BAHCET DISEASE

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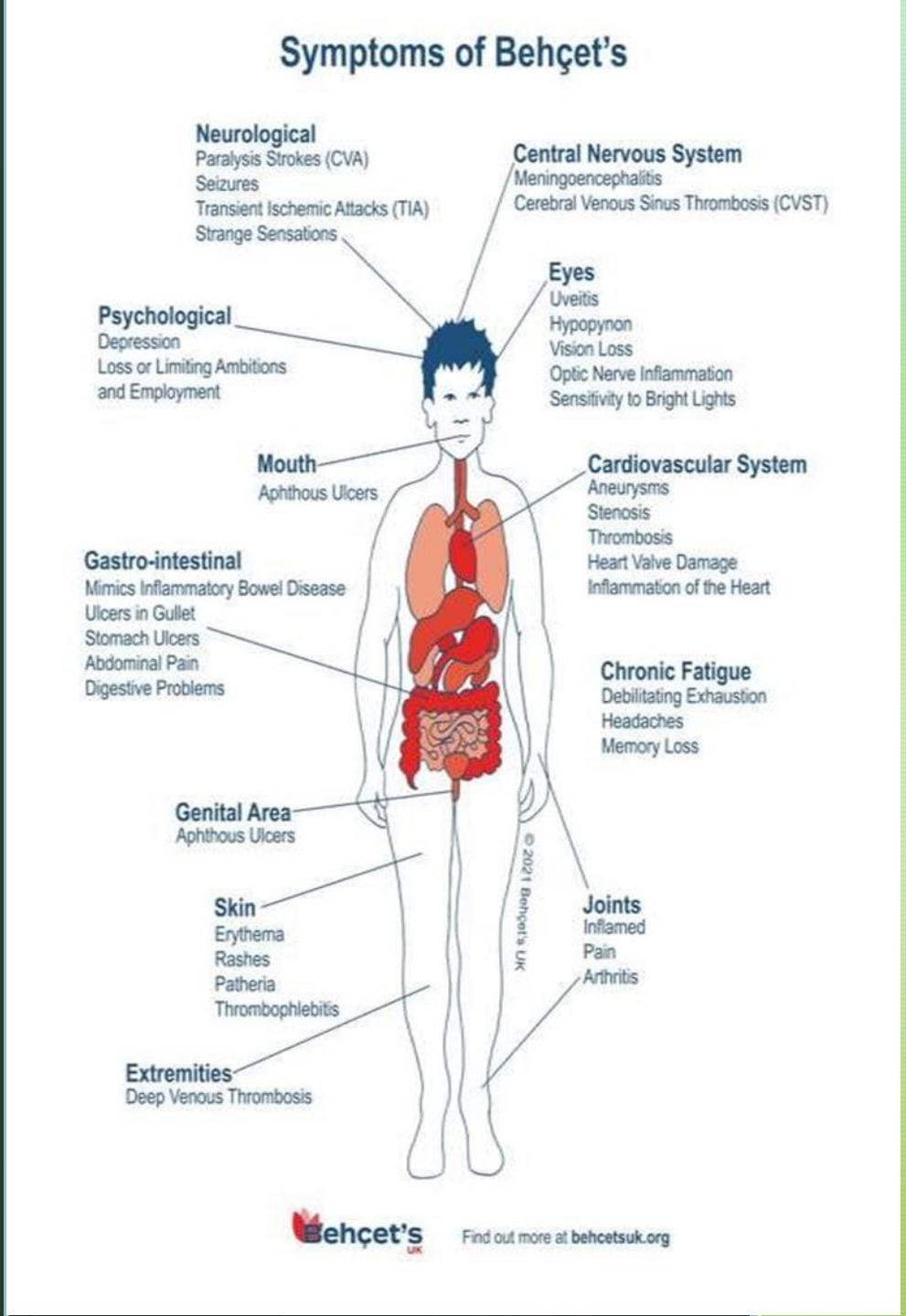
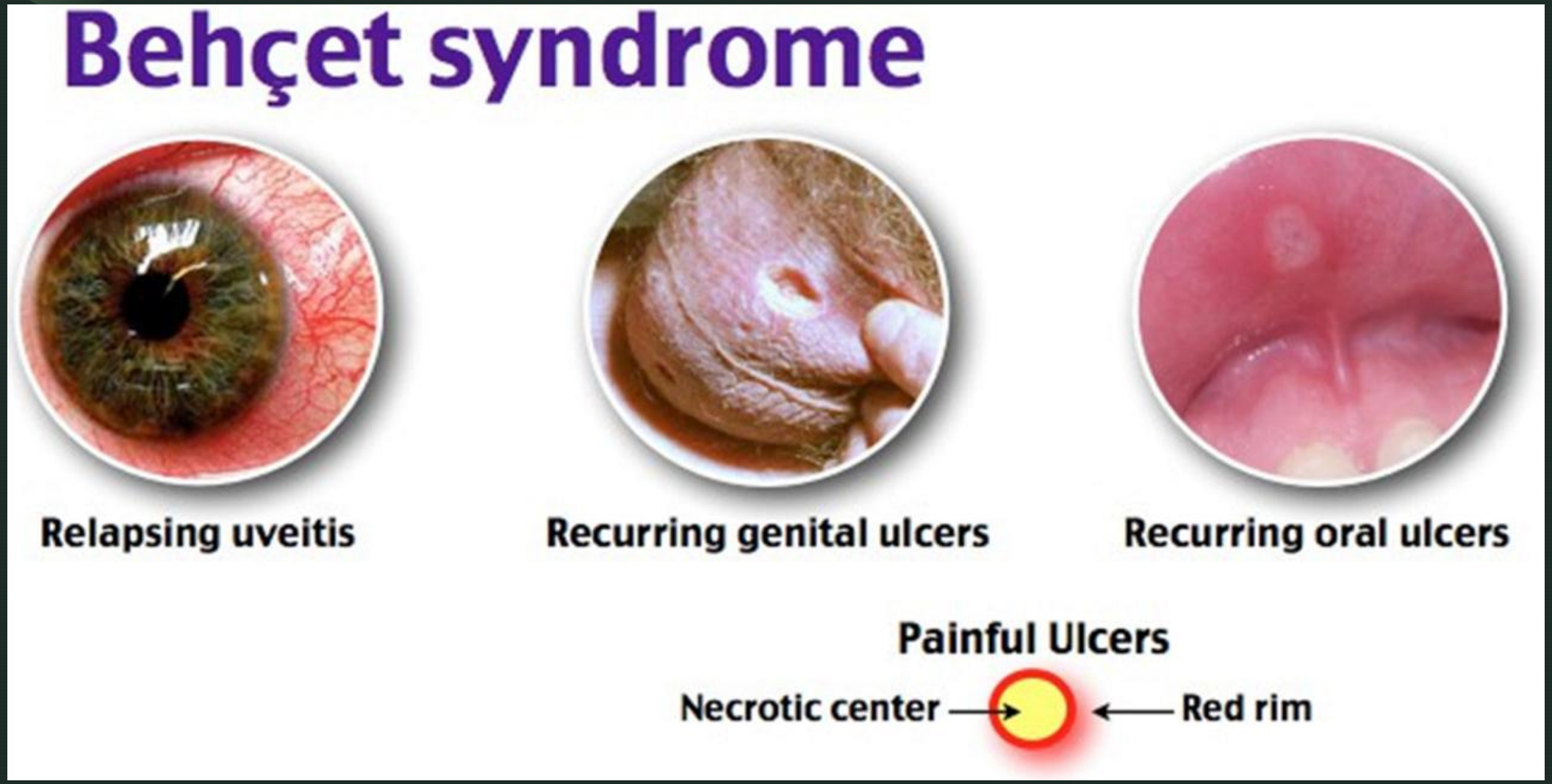


