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CASE REPORT

Behcet's disease presenting with recurrent ocular, oral, and scrotal inflammatory lesions in a young Tanzanian man: a case report

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Key Clinical Message

Behcet's disease (BD) is a rare condition which is diagnosed clinically. Only few cases have been reported in sub-Saharan Africa. We report a Tanzanian male who presented with typical features of BD and was successfully treated. There should be an increase in awareness on BD to improve the management.

Keywords

Behcet's disease, inflammatory lesions, Tanzania.

Introduction

Behcet's disease (BD) is a rare immune-mediated systemic vascular disease (named after the Turkish Dermatologist, Hulusi Behcet in 1937) which is characterized by the recurrent oral aphthae and other several systemic manifestations [1]. It is believed to exist in many parts of the world with high incidences in the Middle East, Far East, and Mediterranean region and in an area of the ancient trading route known as "Old Silk Road" between latitudes 30° and 45° north in Asia and Europe [2, 3]. High prevalence of BD (420 per 100,000) has been reported in Turkey [1] with lowest prevalence of 0.38 per 100,000 being reported in the North America [2]. In sub-Saharan Africa, the prevalence of BD is not known but few cases have been reported elsewhere [4–6], with only one case being reported from Tanzania over 40 years ago [7].

Behcet's disease is believed to be caused by autoimmune response to an infectious or environmental insult in a genetically predisposed individual. HLA-B51 is the most strongly associated genetic risk factor. BD typically presents in the third and fourth decade of life with no specific sex predilection [1, 3, 8]. There are no pathognomonic investigative tests for the disease, and thus, the diagnosis relies on the clinical criteria according to the International Study Group for the Behcet's Disease (ISGBD) [9]. ISGBD requires the presence of recurrent oral aphthae (three times in 1 year) with at least two of the following: recurrent genital aphthae (aphthous ulceration or scarring), eye lesions (retinal vasculitis, cells in vitreous, or uveitis), skin lesions (papulo-pustular lesions, pseudo-vasculitis, acneiform nodules, or erythema nodosum), or a positive Pathergy test [9].

Corticosteroids, immunosuppressant drugs, tumor necrosis factor-inhibitors, and other symptomatic treatments are commonly used in the management of BD [10].

We report a case of young Tanzanian male who presented with typical symptoms and signs of BD and who was successfully treated with corticosteroids alone.

Case Report

We are reporting a 31-year-old black Tanzanian male who presented to our hospital with 2-year history of painful recurrent oral ulcers, 4-month history of scrotal lesions, and 1-week history of intermittent eyes discharge. The mouth ulcers started gradually in the buccal cavity, tongue, and lips. There have been periods of complete healing of approximately 1–2 weeks and recurrences. He reported to have four recurrences in the past 1 year which were neither bleeding nor discharging. In the past 4 months prior to the current admission, he noted the lesions around his scrotum that started like pustules and later ruptured to form painful ulcers. The penis was spared. He denied history of epigastric pain, painful defecation, painful micturition, hematuria, reduced amount of urine, or any history suggestive of sexual transmitted diseases in the past. One week prior to admission, the patient noted to have mild yellowish discharge from both eyes associated with itching and foreign body sensation but not pain. There was no blurred vision or photophobia. Over the course of his illness, the patient had neither fever nor weight loss. He reported to have had several prior admissions due to the same illness and had been treated with multiple courses of fluconazole, nystatin, and several antibiotics with no relief.

He had no known history of allergy and had never been transfused with blood or blood products. He is a fourth sibling in the family of six. All other family members were healthy and none of the other family member has similar illness. He was a manual laborer with no history of smoking or drinking alcohol.

Physical examination revealed young adult who was slightly wasted, full conscious, and afebrile (36.7°C). He had bilateral pus discharging from his eyes (Fig. 1). He also had extensive labial-oral ulcers with patches of thrush on the throat. There were neither Kaposi's sarcoma lesions nor lymphadenopathy. Ophthalmologist examination revealed bilateral conjunctivitis, normal visual acuity (6/6 in both eyes), normal visual fields, and normal optic nerves but with signs of uveitis by slit lamp examination.

