

BEHÇET'S DISEASE

- **Idiopathic multisystem disease**
- **More common in men**
- **Occurs in 3rd - 4th decade**
- **Highest incidence in Mediterranean region and Japan**
- **Associated with HLA-B5**



BEHÇET'S DISEASE

Aetiology

- Unknown
- Various bacteria and viruses suggested
- No good evidence to suggest any of them
- Perpetuated by autoimmune response and CD4 + T-cells
- Tumour necrosis factor (TNF) thought to be important

BEHÇET'S DISEASE

Systemic Involvement (1)

Oral aphthous
ulceration – 100%



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Systemic Involvement (2)

Genital ulceration –
90%



BEHÇET'S DISEASE

Systemic Involvement (3)

- Skin lesions – 80%
 - Erythema Nodosum
 - Acneiform
- Uveitis 70% (inflam. of iris, ciliary body or choroid)



BEHÇET'S DISEASE

Systemic Involvement (4)

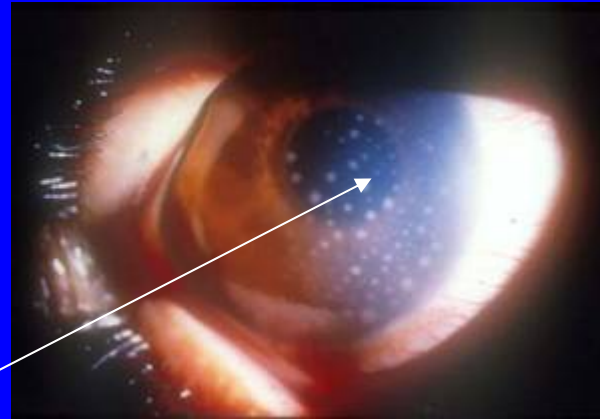
- CNS involvement – strokes, fits
- Major vessels eg superior Vena cava obstruction
- Increased skin response to trauma eg blood taking



BEHÇET'S DISEASE

Ocular Features (1)

- **Acute iritis**
 - Pain, redness & ↓VA
 - Flare (PTN exudation)
 - Inflammatory cells in anterior chamber
 - KPs (Inflammatory cells at posterior surface of cornea)
- **Recurrent hypopyon**
(Fluid level of WBC)
The red or white eye



BEHÇET'S DISEASE

Ocular Features (3)

- **Marked inflammation of the eye**
- **Retinal vasculitis and haemorrhage (inflam. of retinal vessels)**
- **Occlusive periphlebitis (venous sheathing & occlusion)**
- **Retinal microinfarcts**
- **Very damaging to vision: retinal damage and optic nerve atrophy**
- **Cataract or glaucoma**



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Treatment

- Systemic Steroids
- Systemic immunosuppressive agents
- Interferon-alpha may have immunodulating effects
- Anti-TNF monoclonal antibodies may be of help