DEFINTION :

- Behcet disease (BD) is a chronic relapsing inflammatory vascular disease of unknown etiology
- Named after Hulusi Behcet, Turkish dermatologist found this disease characterized by a triad of recurrent oral ulcers, genital ulcers & uveitis

Triad of bahcet disease



- Other clinical features include skin lesions, arthritis, gastrointestinal involvement, neurologic disease, and vascular disease.
- Mucocutaneous and ocular disease are more active in early years, while the vascular and neurologic disease tends to come later on
- Ocular disease has the greatest morbidity, followed by vascular disease generally

- Patients are most commonly from the Middle East, the Mediterranean region, and the Far East; it is most prevalent in Turkey, with a prevalence of 1 in 250 adults.
- It is relatively rare before the late teens and after age 50. The commonest age (20-40 years).
- Males and females are equally affected; however, males frequently have more severe disease and poorer outcomes.
- Some manifestations may show regional differences; for example, gastrointestinal involvement, rare in Turkey, is more common in Japan and is seen in ~30% of patients in the United States.

Pathogenesis

Pathogenesis of BD is not completely clear.

Genetic, trigger factors, and immunological abnormalities are reported.

HLA-B* 51 & HLA-B5101 & HLA-B5103 are common risk factor in BD

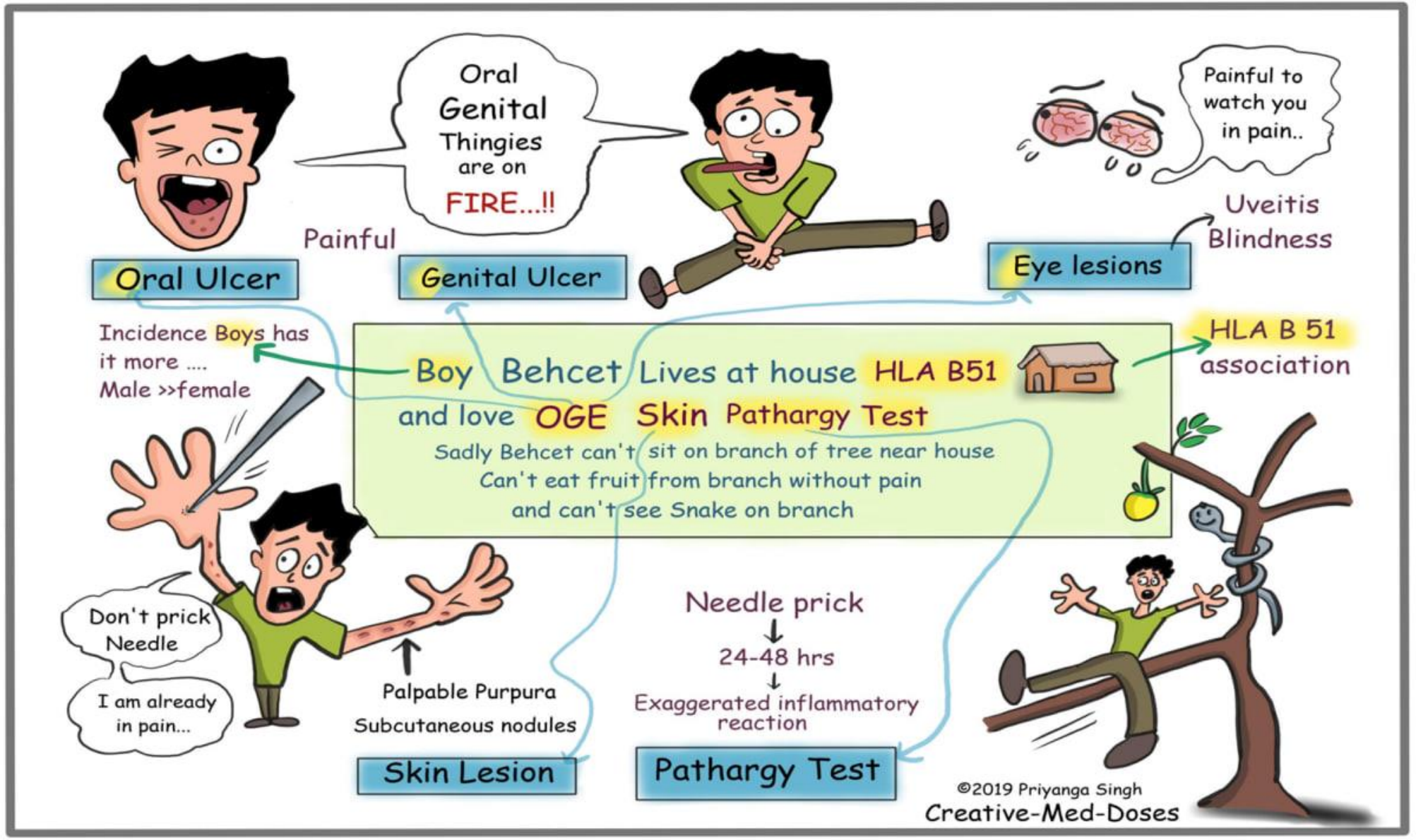
Reduced IL-10 (anti-inflammatory cytokine)

The pathogenesis includes autoimmunity, auto-inflammation & thrombophilia.

