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Behcet's Disease: Difficulties In Diagnosis And Patient Management (Clinical Example)

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Annotation
The article describes a clinical case of Behçet's disease, a rare systemic vasculitis characterized by recurrent orogenital ulcers, ocular and skin manifestations. Detailed anamnesis, clinical examination, laboratory tests and treatment are presented.
Keywords: Behçet's disease, pathogenesis, clinic, treatment.

Introduction

Behçet's Disease (BD) ***(Synonyms: Adamantiades–Behçet disease, Silk Road disease)*** is a chronic, relapsing, multisystem disorder of unknown aetiology characterised by systemic vasculitis involving arteries and veins of various calibres [1–4]. It is the only form of systemic vasculitis capable of leading to secondary amyloidosis. The disease has a well-established immunogenetic basis, with strong associations identified between BD and the HLA-B5, B12, and B51 antigens [2,5].

The major clinical features of Behçet's disease include four main types of lesions:

- **Oral lesions**: deep, painful aphthous stomatitis, gingivitis, glossitis, or pharyngitis.
- **Ocular manifestations**: hypopyon, chorioretinitis, iridocyclitis, and frequently panuveitis with progressive visual loss.
- **Genital ulcerations**: necrotic ulcers followed by coarse scarring.
- **Cutaneous manifestations**: erythema nodosum, ulcerative lesions, and pyoderma.

Minor clinical signs include:

- **Articular involvement** – asymmetric mono- or oligoarthritis of medium-sized joints without destructive changes;
- **Erosive and ulcerative lesions** of the gastrointestinal tract throughout its length;
- **Thrombophlebitis** of large veins (superior and inferior vena cava);
- **Severe central nervous system involvement** – meningoencephalitis, polyneuropathy, or dementia.

Behçet's disease has a unique geographic and genetic distribution. It is most commonly diagnosed in regions once traversed by the ancient Silk Road — including the Mediterranean countries, Central and East Asia [1,6]. Hulusi Behçet originally postulated a viral aetiology for the disease. The presence of antibodies in the serum and oral mucosa supports an autoimmune mechanism in the pathogenesis of the disorder. Furthermore, its frequent association with other autoimmune diseases strengthens the theory of autoimmune pathogenesis.

Pathogenesis

The central pathological process is a systemic vasculitis of immune complex origin. Key pathogenic mechanisms include:

- decreased activity of T-helper lymphocytes and increased levels of circulating autoantibodies directed against mucosal epithelial cells;
- the presence of circulating cytotoxic T-lymphocytes reactive to oral mucosal antigens;
- a reduced number of interleukin-2 receptors on T-lymphocytes;
- decreased levels of secretory IgA in saliva;
- increased chemotactic and phagocytic activity of segmented neutrophils.

These immune abnormalities contribute to endothelial injury and vascular inflammation in both arterial and venous vessels [1,6,7].

Clinical Manifestations

Behçet's disease affects both sexes, though it is more common in men, typically between 20 and 40 years of age. The disease involves multiple organs and systems, with the most frequent manifestations being oro-

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genital ulcerations. The approximate frequency of the main symptoms is as follows: aphthous stomatitis (90–100% of patients), genital ulcers (80–90%), and ocular involvement (60–85%).

Oral lesions are characterised by the appearance of aphthous, thrush-like erosions and ulcers of varying shapes (Fig. 1).

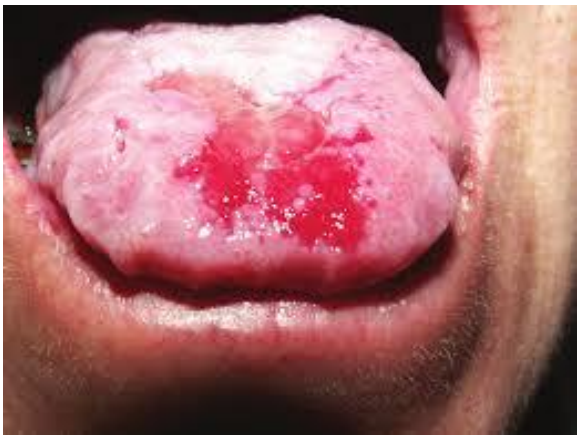

Fig. 1. Aphthous stomatitis of the oral mucosa and tongue.

