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LETTER TO EDITOR

Behcets Disease in an Immunocompromised Female

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A 49 year old female presented to the outpatient department of dermatology withraw areas in oral cavity and genitals. She had multiple reddish elevated lesions with pain and burning sensation all over body. Patient had complaints of watering from eyes, headache, fever,weight loss, left knee joint pain and burning micturition. Past history of three similar episode was present. No history of blood transfusion, IV drug abuse, tattooing, any sexual transmitted diseaseor any significant family could be elicited. No associated comorbidities were elicited.

Cutaneous examination revealed few shallow (6-8) erosions with an erythematous halo over tongue and left side of soft palate. *Figure 1a* Few deep erosions present over labia majora, labia minora and vaginal mucosa. *Figure 1b* Multiple grouped vesicles, papules and pustules present over nape of neck, chest, bilateral axilla, abdomen, back and right leg. There were longitudinal ridges present over nails of right foot. Ophthalmological examination showed few erosions over left upper palpebral conjunctiva of eye. (Figure 1c)

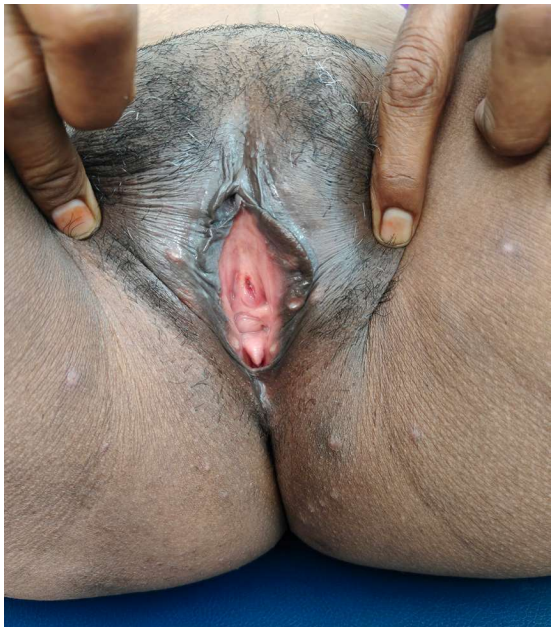

Figure 1a: Few shallow erosions with an erythematous halo over tip and lateral border of tongue

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Figure 1b: Few deep erosions present over labia majora, labia minora

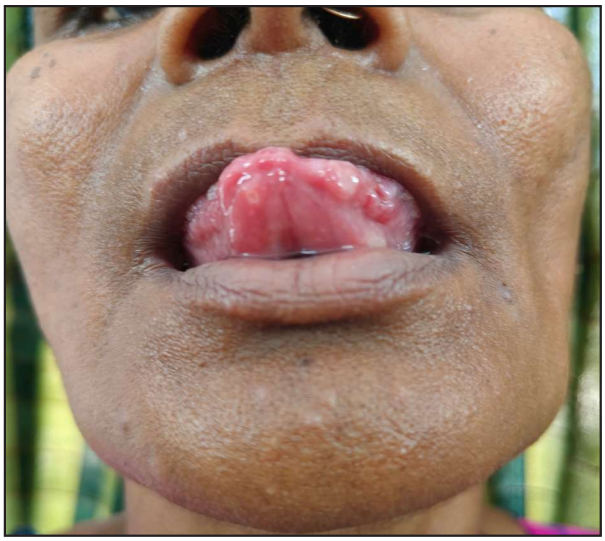

Figure 1c: Erosions present over left upper palpebral conjunctiva of eye

Punch biopsy taken from upper back with differential diagnosis of BD, DH, varicella, zoster, herpes simplex infection and sweet syndrome. Histopathological examination revealed focally ulcerated epidermis with an intense peri-vascular lympho-histiocytic infiltrate in the superficial dermis. There was distinct destruction of the hair follicles by an intense mixed inflammation of neutrophils, lymphocytes, histiocytes and plasma cells. Changes of leukocytoclastic vasculitis with destruction of vessel wall and extravasation of RBCs with lymphocytes, histiocytes, plasma cells and neutrophils were noted. Areas of necrotic debris were`also seen. *Figure 2* Patient was positive for human immunodeficiency virus 1,2 (HIV), Erythrocyte sedimentation rate (ESR) was raised (41mm/hr), total proteins (3.4 mg/dl), and albumin to globulin ratio (0.89) was decreased. Rest of the investigations includingC reactive protein, electrolytes, liver function test, histogram, differential count, urea, creatinine and G6PD were normal. As per the clinical features, blood reports and histopathological examination, patient was labelled as case of Bechet's diseasein HIV patient.