Infectious triggers:

HSV, strept, Staph & E coli have high affinity for HLA-B51 stimulate innate immunity, NK cells, gama delta T cells & neutrophils

Excessive production of pro-inflammatory cytokines (inflammation, stimulation of T cells, tissue destruction, endothelial dysfunction & thrombus formation


Oral Genital Thingies are on FIRE...!!
Painful
Oral Ulcer
Genital Ulcer
Painful to watch you in pain..
Uveitis
Blindness
Eye lesions
Incidence Boys has it more .... Male >>female
Boy Behcet Lives at house HLA B51 and love OGE Skin Pathargy Test
Sadly Behcet can't sit on branch of tree near house
Can't eat fruit from branch without pain
and can't see Snake on branch
HLA B 51 association
Don't prick Needle
I am already in pain...
Palpable Purpura
Subcutaneous nodules
Skin Lesion
Needle prick
24-48 hrs
Exaggerated inflammatory reaction
Pathargy Test
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1990 International Study Group (ISG) clinical criteria for Behçet's Disease (BD):

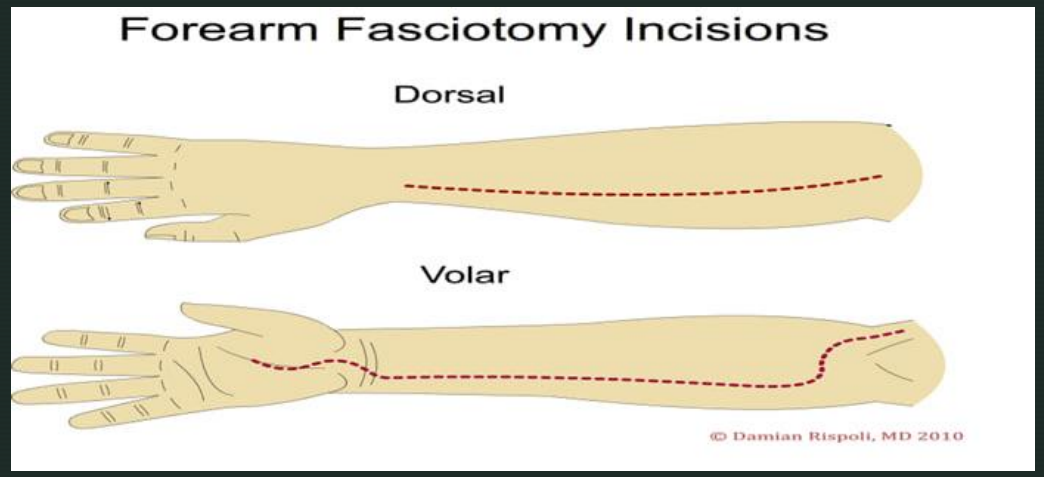
- ***Recurrent oral ulcerations (97-99%) at least 3 times/year plus 2 of the following:***
- Recurrent genital ulceration (80%)
- Skin lesions 80% (papulopustular lesions acneiform nodules, pseudo-folliculitis, EN)
- Eye lesions 50% (uveitis, vitritis, retinitis)
- Positive pathergy test (50%)

international criteria for behcet's disease (icbd), Scoring 4 is diagnostic :

Sign/symptom	Points
Ocular lesions	2
Genital aphthosis	2
Oral aphthosis	2
Skin lesions	1
Neurological manifestations	1
Vascular manifestations	1
Positive pathergy test*	1*

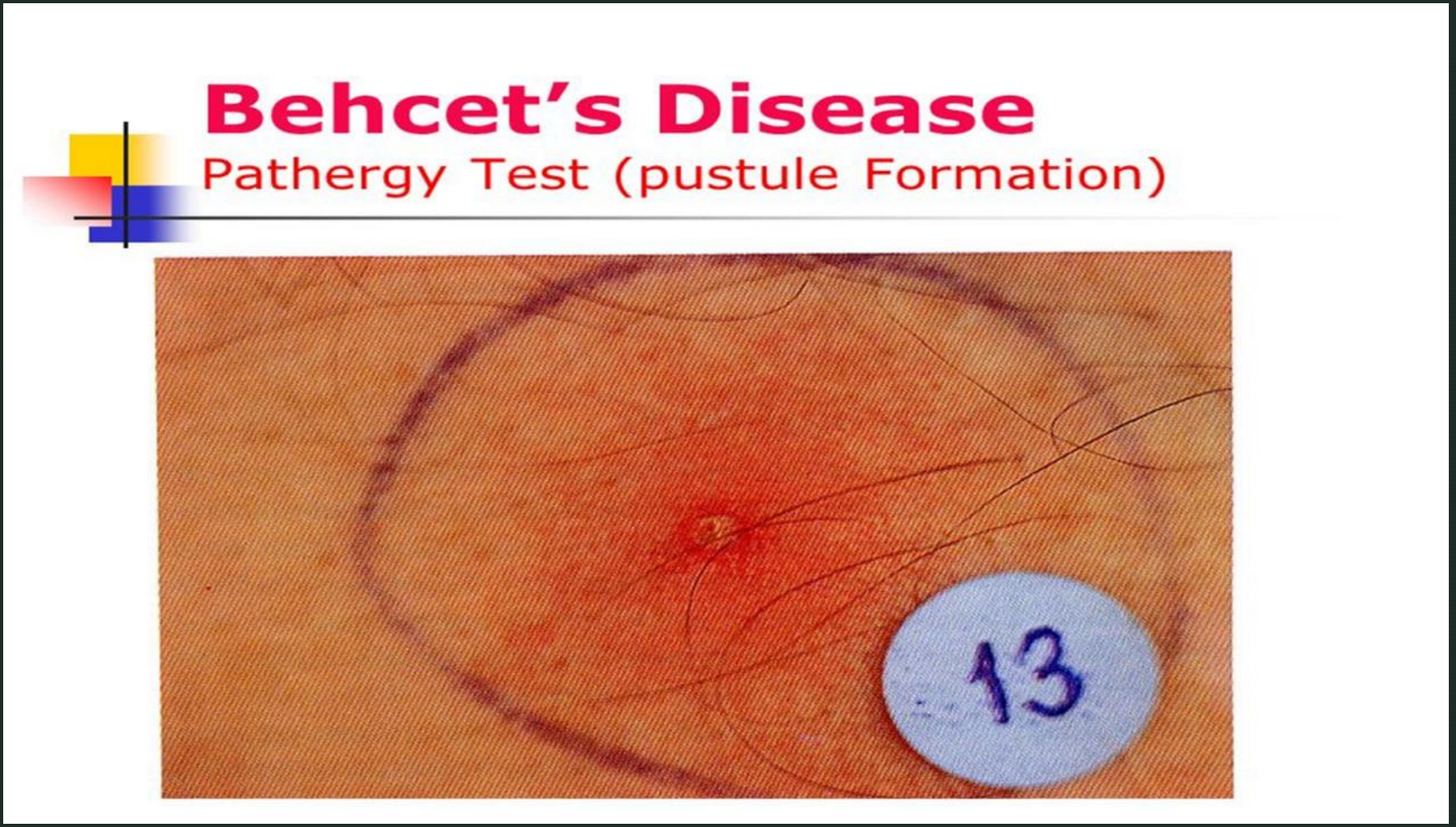
Pathergy test :

