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Behcet's Disease: A Rare Case Report

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1. Introduction

Behcet's disease or Behcet's syndrome is a rare form of vasculitis that can cause symptoms in the oral cavity, skin, eyes, genitals and may also present with other forms of systemic manifestations. This condition is characterised by recurrent aphthous ulcers in the oral mucosa, acneform eruptions on the skin, anterior or posterior uveitis and genital ulcers [1].

It was initially described by Hulusi Behcet in 1973. Since it was initially observed in the middle east, it is commonly known as the "silk route disease" [2]. A search for Behcet's disease on the internet from January 1946 to December 2020 showed, for example 2,115 cases from Turkey 1,183 cases from Japan, 486 cases in China and 137 cases from India have been reported.

It is said to have its highest incidence in Turkey (silk route) where it is around 20 to 602/100,000 in and 0.33/100,000 prevalence in the United States where its incidence is very low [3].

2. Case Report

A 45-year-old female presented to our OPD with complaints of acneform eruptions on her thighs for over 1 year, not decreasing with treatment. Upon further questioning, she also revealed that she had an erosion in her lower lip which kept recurring for a duration of around 1 and a half years.

Physical examination revealed an aphthous ulcer on the mucosal surface of the lower lip as shown in FIG. 1, multiple pustules, and papules on both the thighs shown in FIG. 2 & 3, and genital examination revealed a mucosal erosion at the 12'O clock position near the clitoris shown in FIG. 4, as well as perianal erosions shown in FIG. 5.

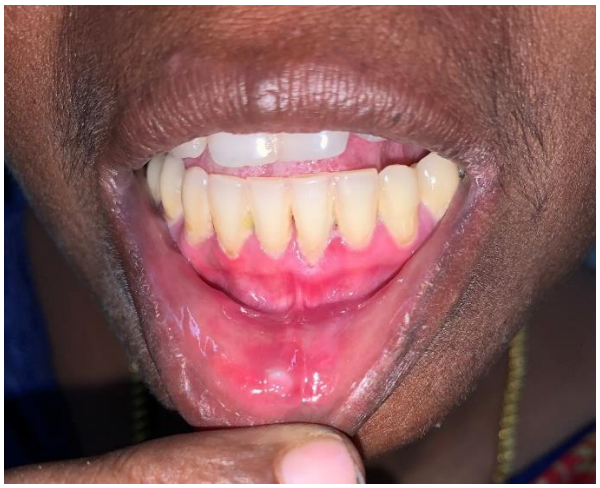

FIG. 1. **Here is a picture depicting an aphthous ulcer on the mucosal surface of the lower lip.**

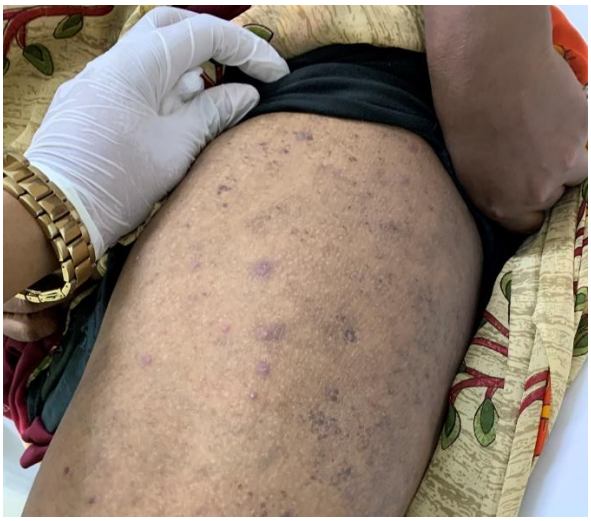

FIG. 2. **Here is a picture depicting acneform eruptions on the left thigh of the same patient.**

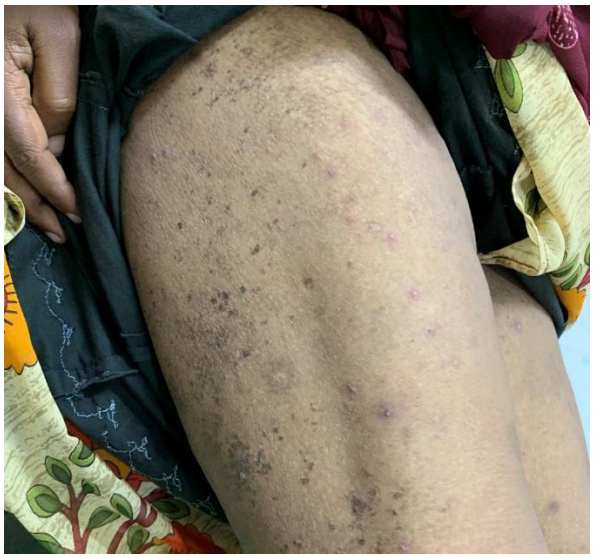

FIG. 3. **Pictured above are a few acneform eruptions on the right thigh of the patient.**

