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%
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Systemic Involvement (3)

• Skin lesions – 80%
  – Erythema Nodosum
  – Acneiform

• Uveitis 70% (inflam. of iris, ciliary body or choroid)
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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
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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
BEHÇET’S DISEASE

Treatment

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