91) Which one of the following is the most common manifestation of Behcet’s disease?

A. Recurrent oral ulceration
B. Recurrent genital ulceration
C. Central nervous system symptoms
D. Anterior uveitis
E. Positive pathergy test

Answer:

Behcet’s Disease
- Multisystem chronic relapsing inflammatory disorder
- Likely vasculitic phenomenon
- Tendency towards venous thrombosis
- Associated HLA B5
- Prevalence varies geographically, more common in Far East, Middle Eastern, Mediterranean
- Affects young males and females equally
- Disease more severe in males
- Disease severity often abates with time, usually normal life expectancy (unless neurological complications)

- International Criteria for diagnosis:
  - Recurrent oral ulcers (> 3 per year) + at least 2 of the following:
    - Recurrent genital ulcers
    - Eye lesions (anterior/posterior uveitis/cells in vitreous humour/retinal vasculitis)
    - Skin lesions (erythema nodosum/papular lesions/acneiform rash)
    - Positive Pathergy Test (Papule > 2mm 24-48 hours post 20G needle skin prick)

- The oral ulcers are painful
- Shallow/deep with central yellow necrotic base
- Singly or in crops
- Anywhere in oral cavity
- Can be found anywhere in GIT mainly terminal ileum, caecum and ascending colon
- Persists 1-2 weeks then resolves without scarring
- May be less common in cigarette smokers

- Genital ulcers less common
- Do not affect glans penis or urethra
- Mainly scrotum (leaving scars) and vulva
- Rarely may cause epididymitis, salpingitis

- Eye involvement occur in 75%
- Worst complication is blindness

Other features:
- Arthritis (not deforming, knees and ankles)
- Superficial/DVT (up to 25%) +/- rarely PE

- Neurological involvement in 5-10% (parenchymal form affecting brain stem with poor prognosis +/- dural vein thrombosis +/- aseptic encephalitis/meningitis)

- Cardiac involvement uncommon
- Renal disease uncommon (may have proteinuria or secondary amyloidosis)
- No diagnostic test: non-specific raised inflammatory markers + ↑ ICAM-1 levels correlating to disease activity
Treatment:
- Mucous membrane: topical glucocorticoids/ colchicine/ IFN alpha
- Thrombophlebitis: aspirin 325 mg/day
- Eye and CNS symptoms: prednisolone +/- azathioprine/ cyclosporine
- Early data for anti TNF Ab promising

Ddx oral ulcers
1) Simple aphthous ulcer
2) Ulcer secondary trauma (biting/ dentures)
3) Herpes simplex ulcers
4) Inflammatory bowel disease (especially Crohns)
5) Other rheumatological diseases eg SLE
6) Drugs eg methotrexate/ chemotherapy
7) Blistering skin disease eg pemphigoid/ pemphigus
8) Lichen planus
9) Linear Ig A disease