Pemphigus

Types
- Pemphigus vulgaris: 3/4 of cases (common in Ashkenazi Jews, Mediterraneans, and Indian)
- Pemphigus vegetans: rare
- Superficial pemphigus
  - Generalized foliaceous
  - Localized erythematous
- Drug eruption: penicillamine
- Paraneoplastic pemphigus
  - Thymoma
  - Castleman's tumour
  - Lymphoma

Cause: All are autoimmune, IgG mediated
- Main antigens: desmoglein 3, desmoglein 1

Presentation
- Pemphigus vulgaris: flaccid blisters
- Pemphigus vegetans: heaped-up, cauliflower-like weeping areas
- Pemphigus foliaceus: so superficial, blisters rupture easily
- Pemphigus erythematous: dominated more by weeping and crusting erosions than blisters

Complications
- Due to high dose of steroids and immunosuppressives
- Severe oral ulcers make eating painful
- Dehydration and electrolytes imbalance

Course
- Severe, life-threatening
- It affects middle age
- 3/4 of cases
- 15% mortality rate
- 1/3 go into remission after 3 years
- Superficial pemphigus is less severe

Investigations
- Biopsy
- Biopsy: Acantholysis
- Direct immunofluorescence
  - Intercellular epidermal deposits of IgG and C3
- Indirect immunofluorescence
  - Titre of these antibodies
- ELISA
  - Tzanck smear

Differential diagnosis
- Pyoderma
- Impetigo
- Ecthyma
- Epidermolysis bullosa
- Aphthous ulcer
- Behçet's disease
- Herpes simplex infection
- Bacterial or fungal infections
- Mouth ulcers
- Widespread erosions
- Differential diagnosis

Treatment
- In superficial pemphigus: small doses of systemic corticosteroids, topical corticosteroids, azathioprine, gold salts, cyclophosphamide, mycophenolate mofetil
- Immunosuppressives: plasmapheresis, intravenous immunoglobulin, Dapsone
- Drug eruption: penicillamine
- Paraneoplastic pemphigus: thymoma, Castleman's tumour, lymphoma

Due to high dose of steroids and immunosuppressives
- Severe oral ulcers make eating painful
- Dehydration and electrolytes imbalance

Presentation
- Most patients develop the mouth lesions first
- Blisters rupture easily
- Leaves widespread painful erosions
- Shearing stresses on normal skin can cause new erosions
- Also positive in toxic epidermal necrolysis

Vegetans variant
- Heaped-up, cauliflower-like weeping areas are present in the groin and body folds

Course
- Prolonged
- Mortality rate: 15%
- 1/3 go into remission after 3 years
- Superficial pemphigus is less severe