Pathergy test

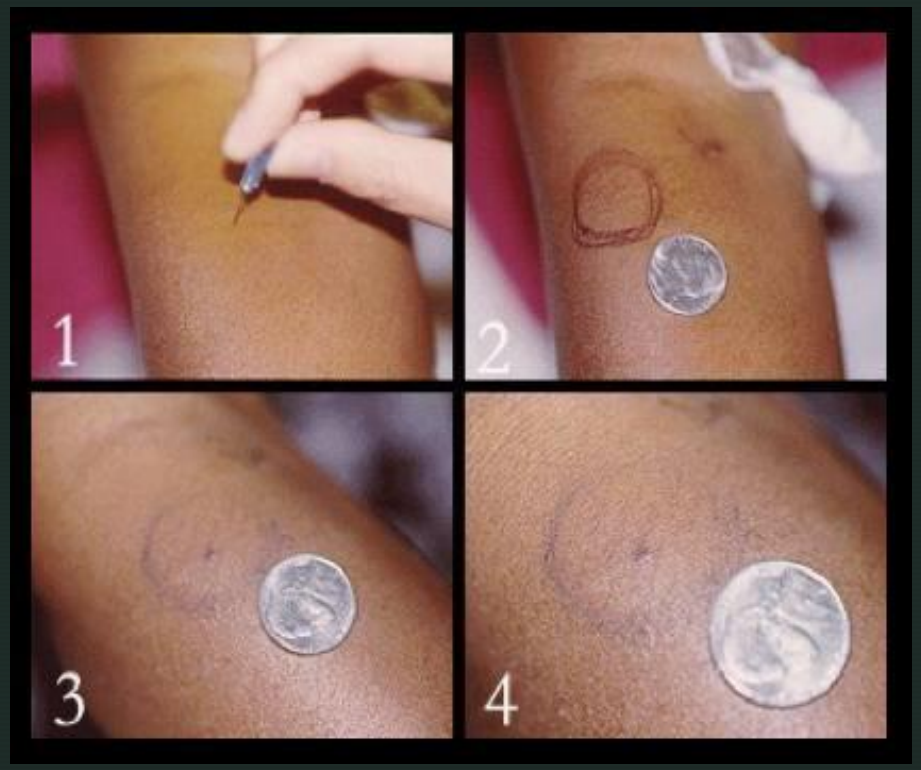


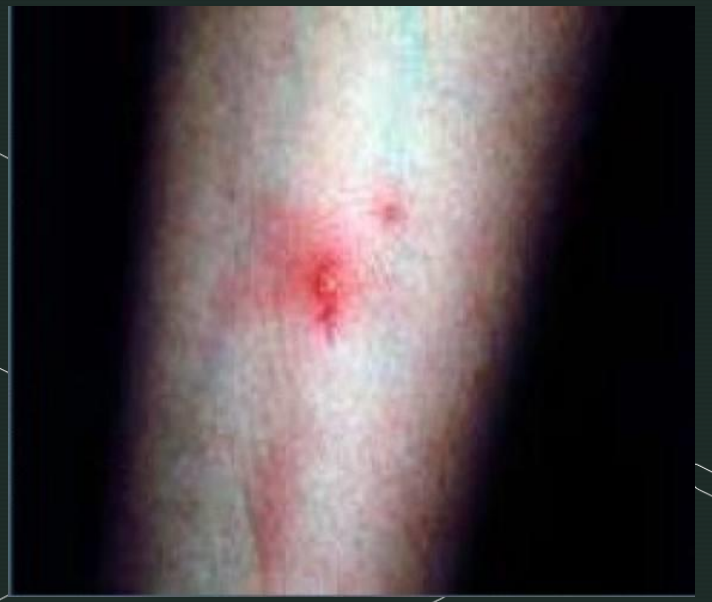
- Site: Commonly volar aspect of forearm.
- Intradermal injection of 0.1 ml isotonic salt solution using 20 G needle without prior disinfection of the injection site.
- 3-5 mm intradermally at an angle of 45 degree.
- Reading – after 24-48 hours
- +ve result- Erythematous papule or pustule (>2 mm) at prick site.

Positive result :



- Pathergy test is not pathognomic.
- +ve results can also occur in patients with,
- Pyoderma gangrenosum
- Rheumatoid arthritis
- Crohn disease
- Genital herpes infection

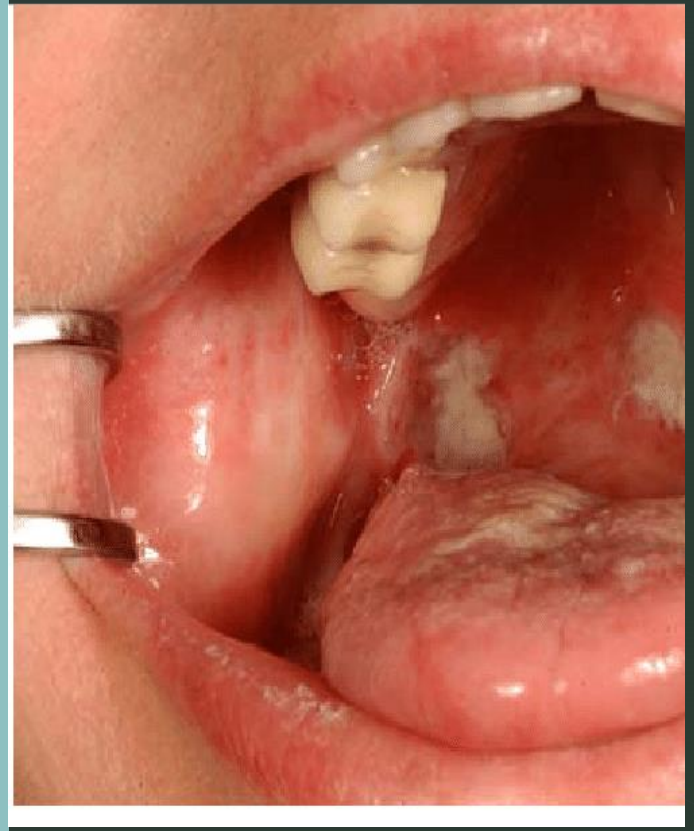


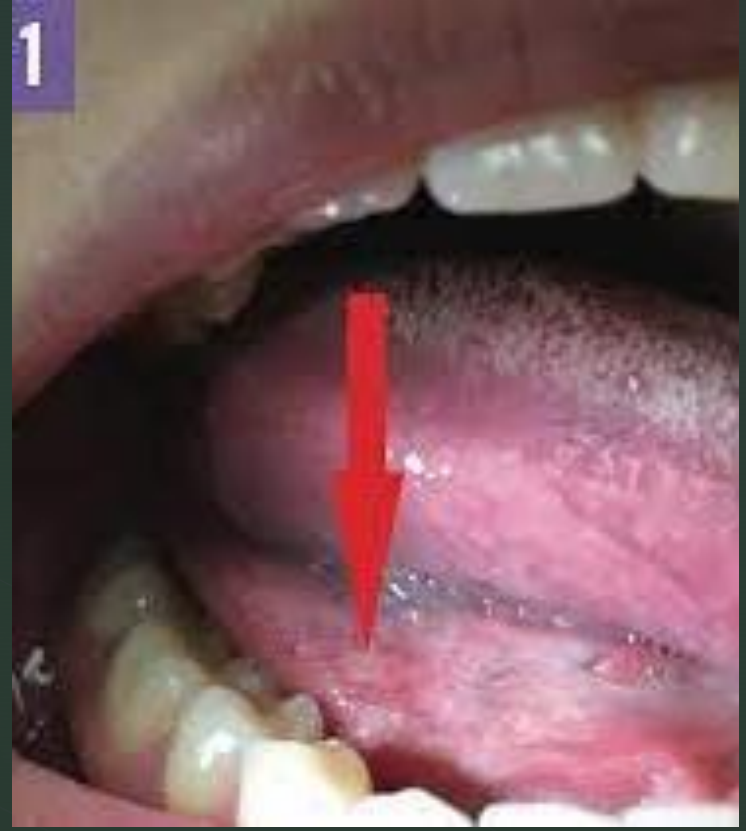


CLINICAL PRESENTATION

Oral Ulcers (97-99%)

