Case Report

Apremilast – A Novel Treatment for Behcet's Disease

Najia Ahmed[1], Kiran Naz Khan[2], Aima Omer[3], Noor us Sabah[4]

Abstract

Behcet's disease is a chronic inflammatory disease involving multi-organ system; it results from a complex interplay of genetic, environmental, infectious, and immunological factors, though its precise etiology remains unclear. The disease is categorized as neutrophilic dermatosis and the clinical manifestations occur due to vasculitis that affects blood vessels of all types and sizes. The disease affects many other organs specifically the eye, gastrointestinal system, central nervous system, joints, kidneys and vascular system. We report the case of an 18-year-old girl having Behcet's disease for the past 4 years with typical recurrent oral aphthosis along with pharyngeal involvement leading to dysphagia and a large ulcerative lesion over labia majora. Notably, her condition showed a remarkable response to a premilasthighlighting its potential therapeutic role in refractory mucocutaneous manifestations of Behcet's disease. This case underscores the expanding treatment landscape and the need for further investigation into targeted immunomodulatory therapies.

Keywords: Apremilast, Behcet's disease, Oral ulcers, Genital ulcers, Orogenital, Novel therapy.

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Authors Affiliation: [1,4]Departement of Dermatology, PNS Shifa Hospital, Karachi; [2]Dermatology, Al-Tibri Medical College, Isra University Karachi, Campus; [3]HITEC Institute of Medical Sciences, Taxila

Corresponding Author: Dr. Kiran Naz Khan, Assistant Professor of Dermatology, Al-Tibri Medical College, Karachi

Email: drkirankhan2010@hotmail.com

Introduction

Adamantiades – Behcet Disease (BD) also known as Behcet syndrome due to its multiorgan involvement. The disease was described by a Turkish dermatologist Madan Basnet et al, as a triad of recurrent oral, genital ulcers and uveitis.[1]

Recurrent oral ulceration (≥3 times per year) is the key diagnostic criterion. These ulcers often resemble recurrent aphthous stomatitis (RAS) but occur more frequently and with deeper, more persistent lesions. Genital ulcers, the second major clinical manifestation, predominantly affect females and labia majora is the most commonly involved site with tendency for scarring.[2]

Ocular involvement is more commonly observed in males which include uveitis (mostly posterior uveitis), conjunctivitis, hypopyon, and retinal involvement, leading to blindness in 25% of cases. Musculoskeletal system involvement is observed in 45–60% of patients. Gastrointestinal, neurological and peripheral vascular involvement are also observed with a male predominance.[3]

Papulopustular lesions or pseudo folliculitis, are the most common cutaneous manifestations, present in 50%–96% of patients, primarily affecting lower extremities.50% of patients may present with erythema nodosum.[4]

Treatment of BD is quite challenging with a chronic course of relapses and remissions. We present a distinct clinical case where apremilast, a phosphodiesterase-4 inhibitor, demonstrated a favorable clinical response. Few studies have been done internationally on this topic but not a single case has been reported from our population.

Case Report

An 18-year-old unmarried female presented to the dermatology outpatient department with a

four-year history of recurrent oral ulcers and a two-year history of genital ulcers. Initially she began experiencing severe pain and burning sensation in oral cavity, followed by multiple aphthous-like ulcers. The lesions progressively extended to the oropharynx, leading to dysphagia. Subsequently, she developed recurrent, painful genital ulcers, significantly affecting her quality of life.

The patient consulted different dermatologists and received multiple combinations of drugs including colchicine, deltacortil, methotrexate, folic acid, omeprazole and oral triamcinolone paste, but the response to treatment remained suboptimal. On examination, the patient exhibited mild pallor. She had an 8mm sized ulcer with an underlying erythematous base over the left border of the tongue (Figure 1a). There were three adjacent ulcers coalescing to form a large ulcer on the right labia majora extending to the labia minora. (Figure1b). Pathergy test was positive. Ocular and systemic examination was unremarkable. Her Hemoglobin was 9.8mg/dl, MCV=76 fl with low iron levels. Chest X-ray, sugar level, liver and renal function tests and urine reports were normal.

