PEMPHIGUS VULGARIS

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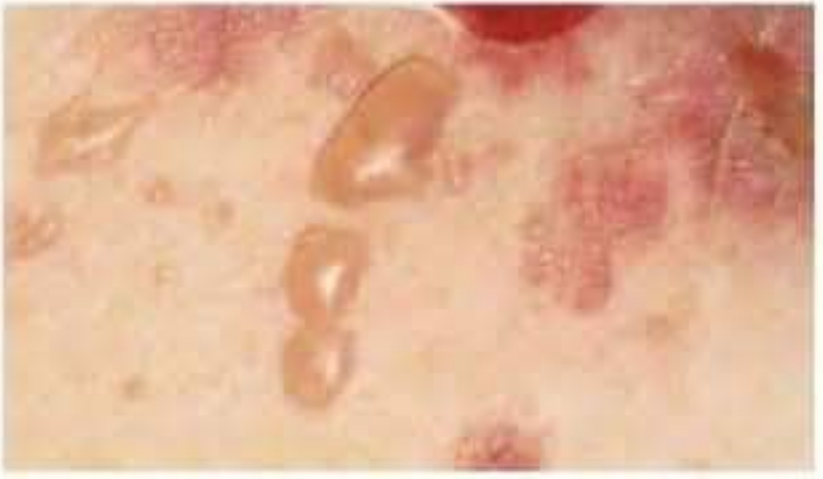
- Pemphigus is derived from the Greek word 'Pemphix' meaning bubble or a blister .

- Pemphigus describes a group of chronic bullous diseases, Originally named by 'Wichman' in 1791.

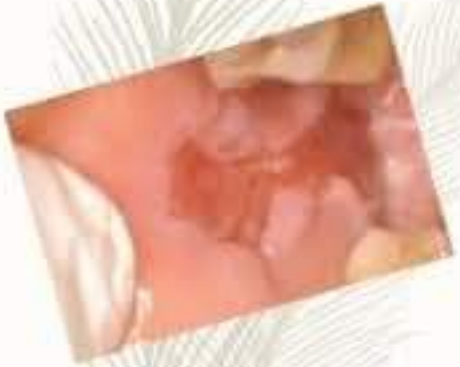
DEFINITION

it is an autoimmune, inta-epithelial, blistering disease affecting the skin and mucous membrane.



Skin blisters

Mucous membrane blisters



EPIDEMIOLOGY

INCIDENCE:

- 0.5 to 3.2 per year per 1,00,000 population
- Male: Female = 1:2
- Ashkenazi Jews and people in Mediterranean origin
- 80-90% patients develop oral lesions, 60% develop oral lesions as first symptom.
- Occasionally associated with other autoimmune disorders, Herpes simplex infection, internal malignancies

[Rai Arpita et al, 2015, ORAL PEMPHIGUS VULGARIS : A CASE REPORT]

PEMPHIGUS VULGARIS

- Accounts 70% of all pemphigus
- **Etiology:**
- **Age:** Middle age, rarely affects children
- **Sex:** Both sexes
- **Races**:Affects in eastern countries
- **Association of other autoimmune disorders**

  Rheumatoid arthritis, Myasthenia gravis, Lupus erythematosus, Pernicious anaemia
- **Drugs:** Penicillamine & Captopril

