NOTES
VESICULOBULLOUS DISEASES

GENERALLY, WHAT ARE THEY?

PATHOLOGY & CAUSES

- Chronic skin blistering diseases; associated with underlying autoimmune, genetic pathology
- Destruction/malfunction of structural, anchoring proteins of skin

SIGNS & SYMPTOMS

- Skin blistering
- Mucosal erosions pruritus

DIAGNOSIS

LAB RESULTS

- Skin biopsy
- Immunofluorescence testing

TREATMENT

MEDICATIONS

- Corticosteroids

OTHER INTERVENTIONS

- Lifestyle modifications

BULLOUS PEMPHIGOID

osms.it/bullous-pemphigoid

PATHOLOGY & CAUSES

- Autoimmune skin disease; bubble-like blisters
  - *Bulla-* = blister, *pemphig-* = bubble, *oid-* = similar
- Chronic, relapsing, remitting, autoimmune subepithelial blistering disease
  - Epithelial lesions (unlike pemphigus vulgaris)
  - Can affect mucous membranes
- Presents with cutaneous bullae, mucosal erosions
- Rare disease, most common autoimmune blistering disorder

CAUSES

- Autoantibodies against hemidesmosomal proteins
  - Bullous pemphigoid antigen 2 (BPAg2)
  - Collagen type XVII
- Autoantibodies may develop in response to certain drugs/infections
- Autoantibody activation → abnormal IgG/complement deposition in basement membrane zone → separation of dermis, epidermis → inflammatory reaction → formation of blisters, inflammatory mucosal erosions

RISK FACTORS

- More common in individuals > 60 years

SIGNS & SYMPTOMS

- Trunk, skin folds, extremities most affected
- May exhibit prodromal phase
  - Pruritic papular lesions, resemble eczema
- Oral, ocular mucositis
- Blisters with inflammatory/noninflammatory base
- Unlike pemphigus vulgaris, bullae tense, difficult to rupture → negative Nikolsky sign
- After rupture, scarring uncommon

DIAGNOSIS

LAB RESULTS

- Histopathological studies (confirm)
  - Skin biopsies, immunofluorescent staining techniques
- Complete blood count (CBC)
  - Eosinophilia
- ↑ IgG antibodies

TREATMENT

MEDICATIONS

- Topical/systemic corticosteroids
  - Decrease blister formation, promote blister healing, improve quality of life

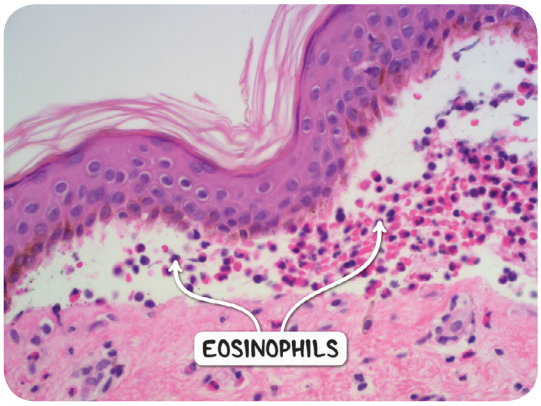

Figure 10.1 The histological appearance of the skin in a case of bullous pemphigoid. In contrast to pemphigus vulgaris, the bullae are subepithelial. The bullae contain an infiltrate of eosinophils.

EPIDERMOLYSIS BULLOSA

osms.it/epidermolysis-bullosa

PATHOLOGY & CAUSES

- Skin breakdown → blisters
  - *Epidermo-* = skin, *lysis-* = breakdown, *bullosa-* = blistering
- Rare condition, inherited group of disorders; blisters, erosions after minor skin trauma due to fragility of epithelial tissue
- May also affect mucous membranes, nails

CAUSES

- Mutations in structural proteins of skin responsible for tissue integrity
  - Keratin, desmosomes, cell junctions, intermediate filaments, etc.
  - Presence of some or all → determine disease severity, clinical presentation

RISK FACTORS

- Genetic inheritance

SIGNS & SYMPTOMS

- Localized/systemic
- Skin blistering following minor trauma/friction
- Nail dystrophy, loss (common)
- Oral lesions