- Painful oral ulcers
- Initial symptom (mostly)or seen at some time during the clinical course.
- Gingiva, tongue, buccal & labial mucosa.
- Round, with sharp, erythematous border & covered with yellowish pseudomembrane
- They last around 10 days without scarring but recur unless treated

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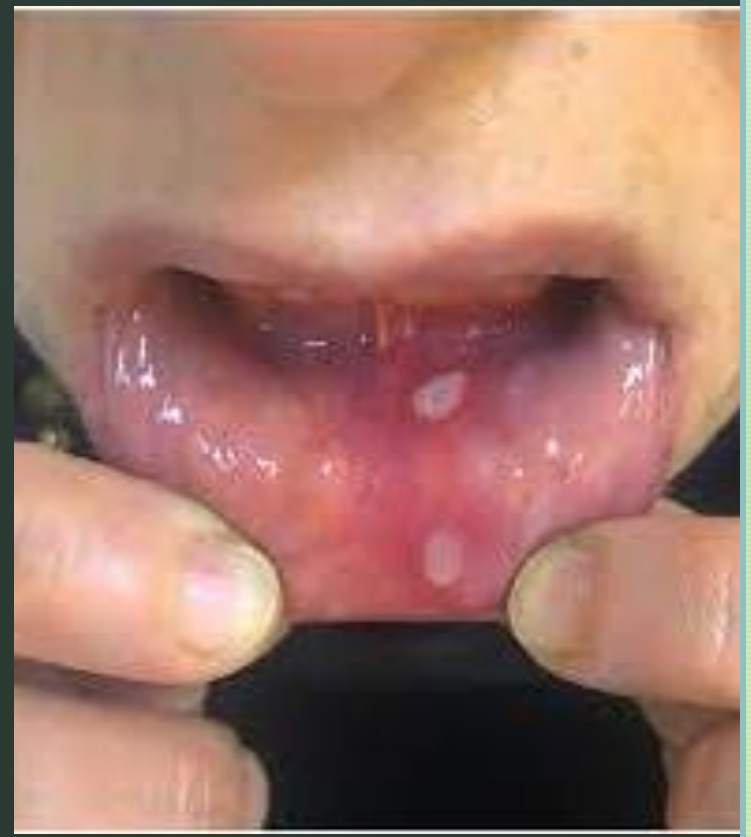

Genital ulcers (75%).

- On the scrotum and penis in males and on the vulva in females.
- Painful and morphologically similar to the oral ulcers but they are larger, deeper, with irregular margin and take longer to heal than oral ulcers and usually leave scars.


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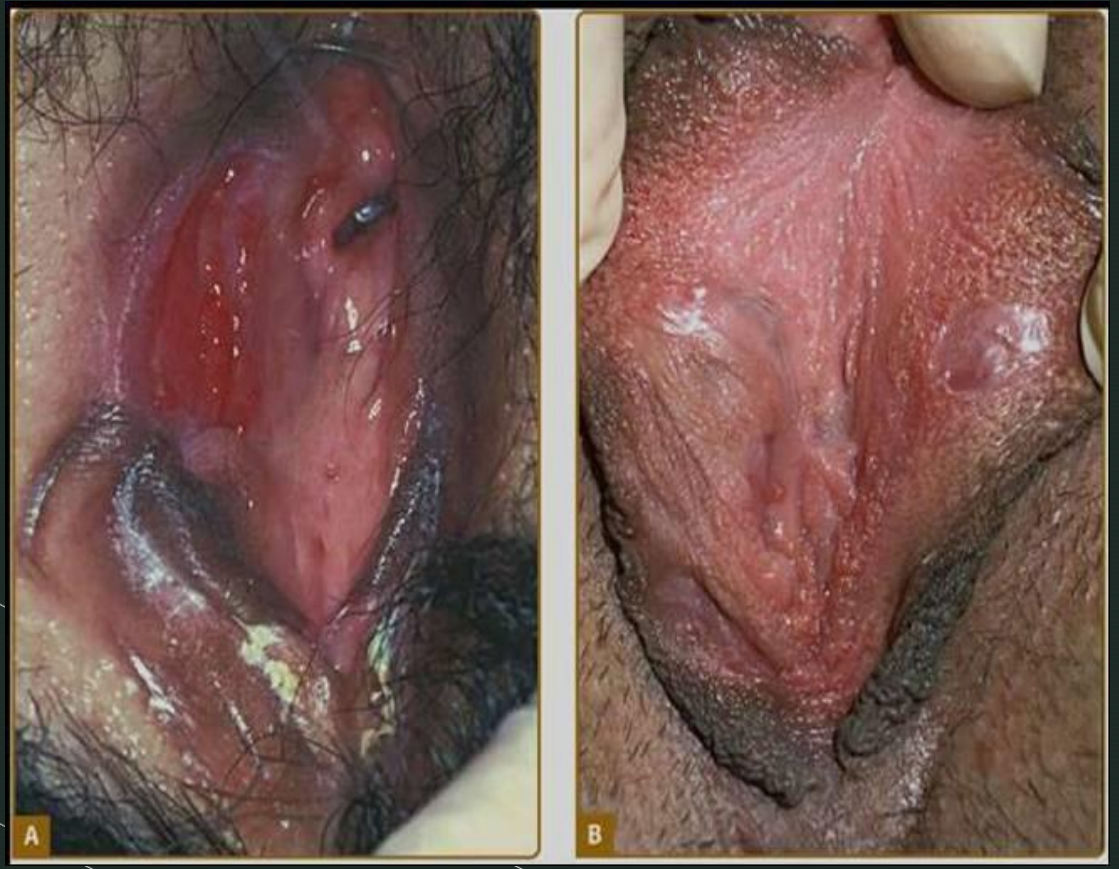
A
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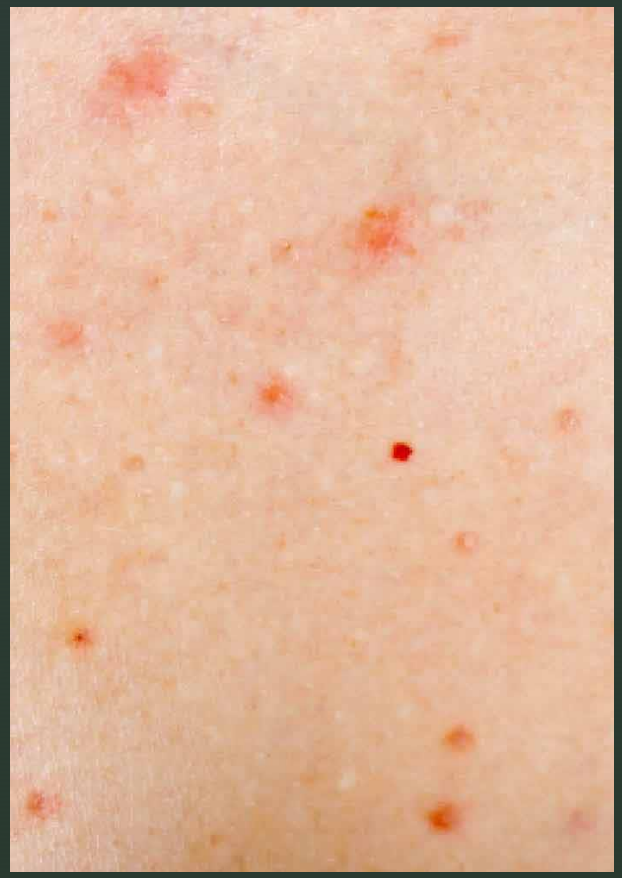
Cutaneous manifestatios (75%)