Lesions develop on the tongue, soft and hard palate, palatine arches, tonsils, cheeks, gums, and lips, accompanied by severe pain. The eruptions usually begin as small, painful indurations of the mucosa, which subsequently evolve into superficial ulcers covered with a fibrinous coating and later into crater-like ulcers surrounded by a narrow hyperaemic border. Ulcers may enlarge up to 2–3 cm in diameter.

Occasionally, the process starts as a superficial aphtha that deepens over 5–10 days as infiltration develops at its base. After healing, soft, superficial, smooth scars remain. Usually, 3–5 lesions coexist simultaneously. Aphthous ulcerations may also develop on the nasal mucosa, larynx, oesophagus, and throughout the gastrointestinal tract.

Genital lesions consist of small vesicles, superficial erosions, and ulcers, sometimes with undermined edges. In men, ulcers are typically located on the scrotum, at the base of the penis, and on the inner thighs. They are irregular in shape, 2–4 cm in diameter, with uneven, seropurulent bases and marked tenderness.

In women, painful ulcers ranging in size from a pea to a five-kopeck coin are commonly observed on the labia majora and minora. In some cases, skin lesions on the trunk and extremities occur, including erythema nodosum, multiform exudative erythema, nodules, pustules, acneiform and haemorrhagic eruptions.

The disease is often accompanied by severe systemic symptoms — fever, intense headache, general weakness, cranial nerve palsies, and joint involvement. Periods of remission and exacerbation alternate; spontaneous remissions lasting weeks or months may occur.

The prognosis is often unfavourable. Ocular involvement may lead to blindness, and prolonged disease frequently results in disability. Prognosis is especially poor in cases with central nervous system involvement.

Histopathology

Histological examination of the ulcer margins on the mucous membrane reveals an inflammatory infiltrate composed of lymphocytes, plasma cells, histiocytes, and neutrophils, along with numerous vessels exhibiting oedematous, loosened endothelium.

In biopsy specimens from genital skin lesions, marked dermal oedema is noted, with numerous congested vessels surrounded by extravasated erythrocytes. The cellular infiltrate mainly consists of lymphocytes, polymorphonuclear leukocytes, and histiocytes.

Differential Diagnosis

Differential diagnosis should include the following conditions: pemphigus vulgaris, Reiter's oculo-genito-urethral syndrome, bullous erythema multiforme, recurrent aphthous stomatitis, acute ulcer of Chapin–Lipschütz, aphthous–ulcerative pharyngitis, and recurrent deep scarring aphthae.

Case Report

Patient R., born in 1962, presented to the **Republican Dermatovenereological Hospital** on **04 November 2020**, residing in the **Kushtipa District of Fergana Region**, with complaints of ulcerative eruptions on the oral mucosa.

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Anamnesis morbi: The patient has considered herself ill for approximately 1.5 years. The disease began with the appearance of a painful, localised induration on the oral mucosa, followed by ulcer formation with a hyperaemic surface. The ulcer gradually enlarged up to 2 cm in diameter. She initially consulted a local dermatologist, who diagnosed ***oral candidiasis***. The patient associated the onset of symptoms with recurrent upper respiratory infections.
She received repeated treatment from dentists, dermatologists, and surgeons with only minor improvement. She also practised self-treatment, applying various creams and tablets, the names of which she could not recall. She was under continuous follow-up at the local dermatovenereologic dispensary.
Anamnesis vitae: Born the second child in a peasant family; parents were not consanguineous. Grew up in satisfactory living conditions. Currently, the patient is a **Group II disabled person**, married, with three children. She has had **stable menopause for 2 years**. Childhood diseases: acute respiratory infections, chickenpox.
Family history: not burdened. No harmful habits. Denies allergic reactions to medications.
Status praesens: General condition satisfactory. Conscious, active, normal body build, normosthenic constitution, moderate subcutaneous fat. Musculoskeletal system: no deformities. Peripheral lymph nodes slightly enlarged and mildly tender on palpation. Breathing even and nasal. Lungs: vesicular breath sounds. Heart: tones clear, rhythmic. Pulse 90/min, BP 140/90 mmHg. Tongue moist and clean. Abdomen soft, non-tender. Liver and spleen within normal limits. Bowel movement regular. Renal percussion tenderness negative bilaterally. Normal urination. Sleep and appetite disturbed. Emotional state unstable.
Status localis: The pathological process is chronic, inflammatory, localised, and asymmetric, affecting the oral mucosa. Symmetrically located ulcers are observed: on the right — pea-sized; on the left — bean-sized. The ulcers are infiltrated, with distinct borders and irregular outlines. The base of the ulcers is uneven; a rim of hyperaemia surrounds the lesions. On palpation: ulcers are soft and sharply painful. Sensitivity preserved in all lesions. Hair and nails not affected. Subjective complaints: severe pain and burning sensation.
Based on anamnesis and clinical findings, the diagnosis of Behçet's disease was established.
Laboratory Investigations:

