

A Case Report On Behcet's Disease

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Abstract

Behcet's disease, also known as Silk Road disease, is a rare and chronic inflammatory condition that can lead to vasculitis. Diagnosis of this condition is mainly based on signs and symptoms as there are no definitive tests. It is characterized by a triple-symptom complex of intermittent oral aphthous ulcers, genital ulcers, and uveitis, with inconsistent systemic manifestations. This case report describes a 16-year-old boy presenting with oral ulcers and skin lesions on the genitalia that eventually turned into ulcers. The patient also complained of intermittent abdominal pain, which was allegedly associated with the lesions. As there is no specific treatment for this disease, the patient was given treatment according to the symptoms, severity, and location of the manifestation.

Keywords: Behcet's disease, Silk Road disease, Oral ulcers, Genital ulcers, Vasculitis

Abbreviation: OPD: Out Patient Department

ENT: Ear, Nose, Throat

FA: Folic acid

MVBC: Multivitamin B complex

BD: Bis in die

IV: Intravenous

PO: Per Oral

LA: Local application

Anti-TNF: Anti Tumor necrosis factor

NSAID: Non-steroidal anti-inflammatory drug

WBC: White blood cell

INTRODUCTION:

Behcet's disease, also known as Behcet's syndrome, Morbus Behcet's, Silk Road disease, or Adamantiades syndrome, is an infrequent and poorly acknowledged condition that results in inflammation of the blood vessels and tissues. It is subdivided into neuro-Behcet, ocular-Behcet, and vasculo-Behcet¹. Although the etiology of Behcet's disease is not fully understood, genetic, environmental, and immunologic factors are thought to play a role². The initial symptom of Behcet's syndrome is generally round or oval aching canker sores with erythematous borders on the mucous membranes that line the mouth (aphthous stomatitis)³. They may precede other symptoms of Behcet's syndrome by a number of years. Sometimes similar sores might appear on the genitals, particularly the scrotum and shaft of the penis, and the vulva in females, which also recurs but, unlike oral sores, might tend to scar⁴. It also affects the eyes, which may include posterior uveitis, anterior uveitis, or iridocyclitis, lacrimation, and hypopyon iritis⁵. It can also be characterized by neurological and gastrointestinal lesions. The exact cause of Behcet's syndrome is not known, but some studies suggest that some people may have a genetic predisposition to the condition⁶. Here, we present a case of Behcet's disease with a classical clinical presentation.

Case Report:

A 16-year-old boy presented to the OPD with complaints of skin lesions over the genitalia for 3 months. Additionally, oral ulcers were seen on the patient's inner aspect of both lips and tongue for 7 months, which were relieved by medication. Initially, there were fewer in number, but gradually increased in size and number. The patient was asymptomatic 5 months ago after which he developed a single white-colored skin lesion on the left side of the scrotum. The skin lesions were not associated with itching but with burning pain and mild intermittent fever which was relieved by medication. The lesions would burst and form raw areas, which eventually turned into ulcers. Both of the lesions never regressed completely but were relieved by medication to some extent. Allegedly, it was informed that swelling of feet and intermittent abdominal pain were associated with the skin lesions.

It was also documented that the patient had unprotected sex 1 year ago. No similar family history was found in the record. Cutaneous examination of the external genitalia revealed the presence of multiple well-defined rounds to oval, tender ulcers with a pale base and yellowish discharge at some areas and crusting at others on the scrotum and undersurface of the penis, along with scarring and oozing present on the penile shaft. The mucosa of both lips and tongue showed multiple superficial ulcers with an ill-defined erythematous rim on the inner aspect.

He had visited the ENT on 6/12/21 and continued the medication for 10 days. His treatment history plan consisted of Cap Doxycycline-100mg, Tab. MVBC, Tab FA, Tab Vit C, Tab PCM for BD, Soframycin cream, and TESS gel, which is a topical corticosteroid.

Laboratory tests showed a fall in the count of lymphocytes (18%) and a high WBC count (16700/cmm).

Treatment plan:

Drug Name	Dose	Route	Frequency
Inj. Dexamethasone	1cc	IV	OD
Inj. Augmentin	1.2gm	IV	12hrly
Inj. Metronidazole	1pint	IV	8hrly
Inj. Infupar	1pint (only if temperature is >101 °F)	IV	
Tab. Colchiline	0.5mg	PO	1-1-1
Tab. Brufen	400mg	PO	1-0-1
Tab. MVBC/FE/FA	101	PO	1-0-1
Tab. Famotidine	40mg	PO	1-0-1
Glycerine Solution		LA	BD
Fucibet cream (scrotum ulcer)		LA	BD
Vaseline Gauze (scrotum ulcer)		LA	BD
Ozenoxacin Cream (oral ulcer)		LA	BD

DISCUSSION:

Behcet's disease is a rare and not well-recognized condition that leads to inflammation in blood vessels and tissues. The exact cause is unknown, although it is believed to have a genetic component. The disease is characterized by painful sores in the genital area and mouth, along with other possible symptoms such as eye, neurological, and gastrointestinal problems. The patient described above had multiple genital skin lesions that were painful, with some having yellowish discharge and crusting. Additionally, the patient had experienced several oral ulcers in the past. Laboratory tests indicated inflammation, with low lymphocyte counts and high white blood cell counts.

While there is no cure for Behcet's disease, treatment is aimed at managing symptoms. This may involve using medications such as corticosteroids, colchicine with apremilast, immunosuppressants, and biologics such as anti-TNF drugs for severe cases. Antibiotics may also be used to prevent infections that can occur due to these medications. In the case of the patient described above, treatment included oral colchicine for mouth and genital sores, IV corticosteroids, and oral NSAIDs to reduce inflammation. The patient was also given a topical corticosteroid cream (Fucibet) with Vaseline gauze to apply locally twice a day for genital ulcers and an antibiotic cream (Ozenoxacin) to apply locally twice a day for oral ulcers. The patient responded well to the treatment with no reported systemic complications.

CONCLUSION:

Behcet's disease is a chronic and heterogeneous condition, which is intriguing in nature ⁷. We present this case because it is a rare multisystem inflammatory disorder that is characterized by ulcers. It is uncommon among Indians, and since the diagnosis is primarily based on clinical judgement, primary care physicians should be familiar with the disease's characteristics. Treatment of Behcet's disease is typically based on the symptoms, severity, and location of its manifestations. The patient in this case received supportive and symptomatic therapy. In Behcet's disease, patients can experience genital and oral ulcers for months without a definitive diagnosis, and only when severe complications arise do they consult their physician, as was the case with the patient described above. Therefore, any delay in the diagnosis increases the morbidity and mortality rates in patients with Behcet's disease.

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