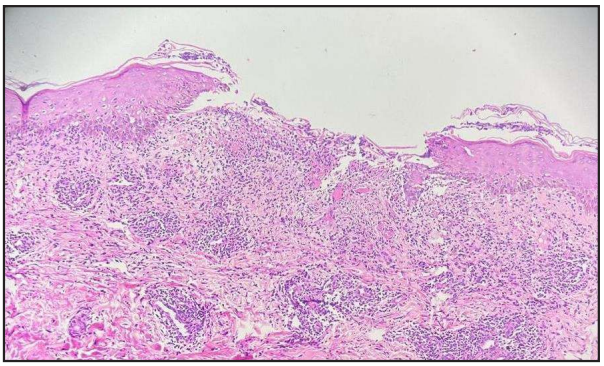

Figure 2: Histopathological examination revealed focally ulcerated epidermis with an intense peri-vascular lympho-histiocytic infiltrate in the superficial dermis. (H&E staining, 10x)

Patient was treated with oral steroids in tapering doses, azithromycin and colchicine 0.5 mg three times a day. For oral involvement benzocaine and chlorhexidine gel was prescribed. Eye lesions were treated with chloramphenicol plus dexamethasone and flurometholone eye drops. The skin and eye lesions started improving after 1 week of starting treatment. Patient was referred to ART (Anti- retroviral therapy) centre for further management.

Behcet's disease (BD) is a chronic, recurrent, multi-system inflammatory disorder of unknown aetiology.[1] It occurs primarily in young adults with the mean age of onset being 25-30 years. Most cases are sporadic although a familial aggregation has been reported. The disease has a worldwide distribution, common among populations with a higher prevalence of human leukocyte antigen (HLA) B5 and its split, HLA B51.[1] BD is rarely reported in HIV patients.[2-4]

BD is believed to be due to an autoimmune process triggered by an infectious or environmental agent in genetically predisposed individuals. T cell homeostasis perturbation, especially Th1 and Th17 expansion and decreased regulation by T-regs is supposed to be the cornerstone of disease pathogenesis.[1]

Presence of oral aphthous ulcerations plus two of the following manifestations recurrent genital ulcerations, cutaneous manifestations (erythema nodosum-like lesions, pseudofolliculitis, papulopustular lesions, or acneiform nodules), ocular involvement, and positive pathergy test is required for the diagnosisbased on international Study Group Criteria[5]

Occurrence of BD in patients with HIV infection may pose a diagnostic challenge. Kharkar V. *et al* has reported a case of BD with false positive VDRL.[6] Occurrence of both BD and HIV infection may be coincidental, a Behcet's-like presentation of the complications of HIV disease, or HIV infection causing or predisposing to a Behcet's-like illness.

First case of BD in an HIV infected individual was reported by Buskila in 1991. The pathophysiology of Behcet's disease in HIV patients is still unclear; but it is proposed to be due to disturbances in the immune system caused by virus resulting in susceptibility to autoimmune disease and vasculitis.[1]

Cicalini *et al.*[7] have postulated that induction of BD in HIV-associated cases might be a direct effect of viral replication or through HIV-induction of autoimmune mechanisms. A case of BD has been reported after acute HIV infection when the CD4 count was not low, suggesting that, if there is a true relationship, HIV replication itself could be the relevant HIV parameter causing immune dysregulation.

The cornerstone of therapy is corticosteroids whereas immunomodulatory and immunosuppressive agents such as colchicine, azathioprine, cyclosporine-A, interferon-alpha, and cyclophosphamide are used as steroid-sparing agents and to prevent further relapses.

Support: NIL

Conflicts of interest: NIL

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