Pemphigus

Types
- pemphigus vulgaris: 3/4 of cases
- Pemphigus vegetans: rare
- Pemphigus foliaceus: common in Ashkenazi Jews, Mediterraneans, and Indian
- Pemphigus erythematosus: localized erythematous
- Drug eruption
- Paraneoplastic pemphigus
- Penicillamine
- 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 mouth lesions first
  - Blisters rupture easily
  - Wakes widespread painful erosions
  - Positive Nikolsky sign
  - Shearing stresses on normal skin can cause new erosions
- Pemphigus vegetans: heaped-up, cauliflower-like weeping areas are present in the groin and body folds
- Pemphigus foliaceus: blisters
  - So superficial
  - So easily ruptured
  - Dominated by weeping and crusting erosions than by blisters
  - Facial lesions
  - Pink, rough, and scaly
- Pemphigus erythematosus: facial lesions
  - Pink, rough, and scaly

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

Complications

Drug eruptions
- Penicillamine
- Thymoma
- Castleman’s tumour
- Lymphoma

Investigations
- Biopsy
- Vesicles are intra-epidermal
- Acantholysis
- Rounding keratinocytes floating freely within the blister cavity
- Direct immunofluorescence
- Intercellular epidermal deposits of IgG and C3
- Indirect immunofluorescence
- ELISA
- Serum from a patient with pemphigus contains antibodies that bind to the desmogleins in the desmosomes of normal epidermis
- Titer of these antibodies correlates loosely with clinical activity

Differential diagnosis
- Pyoderma
- Impetigo
- Ecthyma
- Epidermolysis bullosa
- Aphthous ulcer
- Behçet’s disease
- Herpes simplex infection
- Bacterial or fungal infections
- Mouth ulcers
- Widespread erosions
- Scalp erosions
- Due to high dose of steroids and immunosuppressives
- Severe oral ulcers make eating painful

Treatment
- In superficial pemphigus: small doses of systemic corticosteroids, topical corticosteroids, high doses of systemic steroids, azathioprine, gold salts
- Immunosuppressives: cyclophosphamide, mycophenolate mofetil, plasmapheresis, intravenous immunoglobulin, Dapsone
- Drug eruption: penicillamine
- Paraneoplastic pemphigus
- Thymoma
- Castleman’s tumour
- Lymphoma
- Superficial pemphigus: small doses of systemic corticosteroids, topical corticosteroids, high doses of systemic steroids, azathioprine, gold salts, cyclophosphamide, mycophenolate mofetil, plasmapheresis, intravenous immunoglobulin, Dapsone
- Pemphigus vegetans: penicillamine
- Pemphigus foliaceus: topical corticosteroids
- Pemphigus erythematosus: high doses of systemic steroids, azathioprine, gold salts, cyclophosphamide, mycophenolate mofetil, plasmapheresis, intravenous immunoglobulin, Dapsone

Signs
- Positive Nikolsky sign
- Shearing stresses on normal skin can cause new erosions