DIAGNOSIS

LAB RESULTS

- Skin biopsy
- Immunofluorescence testing

OTHER DIAGNOSTICS

- Family history

TREATMENT

- No specific therapy

OTHER INTERVENTIONS

- Symptomatic care, wound care, infection prophylaxis, pain management

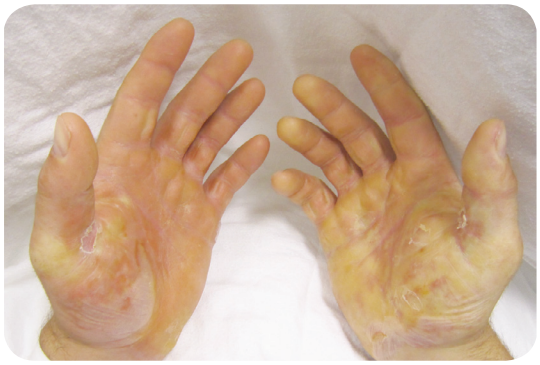

Figure 10.2 The hands of an individual with epidermolysis bullosa. Numerous consecutive bullae have caused scarring and induration of the skin, leading to contractures.

PEMPHIGUS VULGARIS

osms.it/pemphigus-vulgaris

PATHOLOGY & CAUSES

- Autoimmune, life-threatening blistering disorders of skin, mucous membranes
- Epithelial lesions
  - Unlike bullous pemphigoid, which presents with subepithelial lesions
- *Acantholysis:* defining mechanism (*acanthus-* = thorny, *lysis-* = breakdown)
  - Impaired adhesion between cells in spinous layer of epidermis

CAUSES

- Autoantibodies against desmoglein
- Autoantibody activation → attack of adhesion molecules → breakdown of intercellular adhesion → inflammatory reaction → blister formation

RISK FACTORS

- Adults
- Jewish people of Middle Eastern origin

SIGNS & SYMPTOMS

- Oral mucosa (most common); can affect all mucosal surfaces
- Nikolsky sign → blister ruptures with pressure/friction
  - Unlike bullous pemphigoid, where blisters difficult to rupture
- Easily-eroding painful blisters over erythematous skin
- No pruritus

DIAGNOSIS

LAB RESULTS

- Skin biopsy
- Immunofluorescent staining
- Serum antibodies

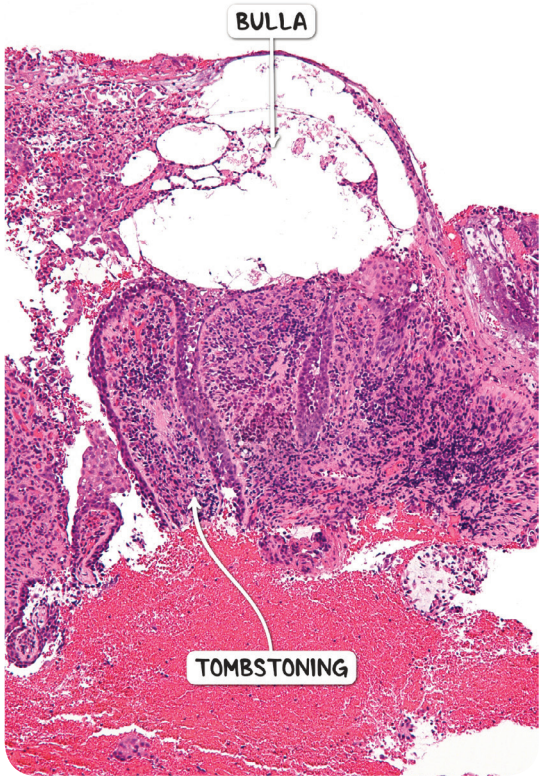

Figure 10.3 A histological section of the skin in a case of pemphigus vulgaris. There is intraepidermal bulla formation in the superficial epidermis and characteristic tombstoning of the dermoepidermal junction.

TREATMENT

MEDICATIONS

- Systemic steroids
- Immunosuppressive agents