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Behcet's disease: A study of two cases

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Abstract
Background: Behcet's disease (BD) is a rare, chronic, and recurrent systemic vasculitis characterized by recurring oral and genital ulcers, along with other auto-inflammatory symptoms. It primarily affects the small, medium, and large vessels, with a predilection for venous involvement, particularly in the pulmonary circulation. The disease is more common along the Silk Road, but its prevalence has increased globally. Early diagnosis and proper treatment are essential to prevent long-term morbidity and mortality. This paper reports two cases of BD from the Dermatology Hospital of Bamako, Mali.
Case Reports
Case 1: A 28-year-old Malian female presented with recurrent oral and genital ulcers for four years. Despite previous treatments, lesions persisted, leading to the diagnosis of BD. She was successfully treated with methotrexate and prednisone, resulting in significant improvement after 21 days.
Case 2: A 34-year-old female schoolteacher with recurrent genital and oral ulcers sought treatment after a year of flare-ups. The diagnosis of BD was confirmed based on clinical criteria, and the patient experienced improvement after medical management.
Discussion: BD is primarily diagnosed through clinical evaluation, with significant concerns regarding neurological and ocular involvement, which can result in permanent disability. Early intervention and multidisciplinary management, including corticosteroids and immunosuppressive agents, are key to controlling disease flares and preventing complications. Both cases highlight the challenges in patient education and the importance of timely diagnosis and treatment, particularly in resource-limited settings.
Conclusion: BD remains a challenging disease to diagnose and manage, requiring an integrated approach for optimal outcomes. Proper patient education, early diagnosis, and adherence to treatment protocols are crucial to improving prognosis and quality of life for affected individuals.

Keywords: Behcet's disease, vasculitis, oral ulcers, genital ulcers, systemic involvement

Introduction
Behcet's disease is a variable, recurrent, and remitting vasculitis characterized by recurring cutaneous and mucosal ulcers that can affect almost all organ systems in the body. Indeed, the presence of recurrent oral or genital ulcers accompanied by other auto-inflammatory symptoms should raise suspicion of this elusive disease. It is unique among vasculitides in that it can affect small, medium, and large vessels and tends to involve the venous circulation rather than the arterial circulation. Its effects on the pulmonary venous circulation are particularly notable for their role in mortality. Classically observed in Mediterranean, Middle Eastern, and East Asian countries and relatively rare in the United States, the disease's prevalence has increased, prompting the need for internists to be aware of the clinical presentation and treatment of Behcet's disease. The disease shows higher prevalence along the Silk Road, suggesting the role of environmental and genetic factors. Despite significant progress in understanding its clinical characteristics and therapeutic approaches, gaps remain in our understanding of its pathogenesis [1–3].
We report two cases of Behcet's disease at the Dermatology Hospital of Bamako in female patients aged 28 and 34, respectively.

Case report 1

This was a case of a 28-year-old Malian housewife with no significant medical or surgical history, who presented with painful oral ulcers that had been ongoing for 4 years, with the most recent recurrence in April 2024. Initially affecting both oral and genital areas, she sought treatment at a local clinic, where she was prescribed methylprednisolone and ceftriaxone, which provided temporary relief. Due to the persistence of lesions, the patient consulted a foreign center four times, where several tests were conducted, including acolonoscopy, autoimmune workup, hepatitis and HIV serologies, HSV PCR, TPHA, VDRL, and urine cultures (ECBU), all of which were unremarkable.

The diagnosis of Behcet's disease was considered based on clinical findings, and the patient was treated successively with colchicine, azathioprine, prednisone, and dapsone at six-month intervals.

A new flare-up three weeks prior led her to consult at the Dermatology Hospital of Bamako.

Upon examination, there were multiple ulcers of varying sizes and shapes, some confluent in areas, giving a "fresh butter" appearance on the inner sides of the lips and palate. The tongue was coated, and the skin-mucous junction of the lips had multiple ulcerative-crusty lesions (Figures 1, 2, and 3). There were no skin lesions resembling prurigo, and no ocular involvement. A number of tests, including a complete blood count (CBC) and hepatitis B and C serologies, were requested and came back normal. The patient was started on methotrexate at 20 mg per week and prednisone (Cortancyl) at 1 mg/kg of body weight. The condition showed favorable improvement after 21 days of treatment.

Case report 2

The case involved a 34-year-old Malian schoolteacher who had been experiencing symptoms for one month and sought consultation for recurrent genital and oral ulcers that had been flaring up for one year. The most recent recurrence was two months prior, starting with oral ulcers followed by genital ulcers four days later. The persistence of these flare-ups prompted her to seek care at the Dermatology Hospital of Bamako.

On examination, multiple ulcers were observed, the largest being roughly oval in shape, measuring 5 x 5 cm in diameter, with a flesh-colored, clean red base and well-defined borders, surrounded by an erythematous halo, located in the genital area. The palate showed signs of scarred lesions, and multiple prurigo-like lesions were present on the lower limbs (Figures 4 and 5).

The general workup was normal; tests for HIV, hepatitis, syphilis, Chlamydia PCR, and Donovan body examination were all negative. An ophthalmologic consultation revealed a decrease in visual acuity.

The diagnosis of Behcet's disease was considered based on clinical findings.

Argument

Behcet's disease is a serious vasculitis with a diagnosis that is primarily clinical. Neurological and ocular involvement are significant causes of long-term disability in this disease. Cyclophosphamide appears to be associated with infertility and an increased risk of malignancies in patients with Behcet's disease, prompting consideration of shorter treatment durations. Behcet's syndrome is a variable vascular vasculitis with heterogeneous clinical features. Skin, mucosal, and joint involvement may impair quality of life but typically does not cause permanent damage. However, untreated involvement of the eyes, blood vessels, nervous system, and gastrointestinal system can lead to severe damage or even death.