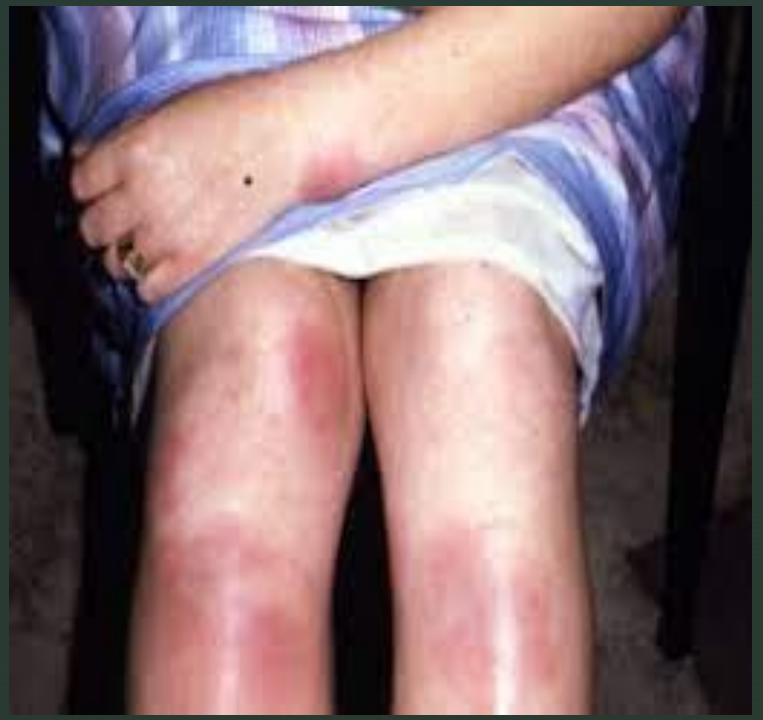
- Papulopustular acneiform lesions
- Pseudo-folliculitis,
- Erythema nodosum like lesion.
- Cutaneous small vessel vasculitis with ulcers.
- Pyoderma gangrenosum
- Hypersesitive skin (+ ve. Pathergy test)