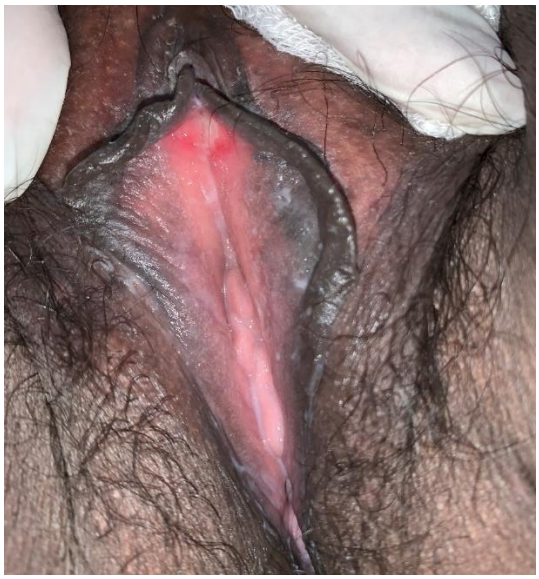

FIG. 4. **Pictured above is a vulval erosion at the 12 O'clock position in our patient.**

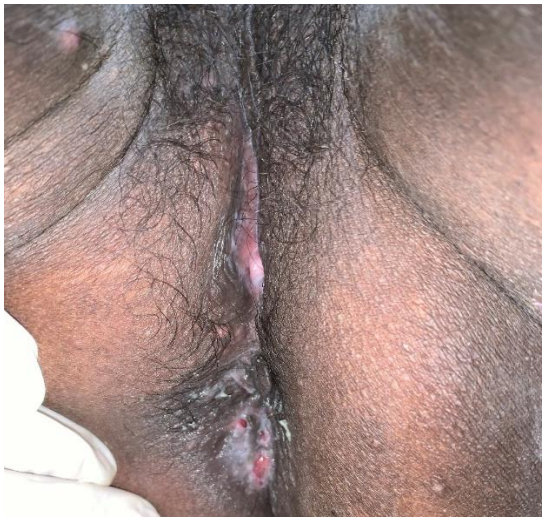

FIG. 5. **Here is a picture showing the perianal erosions.**

3. Investigations

A provisional diagnosis of Behcet's was made and thus the patient was taken for an ophthalmological check-up which showed no signs of anterior uveitis.

Pathergy test was done which was found to be positive.

A biopsy was taken from a pustule from the right thigh which showed -

3.1 Histopathologic Picture

Showed a dense perivascular lymphocytic infiltrate as shown in FIG. 6. Epidermis was absent.

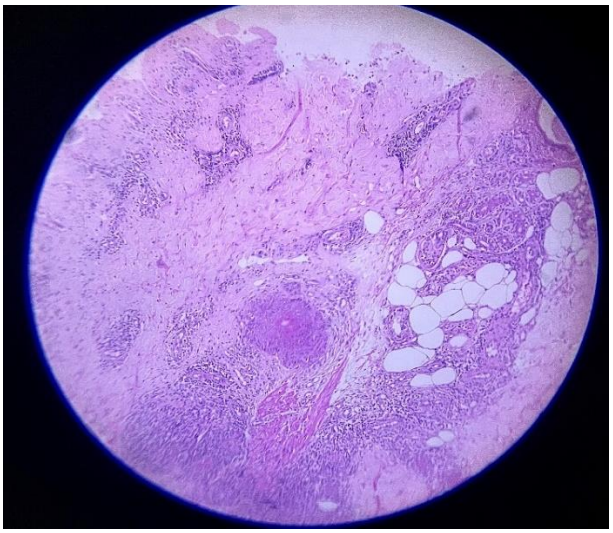

FIG. 6. **Histopathological image.**

3.2 Treatment history

Initially she was given Triamcinalone oral paste twice a day for the aphthous ulcer. She was also given Mupirocin ointment for the skin lesions which showed no improvement and was then given oral Clindamycin at a dose of 300 mg three times a day for five days. Despite this treatment the skin lesions persisted. A Diagnosis of Behcet's disease was made and thus she has been started on a short course of systemic corticosteroid therapy - Tab. Methylprednisolone 4mg twice daily and was given topical therapy for her skin and mucocutaneous lesions. She is being reviewed under regular follow up.

4. Discussion

In Behcet's disease there is inflammation of the blood vessels ranging from the small arteries to the big ones, sometimes including inflammation of the veins as well [2], however there is no necrosis or giant cell formation associated with this type of vasculitis [4]. It is said to be associated with the HLAB51 gene, but not every person carrying this gene is necessarily affected. There have also been certain environmental and infectious triggers that have been reported to have caused manifestation of the disease in certain individuals through an exaggerated autoinflammatory response in genetically predisposed individuals. The disease course tends to be chronic and relapsing in most cases [1].

The clinical criteria to diagnose Behcet's is known as the ICBD (International Criteria for Behcet's disease) criteria and includes the following [5]-

Occular lesions	2 points
Genital Aphthous ulcer	2 points
Oral aphthous ulcer	1 point
Skin lesions	1 point
Neurological manifestations	1 point
Vascular manifestations	1 point

Positive pathergy test - 1 (Pathergy test is optional criteria, 1 extra point may be added in case a pathergy test is conducted and reported positive).

A minimum score of 4 indicates Behcet’s disease.

Our patient had a total score of 5, thus indicating in favour of Behcet’s disease.

5. Conclusion

Thus, based on the clinical findings and after histological correlation, a diagnosis of Bechets disease was made. As physicians we must not exclude the thought of Behcet’s disease when we see patients with recurrent, non-healing aphthosis, Uveitis and Genital ulceration although the disease itself is a rarity.

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