Management of Behcet's disease requires a multidisciplinary team to ensure faster, more accurate diagnoses and well-integrated treatment strategies. Corticosteroids are the cornerstone of treatment. Other agents used for induction and/or maintenance therapy include colchicine, azathioprine, cyclosporine-A, cyclophosphamide, IFN alpha, and tumor necrosis factor-alpha inhibitors.

In our first case, the therapeutic pathway taken by the patient was influenced by her misunderstanding of the disease's course (flare-ups and remissions). She believed that her condition would be cured after the initial treatment, which led her to repeatedly seek care abroad, resulting in significant financial costs and time loss, despite the availability of a simpler, less costly treatment, provided adherence to the prescribed therapy was maintained.

In the second case, the diagnosis of Behcet's disease was evident based on more than three criteria. However, what should be highlighted is the delay in seeking medical attention, which made the patient's daily life more difficult [4–6]. These two cases have provided us with insights into different aspects of Behcet's disease in our region.

Case 1

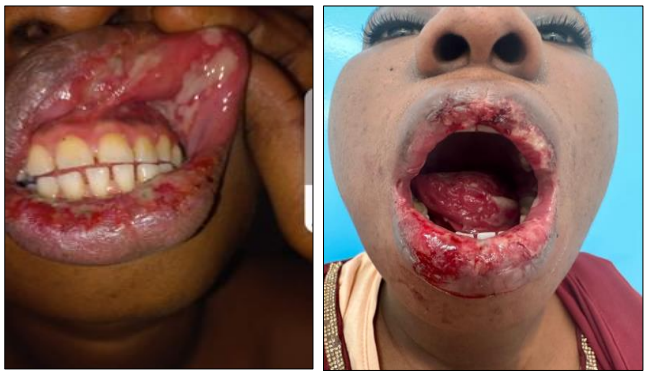

Fig 1: Oozing oral ulceration with a fresh butter appearance.

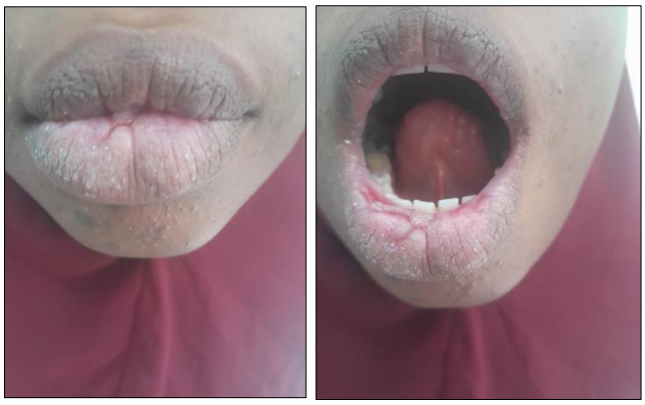

Fig 2: Progression after 3 weeks of treatment.

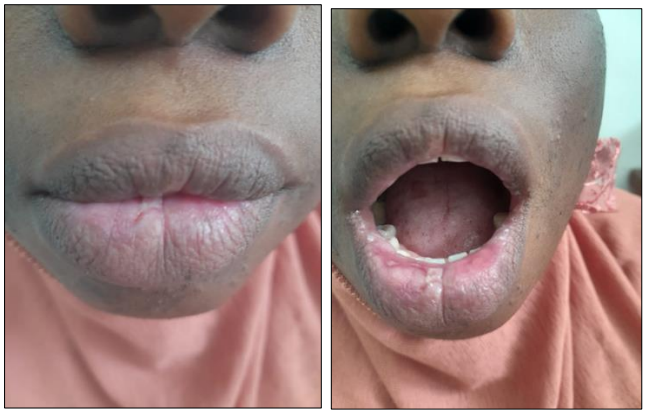

Fig 3: Progression after 5 weeks of treatment.

Case 2

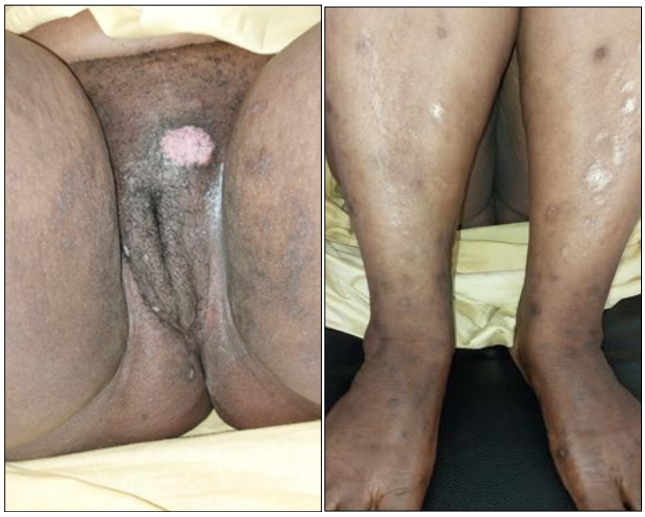

Fig 4: Ulcer in the process of healing on day 21. Face 5: Prurigo lesions.

Conclusion

Behcet's disease is a relapsing vasculitis that requires effective patient education for proper management. In the presence of any oral or genital ulceration, it is crucial to thoroughly examine the patient and establish an early diagnosis to avoid diagnostic delays, which can lead to delayed treatment.

Conflict of Interest: None

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