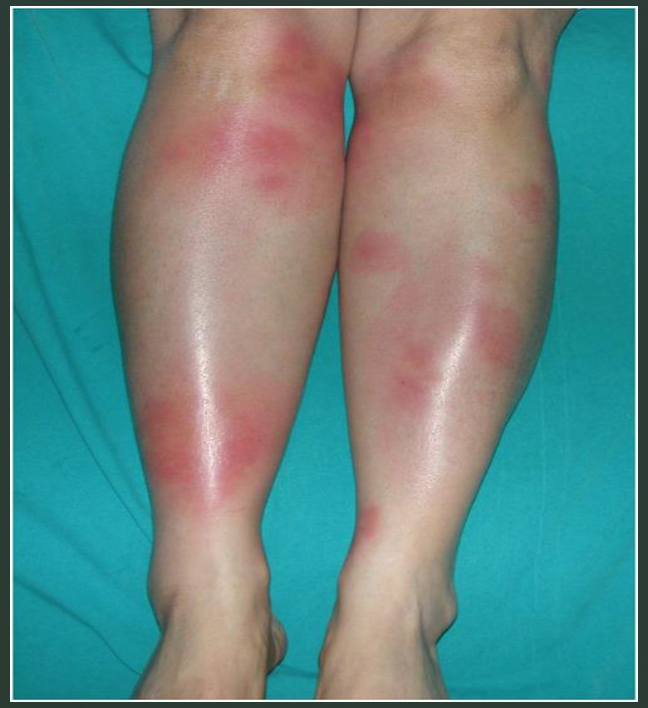
papulopustular acneiform lesions



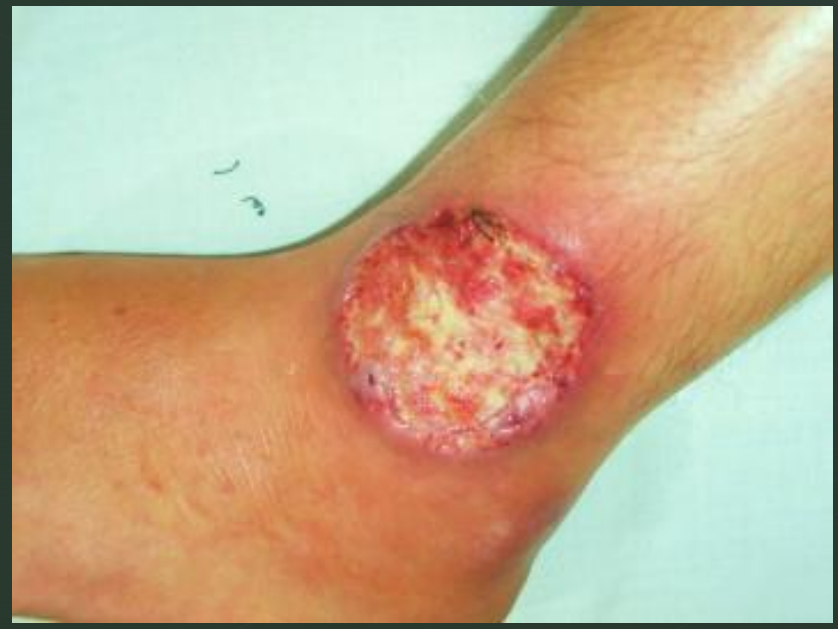
Pseudo-folliculitis

Erythema nodosum





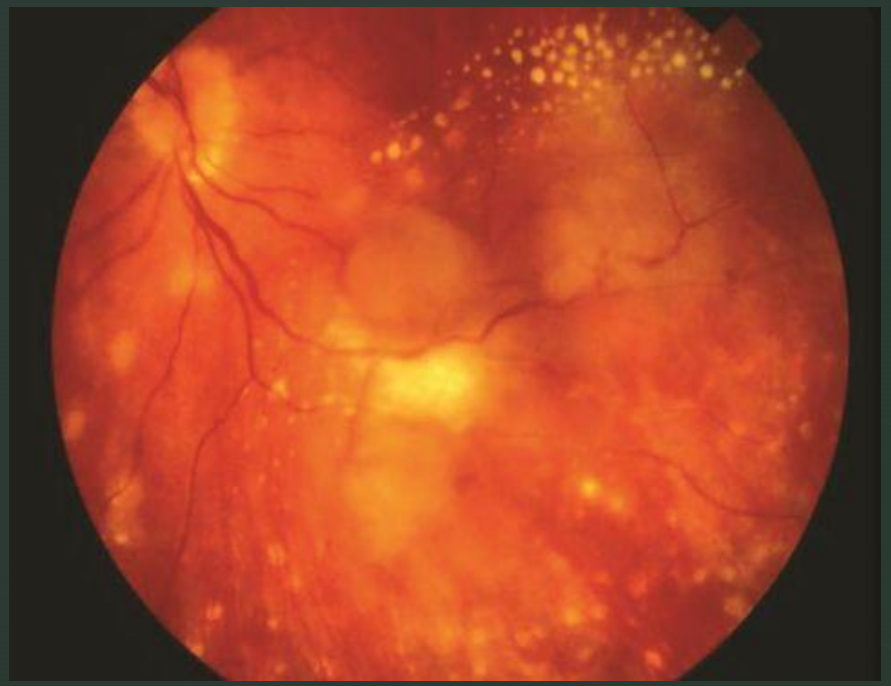
Pyoderma gangrenosum

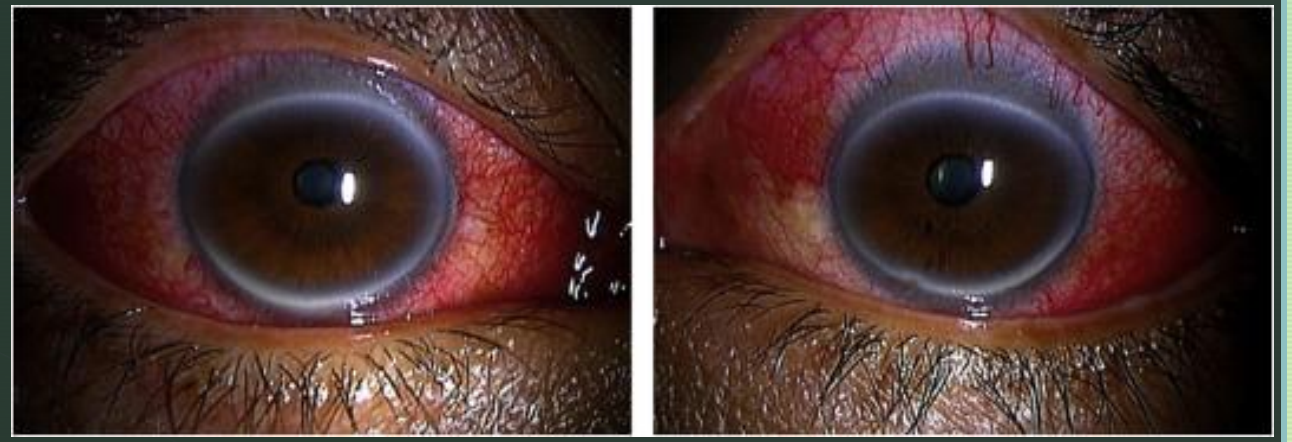


Inflammatory eye disease (50%)

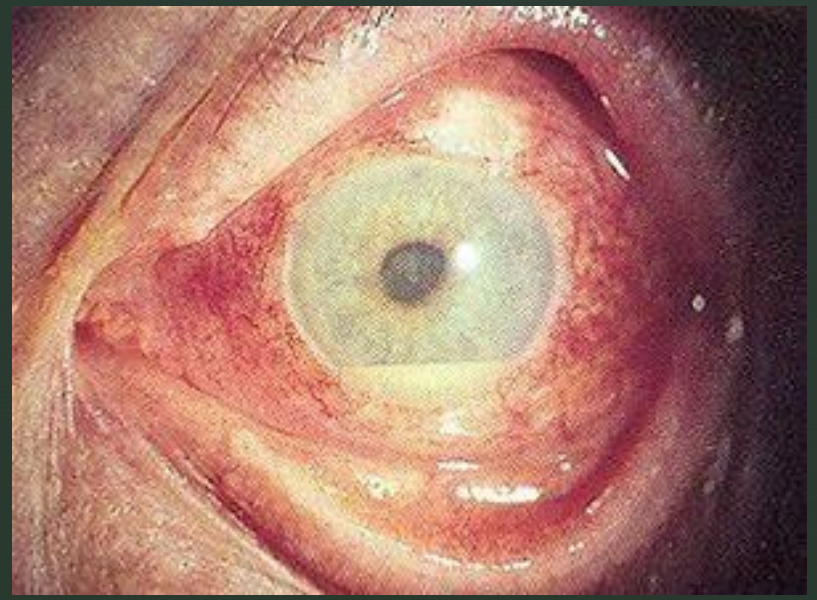
- More common in males of young age.
- Occurs latter on within 2-3 years of onset
- Better prognosis with later onset.
- Ocular Behcet's may be only presentation
- Poor prognosis: male, posterior involvement, frequent attacks (>3 per year), strong vitreous opacity, and macular edema
- It is most commonly a bilateral panuveitis
- A hypopyon, seen in ~10% of patients with eye disease, is an intense inflammation in the anterior chamber and is quite specific for Behçet syndrome.

panuveitis





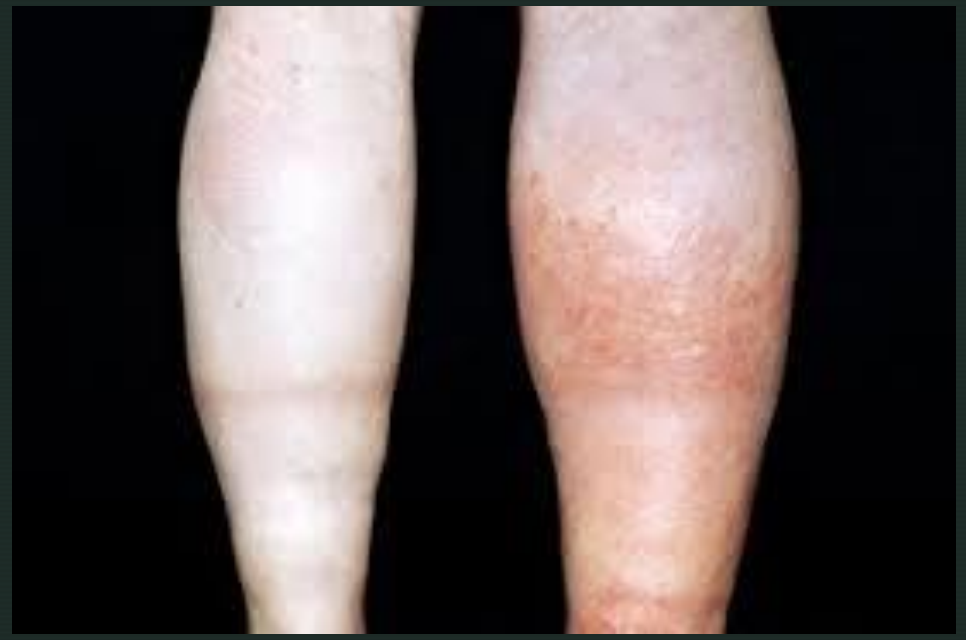
hypopyon



Vascular involvement (40%)