Test	Findings
Complete blood count	Hb 110 g/L; RBC 4.5×10^{12}/L; WBC 9.7×10^{9}/L; eosinophils 7%; lymphocytes 32%; monocytes 7%; ESR 18 mm/h
Biochemical blood analysis	Total protein 72 g/L; bilirubin 11.6 μmol/L; glucose 5.4 mmol/L
Urinalysis	Protein 0.033 g/L; no abnormalities
Vaginal/cervical/urethral smears	Leukocytes: Vag 18–20, Cervix 25–30, Urethra 6–9; epithelial cells normal; no gonorrhoea or trichomonads detected
Mycology	Yeast-like fungi isolated from the oral mucosa

Treatment:

- *Sol. Diflucani* 50 ml IV, No. 5
- *Sol. Vitreous body* 2.0 ml IM, No. 10
- *Sol. Actovegini* 5.0 ml IM, once daily, No. 10
- *Tab. Diflucan* 200 mg, No. 4
- *Tab. Prednisolone* 5 mg (40 mg/day)
- *Tab. Asparkam* 1 tablet 3 times daily, No. 10
- *Topical therapy:* gargling with antiseptic solutions; crushed fluconazole tablets mixed with butter applied twice daily for 10 days

Treatment outcome: By day 4, epithelialisation of the lesions was noted; by day 10, residual hyperaemic halos remained.
Comment:
This case represents a rare occurrence of Behçet's disease and a diagnostic error by dentists, who are often the first specialists to encounter such patients. Given the potential complications of the disease, early diagnosis

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and treatment are essential, especially considering the increasing incidence among women of reproductive age.

Discussion

Behçet's disease remains a diagnostic challenge for clinicians of various specialities due to its multisystemic nature and variable presentation. The disease can mimic several other disorders, such as aphthous stomatitis, erythema multiforme, or autoimmune mucocutaneous conditions, which often leads to misdiagnosis and delayed treatment.

In the present case, the patient initially received prolonged local therapy for presumed oral candidiasis and stomatitis, with minimal clinical improvement. Only after persistent ulcerations and characteristic symmetrical oral lesions were observed was Behçet's disease correctly diagnosed.

The immunopathogenesis of BD is thought to involve aberrant activation of both innate and adaptive immune responses. Enhanced neutrophil activation and oxidative burst, combined with endothelial dysfunction, result in immune-complex–mediated vasculitis affecting both arterial and venous systems. T-lymphocyte dysregulation, autoantibody production, and a decrease in interleukin-2 receptor expression further contribute to chronic inflammation and tissue injury.

The case presented demonstrates the importance of multidisciplinary evaluation, involving dermatologists, ophthalmologists, rheumatologists, and neurologists. Early recognition of systemic features and institution of immunosuppressive therapy—particularly corticosteroids—can prevent serious complications, including blindness and neurological deficits.

Conclusion

Behçet's disease requires vigilance from physicians across multiple disciplines. Early recognition of systemic manifestations and prompt initiation of combined therapy help to stabilise the disease course and improve prognosis.

Multidisciplinary management remains the key factor in achieving optimal outcomes for patients with Behçet's disease.

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