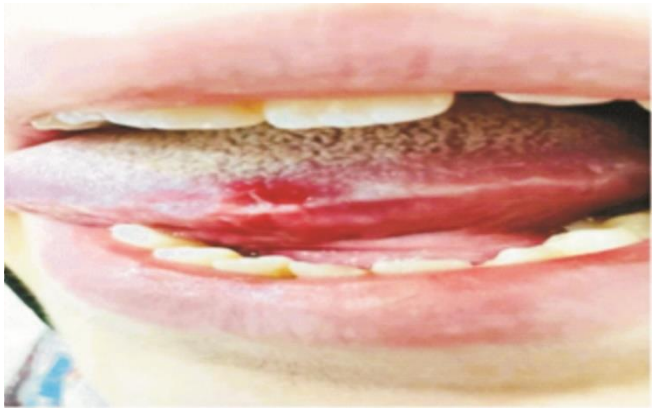

Figure (1a): *Shows a solitary erythematous ulcerative lesion over the left lateral border of tongue (Pre-treatment).*

Apremilast tablet was initiated, gradually building the dose to 30mg twice daily resulting in a significant clinical improvement. The ulcer healed in 3 weeks. However, the treatment continued for 12 weeks and the patient remained ulcer free. She was followed for a further three months without any recurrence of ulcers.

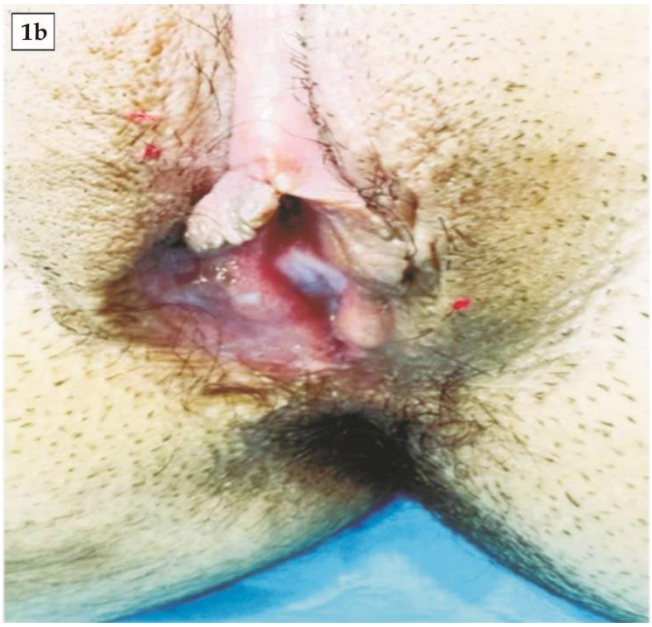

Figure (1b): *Single large healed ulcer over the right labia majora. (Post treatment).*

Discussion

BD, a rare systemic vasculitis with multiorgan involvement, usually occurs in the 3rd and 4th decade of life. Severe morbidities are more common in males.[5]However; our patient was a female in her teenage. Its exact etiology is unknown and is thought to be multifactorial with HLA-B*51 being the strongest genetic factor involved which is positive in about 60% of patients.[1] In our patient genetic testing was not performed due to a lack of resources.

Behcet's disease is diagnosed clinically using the International Criteria for Behçet's Disease (ICBD) which allocates 2 points each for oral ulcers, genital ulcers, and ocular lesions while 1 point is designated each for cutaneous, vascular, neurological involvement, and positive pathergy test. A total score of ≥4 supports the diagnosis of Behçet's disease.[6] The patient in our case report had recurrent oral aphthosis, and genital ulcers along with a positive pathergy test, hence, total score came out to be 5.

Carmona-Rocha E,[7]reported that Apremilast is a

novel treatment that inhibits phosphodiesterase-4 and increase the levels of cAMP, which restores the equilibrium between pro and antiinflammatory mediators like IL-10, IL-23, TNFα, IFNγ and produces good results in several chronic inflammatory diseases like psoriasis, systemic lupus erythematosus, atopic dermatitis, vasculitis, and uveitis. It produced excellent results in our patient.

Treatment with apremilast in BD was related to enhanced quality of life by Gulen Hatemi et al,[8] which was true for our patient. Zeyuan wang et al,[9] stated in his study that colchicine is an effective treatment option for BD and has been used widely but our patient was not satisfied with colchicine and got remarkable improvement with apremilast. Given the limited research on apremilast in BD, this case provides valuable clinical insight into its potential role in disease management.

Conclusion

This case report highlights apremilast as a novel treatment option for BD. Its role in other chronic inflammatory diseases is well established but its effectiveness in BD remains underreported. With this case report we set forth a path for further research on the effects of apremilast on Behcet's disease.

Author's Contribution

NA: Conceived, designed, edited the manuscript, given final approval of the version to be published, critical revisions.

KNK: Manuscript writing, final approval of the version to be published, agree to be accountable for all aspect of the work.

AO: Manuscript writing.

NUS: Manuscript writing.

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