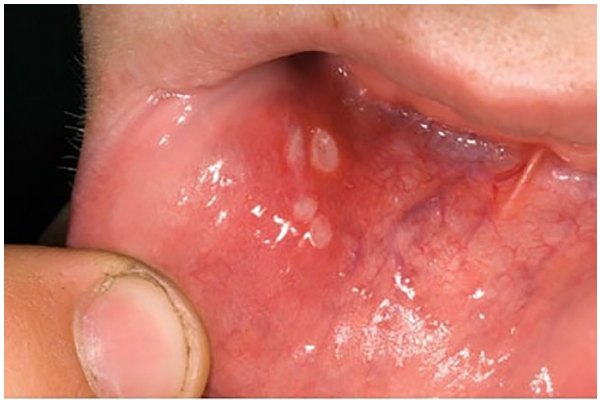

Figure 2. Mouth ulcers.

Figure 3. Dermatographia (cris-cross pattern).

Patient perspective

The patient was anxious when he first visited our clinic in the eye hospital. He was very much worried due to his sudden loss of vision. As the patient was young, he was concerned about his prognosis and prospects. However, the eye specialists provided him with detailed information about his disease process and counseled him and his family members (father and uncle). With the passage of time, as he was getting better and as his symptoms improved, he was satisfied with the treatment provided at the eye hospital. The patient was also regularly following specialty doctors as he was advised. However, he developed serious systemic side effects which were managed by the medical physician and rheumatologist with tapering off of steroids and continuation of immunosuppressants and biological agents in consideration with the systemic reports of main organs of the body such as liver and kidney status.

Discussion

Typically, BD affects younger patients and mostly develops before puberty.[7] However, our patient was presented in the early third decade. The prevalence of this ocular disease varies around the world.[4] In some ethnicities, inflammatory eye disease occurs in over 50% of patients.[4] Uveitis is the most common ocular manifestation as was found in our patient and can be present in up to 90% of cases.[8] Behçet's uveitis can be very difficult to manage and even if treated, up to 74% of patients lose vision in the decade following the onset of ocular symptoms.[9] However, in our patient, the right eye vision was retained and the left eye vision improved. One of the current challenges is ophthalmologists' understanding of rare diseases and ophthalmic manifestations worldwide.[10] There is a list of systemic multiorgan diseases that affect different layers of eyeball including Sjogren disease, rheumatoid arthritis (RA), and connective tissue disorders such as systemic lupus erythromatosis (SLE) which have histocompatibility complex (HLA) predilection like granulomatosis with polyangiitis (GPA) are usually associated with scleritis (inflammation of outer layer of eye).[11] Therefore, a multidisciplinary approach is required in each of these cases to manage the patient's desired outcomes. There is a scarcity of information and data related to the treatment of BD in Pakistan and only a few case reports are available[12–14]; therefore, this case report is a piece of evidence that future clinical trials and interventional studies are required to treat rare diseases with systemic involvement.

Conclusion

BD is most common in those whose roots are based in the Mediterranean Basin to the Far East.[4] This was the first ever BD case in our hospital after 20 years. Our hospital is a research and postgraduate educational institution that deals with complex eye diseases. The range of investigations and clinical exams helped clinical decision-makers in evaluating the patient's diagnosis.

Declarations

Ethics approval and consent to participate

Ethics approval is not applicable due to institutional/IRB policy for single case report. The patient involved in the case report gave written informed consent to participate in the study.

Consent for publication

The patient involved in the case report gave his written informed consent authorizing the use, disclosure, and publication of his health information.

Author contributions

Tabish Ali Shalwani: Conceptualization; Data curation; Investigation; Supervision; Validation; Visualization; Writing – original draft; Writing – review & editing.

Alishan Khowaja: Visualization; Writing – review & editing.

Narmeen Punjwani: Data curation; Visualization; Writing – review & editing.

Acknowledgements

We would like to acknowledge Professor PS Maher and Professor Dr Saleh Memon for providing us with a facility of tertiary eye care setup where we see patients with variety of rare diseases.

Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

Competing interests

The authors declare that there is no conflict of interest.

Availability of data and materials

No specific dataset was used in the current case report

ORCID iD

Tabish Ali Shalwani https://orcid.org/0009-0007-1306-0589

Supplemental material

Supplemental material for this article is available online.

References

1. Jin ZY, Lu B and Wang Y, eds. *Cardiac CT: diagnostic guide and cases*. Singapore: Springer Nature, 2020.
2. Ortiz-Fernández L and Sawalha AH. Genetics of Behcet's disease: functional genetic analysis and estimating disease heritability. *Front Med* 2021; 8: 625710.
3. Alibaz-Oner F and Direskeneli H. Advances in the treatment of Behcet's disease. *Curr Rheumatol Rep* 2021; 23(6): 47.
4. Davatchi F. Behcet's disease: global perspective. *Indian J Rheumatol* 2007; 2(2): 65–71.
5. Nair JR and Moots RJ. Behcet's disease. *Clin Med* 2017; 17(1): 71.
6. CARE checklist. https://www.equator-network.org/reporting-guidelines/care/ (accessed 11 December 2024).
7. Mendes D, Correia M, Barbedo M, et al. Behçet's disease–a contemporary review. *J Autoimmun* 2009;32(3–4): 178–188.
8. Zając H and Turno-Kręcicka A. Ocular manifestations of Behçet's disease: an update on diagnostic challenges and disease management. *J Clin Med* 2021; 10(21): 5174.
9. Tugal-Tutkun I, Onal S, Altan-Yaycioglu R, et al. Uveitis in Behçet disease: an analysis of 880 patients. *Am J Ophthalmol* 2004; 138(3): 373–380.
10. Pinazo-Durán MD, Zanón-Moreno V, García-Medina JJ, et al. Eclectic ocular comorbidities and systemic diseases with eye involvement: a review. *BioMed Res Int* 2016; 2016.
11. Uddin Z, Khan A, Haroon M, et al. Spectrum of ocular manifestations of rheumatic autoimmune diseases: a tertiary care experience in Pakistan. *J Ayub Med Coll Abbottabad* 2021; 33(4): 612.
12. Ishaq S, Saeed A and Jawad M. Regression of severe Behcet's eye disease with infliximab therapy; first two cases treated in Pakistan. *J Pak Med Assoc* 2012; 62(11): 1242–1244.
13. Jafri L, Nasir N and Almas A. Multifocal venous thrombosis in Behcet's disease. *J Coll Physicians Surg Pak* 2012; 22(11): 730–732.
14. Khan A, Haroon M, Bashir F, et al. Behcet's disease: Pakistani experience. *Pakistan J Med Sci* 2020; 36(5): 1005.