- Venous involvement as DVT or superficial thrombophlebitis is more common than arterial involvement (about 30%)
- Showering of pulmonary emboli is rare?
- The superior and inferior vena cava could be obstructed producing a dramatic clinical picture.
- Hepatic vein (Budd-Chiari syndrome)

DVT superficial thrombophlebitis



Pulmonary vasculitis

- Pulmonary artery thrombosis,
- Pulmonary artery hypertension.
- Pulmonary artery aneurysms (common complication, It is fatal complication manifests as hemoptysis, cough, chest pain, or dyspnea and needs urgent management).

Arterial involvement

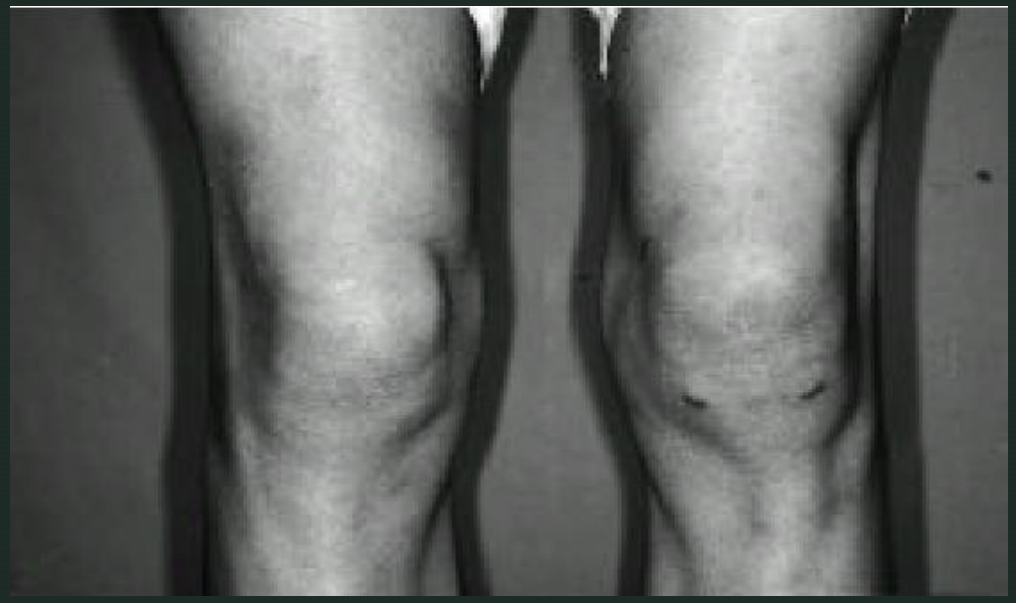
- Occurs in 5% of patients,
- More in males
- Presents as arterial aneurysm or thrombosis,
- Aorta and main branches could be affected

Neuro-Behcet disease (NBD).

- Like eye disease, is commoner in males and seen in (5-10%) of patients.
- Increased frequency of HLA-B5103
- Develop latter on, few years after the onset of other systemic features of BD
- It is either parenchymal or nonparenchymal
- Parenchymal involvement usually affects the telencephalic-diencephalic junction, brainstem, and spinal cord.
- Patients may present with a subacute onset of severe headache, cranial nerve palsy, dysarthria, ataxia, and hemiparesis.

Joint affection

- Non-deforming arthritis or arthralgias are seen in a 50% of patients
- it is usually a mono- or oligoarthritis in the lower extremities mainly knees & ankles.



Gastrointestinal

- Abdominal pain; ulcerative lesions at any level but mainly ileocaecal region;
- Clinical differentiation from IBD is difficult.
- it is difficult to distinguish Behçet syndrome from Crohn’s disease unless extraintestinal lesions are present.

Cardiac manifestations (5-10%):

- Coronary vasculitis and thrombosis,
- Pericarditis, myocarditis, endocarditis e granulomatous changes or fibrosis, and diastolic dysfunction

- Nephrotic syndrome.
- Epididymitis, urethritis &neurogenic bladder
- Materno-fetal problems (hypercoagulability)

investigations

- Complete blood count: neutrophilia
- C –reactive protein- raised
- ESR-raised
- HLA-B51 test-positive
- Skin Biopsy
- CSF analysis
- MRI/CT Scan
- Chest X ray
- Endoscopy
- Pathergy test

TREATMENT

- Treatment is guided by type and severity of involvement, with the goal of preventing long-term damage.
- Most new manifestations present within the first 5 years, and for most patients, the natural course is one of diminishing symptoms culminating in potential remission, frequently not requiring ongoing treatment with medications.
- Patient characteristics, such as being young and male, need to be kept in mind as these patients tend to have a worse prognosis.
- For most patients, tapering and/or stopping their medications in 2–3 years after the symptoms have improved should be attempted.

Inflammatory eye disease

- glucocorticoids and longer-term treatment with an immunosuppressant, azathioprine is usually the preferred agent.
- Infliximab, adalimumab, or cyclosporine can also be used, in combination with systemic glucocorticoids and azathioprine, for control of disease activity
- Glucocorticoids can be tapered in many patients after active disease has been controlled, whereas immunosuppressants are generally continued for at least 2 years

Oral ulcers

- can be managed with topical glucocorticoids and on an as-needed basis if mild
- Lesions resistant to local measures may require systemic treatment with colchicine, oral glucocorticoids, immunosuppressants such as apremilast, azathioprine, or a tumor necrosis factor-α inhibitor such as infliximab
- Patients may need a combination of medications, at least initially, to control disease activity.
- A similar treatment approach can be used for genital ulcers and other mucocutaneous manifestations.

- **Gastrointestinal involvement:** is treated with a glucocorticoid plus an immunosuppressant such as azathioprine alone or in combination with infliximab

- **Venous thrombotic events:** are treated by controlling systemic inflammation with immunosuppressive medications (usually azathioprine or, for more severe cases, cyclophosphamide), rather than using anticoagulants.

- **For central nervous system involvement:** the combination of azathioprine and a tumor necrosis factor inhibitor is usually the first choice.

عَلَى هَذِهِ الأَرْضِ مَا يَسْتَحِقُّ الحَيَاةْ