The blood pressure was 120/60 mmHg, the pulse rate was 72 beats per minute regular, the respiratory rate was 18 cycles per minutes, and the oxygen saturation was 95% in room air. Urogenital system examination revealed normal male genitalia with scattered wet ulcerations on the scrotum (Fig. 1). The physical examination of the rest of the systems was essentially normal.

The results from laboratory analysis done were complete blood count (normocytic moderate anemia with hemoglobin level of 10 g/dL and a mean corpuscular volume 87 fl., other cells' count were within normal ranges), serum for syphilis antibodies by Venereal Disease Research Laboratory (nonreactive), HIV rapid serological test (negative), scrotal and ocular ulcers' swabs for Gram staining (no bacteria found), renal function test (normal range), random blood glucose (5.1 mmol/L), and Pathergy test (negative). Oesophago-gastroduedenoscopy was also performed and was normal. The diagnosis of BD was made according to the ISGBD [9] basing on the presence of recurrent oral aphthae (>3 times in 1 year) together with recurrent genital aphthae and posterior uveitis with conjunctivitis.

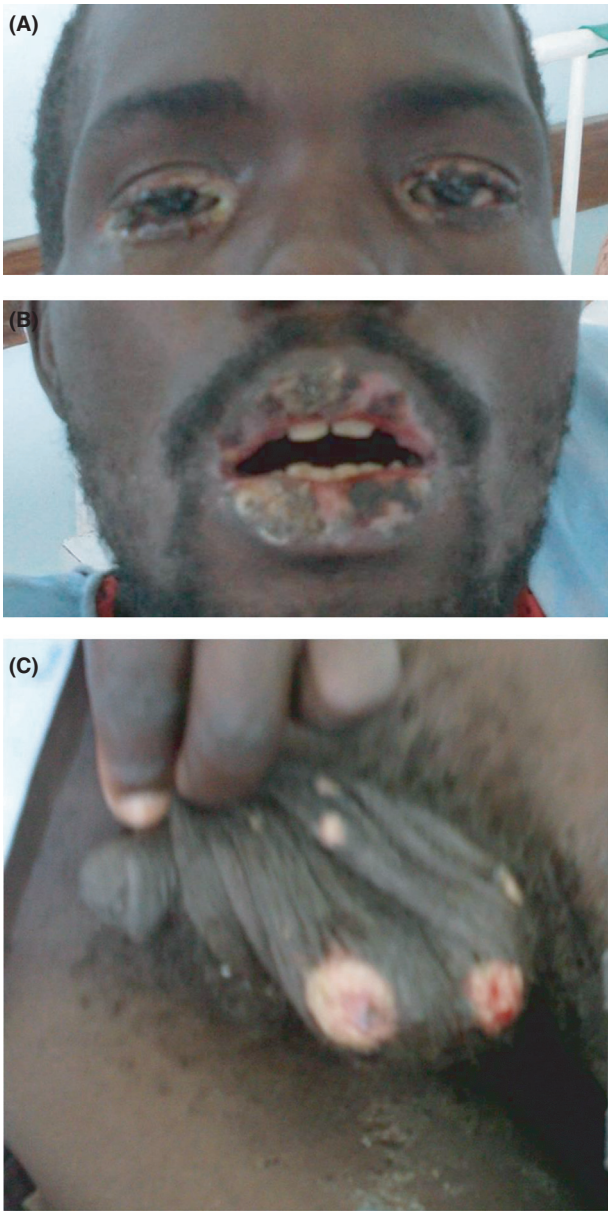

Figure 1. Characteristic ulcerative inflammatory lesions on the (A) eyelids, (B) mouth, and (C) scrotum in the index patient with Behcet's disease.

