Pemphigus

- Treatment
  - In superficial pemphigus
    - small doses of systemic corticosteroids
    - topical corticosteroids
  - high doses of systemic steroids
  - mmunosuppressives
    - azathioprine
    - gold salts
    - cyclophosphamide
    - mycophenylate mofetil
  - plasmapheresis
  - intravenous immunoglobulin
  - Dapsone
- Investigations
  - Biopsy
    - vesicles are intra-epidermal
    - rounded keratinocytes floating freely within the blister cavity
    - Acantholysis
  - Direct immunofluorescence
    - intercellular epidermal deposits of IgG and C3
  - Indirect immunofluorescence
    - serum from a patient with pemphigus contains antibodies that bind to the desmogleins in the desmosomes of normal epidermis
  - ELISA
  - titre of these antibodies
    - correlates loosely with clinical activity
  - Tzank smear
    - acantholysis
- Differential diagnosis
  - Widespread erosions
    - pyoderma
    - impetigo
    - ecthyma
    - epidermolysis bullosa
  - **Mouth ulcers**
    - Aphthous ulcer
    - Behçet's disease
    - herpes simplex infection
  - Scalp erosions
    - bacterial or fungal infections
- Complications
  - Due to high dose of steroids and immunosuppressives
  - severe oral ulcers make eating painful
  - dehydration and electrolytes imbalance
- severe
  - life-threatening
  - it affects middle age
- Types
  - pemphigus vulgaris
    - 3/4 of cases
    - Pemphigus vegetans
      - rare
    - common in Ashkenazi Jews, Mediterraneans and Indian
  - superficial pemphigus
    - generalized foliaceus
    - localized erythematosus
  - 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
      - On skin
        - Trunk
        - flexures
        - scalp
      - In mouth
    - Most patients develop the mouth lesions first
    - blisters rupture easily
    - Leaves widespread painful erosions
    - Positive Nikolsky sign
      - 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
  - pemphigus foliaceus
    - Blisters
      - so superficial
      - rupture so easily
    - Dominated more by weeping and crusting erosions than by blisters
  - pemphigus erythematosus
    - facial lesions
    - pink, rough and scaly
  - patient is in poor health bcoz of the diseases it self
- Course
  - Course is prolonged
  - mortality rate
    - 15%
  - 1/3 go into remission after 3 years
  - Superficial pemphigus is less severe