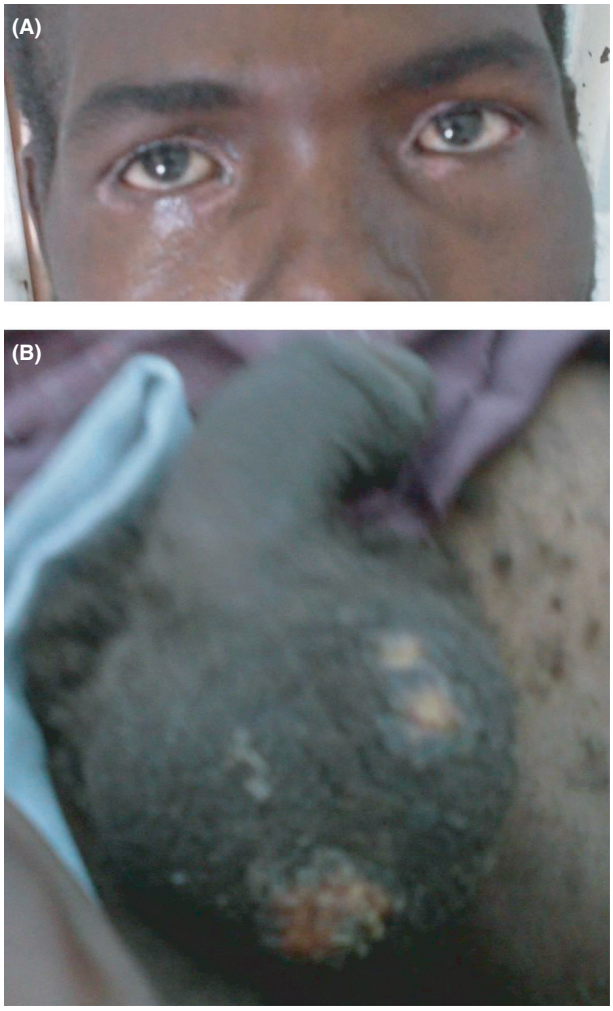

Figure 2. Pictures of the (A) eyelids and (B) scrotum showing good response to therapy 1 month after the initiation of corticosteroid therapy.

We treated this patient with intravenous dexamethasone 12 mg 8 hourly for 5 days followed by oral prednisolone 30 mg 12 hourly for 30 days then tapered gradually over 30 days. We also gave erythromycin eye ointment two drops 8 hourly for 7 days. Toward the end of the first week of corticosteroid therapy, the patient started improving significantly and the ulcers were completely healed after 18 days of medications. The patient was discharged from the hospital on day 21. He was followed up in the outpatient clinic on days 30, 60, and 90 as well as at sixth month and 1 year and without any recurrence of the ulcers or conjunctivitis (Fig. 2).

Discussion

The prevalence of BD in sub-Saharan Africa is unknown but only a few cases have been reported in Tanzania, South Africa, Ethiopia, and Nigeria [4, 7, 11, 12]. In light of these reports, we assume that many cases remain undiagnosed or unreported in this region due to lack of awareness regarding BD among clinicians in this region. The long duration of speculation prior to BD diagnosis as shown in the present case (2 years) and other similar case reports (5.5 years in Comoro and 2 years in Nigeria) [6, 12] further exemplify the delay in diagnosis that typically results from this lack of BD awareness. Our patient had typical presentation for BD according to the ISGBD criteria [9] (Fig. 1), but he was initially misdiagnosed at multiple health facilities. In particular, this patient was misdiagnosed as having candidiasis and bacterial infections.

In four case series of adults with BD of Africa ancestry, oro-genital aphthae were found to be the most common presenting feature [6, 11, 13, 14], as was also observed in our patient. Furthermore, the age and sex of our patient (31-year-old male) is similar to the patients' mean age at the time of diagnosis reported among one case series of 14 African adults living in the Union of the Comoros (33.1 ± 5.8 years) with male preponderance (71.4%, 10/14) [6]. Of note, in this series from Comoros, most BD patients were HLA-B51 negative leading the authors to conclude that BD associated with HLA-B51 negativity presents with severe manifestation with undue sequelae in East African adults as opposed to those with HLA-B51 positivity in other regions [1, 3].

Generally, BD responds well to steroids, with the combination of corticosteroids and immunosuppressant drugs being indicated when vital organs are involved [10]. Our patient responded very well to the steroids alone within 18 days of treatment with no vision sequelae as well as no systemic complications (Fig. 2).

Conclusion

This case emphasizes the need to increase awareness among clinicians in sub-Saharan Africa on BD so as to timely diagnose and offer prompt treatment. Further studies are needed to ascertain the prevalence and distribution of BD in Africa as well as associated genetic factors.

Acknowledgments

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Conflict of Interests

None declared.

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