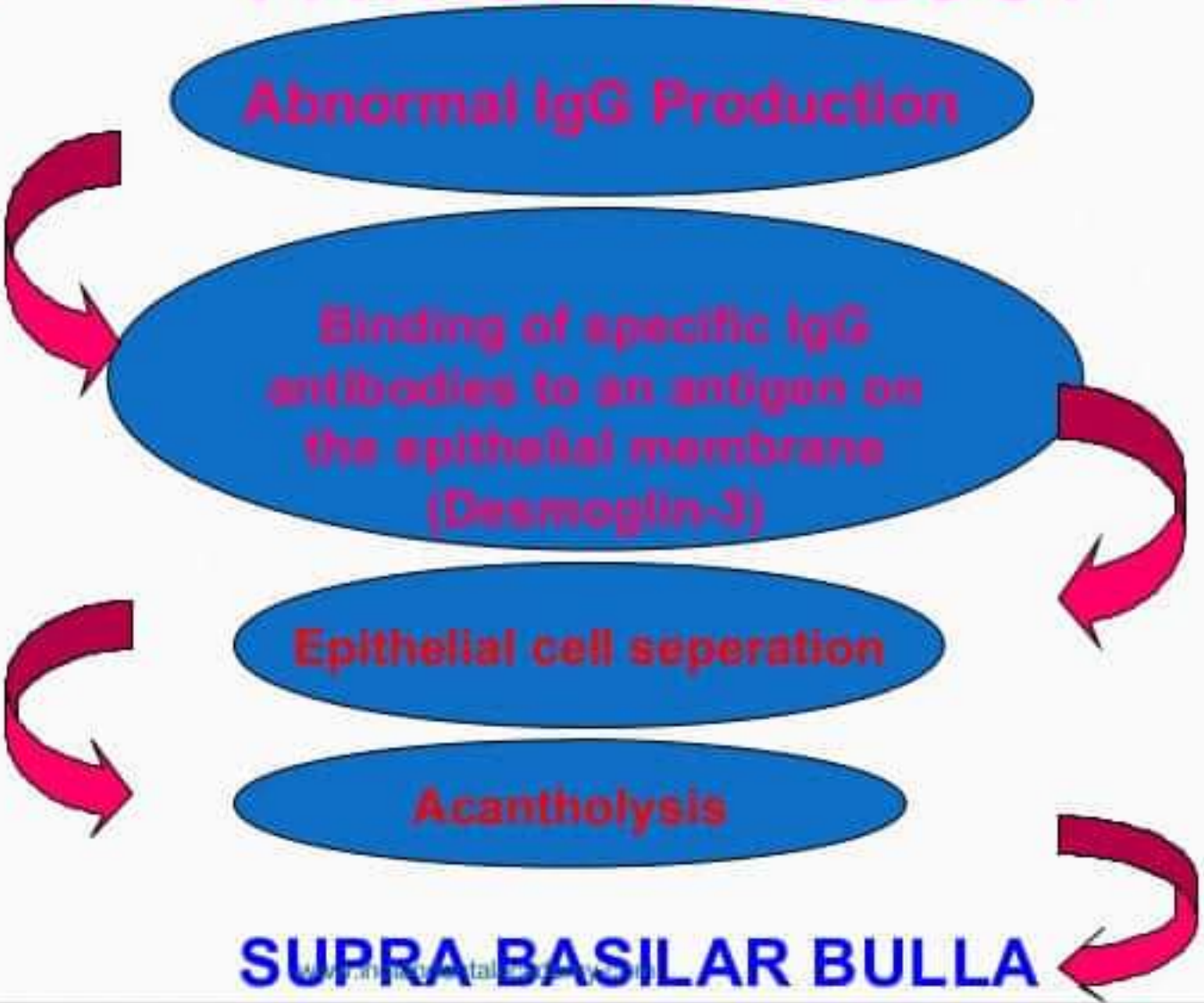
Pemphigus vulgaris

- Etiology
  - Genetic predisposition: HLA-DRB1*0402, -DQB1*0503.
  - Ab against desmoglein 3 (Dsg3) and later desmoglein 1 (Dsg1)
    - The bound Ab activate proteases that damage the desmosome → Acantholysis
  - Drugs, esp. without sulfhydryl groups
    - Beta-blockers, cephalosporins, penicillin, and rifampicin

Sterry, et al., Dermatology , 2006.

DL Longo, et al., Harrison's Principle of Internal Medicine, 18th edition , 2012.

PATHOPHYSIOLOGY



Histopathology

Pemphigus Vulgaris

- The earliest changes are intercellular edema and disappearance of the intercellular bridges in the lowermost epidermis.
- Loss of coherence between epidermal cells (acantholysis) leads to the formation of clefts and then of bullae in the suprabasal zone.
- The basal cells remain attached to the dermis, producing a 'tombstone' appearance

PEMPHIGUS VULGARIS

CLINICAL FEATURES:

- Painful ulcers or bulla are formed which are fluid filled.
- They can be formed any where in the oral cavity .
- The bulla is rapidly ruptured leaving a collapsed roof of grayish membrane with a red ulcerated base.The ulcer may look like an apthous ulcer or may be large map shaped.
- Nikolsky sign is positive.

Pemphigus Vulgaris

- Diagnosis
  - Histology
  - Immunofluorescence pattern of perilesional skin
  - Indirect ımmunofluorescence (IIF) testing of serum
  - Enzyme-linked immunosorbent assay testing for anti-desmoglein (Dsg)1 and –Dsg3 autoantibodies
  - As in other autoimmune diseases, specific antibodies may be present in relatives of patients who do not manifest any signs of diasease

Diagnostic tests for Pemphigus

- Positive Nikolsky sign
- Indirect fluorescent antibody (IFA)

  -the qualitative and semi-quantitative detection of antibodies associated with pemphigus
- The direct immunofluorescence test (DIF)

  -detects the antibody deposition in the tissues
  -very reliable diagnostic test for pemphigus
  -can remain positive for several years after regression of the disease

Pemhigus vulgaris

- DIF testing is very reliable & sensitive diagnostic test for pemphigus vulgaris, in that it demonstrates lacelike IgG in the squamous intercellular/cell surface areas in upto 95% of cases, including early cases & those with very few lesions,& in upto100% cases with active disease.

- IIF shows circulating IgG autoantibody in squamous intercellular substance in 80%-90%of cases.

Types of Nikolsky's sign

- Direct Nikolsky's sign-
  - When sign is elicited in normal skin away from the blister
  - Indicates severe disease activity.
- Marginal Nikolsky's sign-
  - When sign is elicited in normal skin near blister.
- Pseudo Nikolsky's sign-
  - Shearing or tangential force cause peeling of skin which is due to necrosis of the cells in contrast to acantholysis in pemphigus. Seen in Stevens-Johnson syndrome, Toxic epidermal necrolysis, burns.

Pemphigus vulgaris

- Treatment
  - Systemic corticosteroids
    - Main cause of morbidity and mortality is corticosteroid side-effects → always combined with steroid-sparing agents
    - Screen for osteoporosis and latent tuberculosis
  - Treatment of choice.
    - Pulse of prednisolone 1 g/day q 3-4 wk
    - Plus single dose of cyclophosphamide 7.5–15 mg/kg divided into 1–2 mg/kg/day

Sterry, et al., Dermatology , 2006

Associated features and complications

- Secondary infection
- Extensive lesions associated with water and electrolyte imbalance
- Complication of steroids and immunosuppresive drugs.

NURSING DIAGNOSES

- Acute pain of skin and oral cavity related to blistering and erosions
- Impaired skin integrity related to ruptured bullae and denuded areas of the skin
- Anxiety and ineffective coping related to the appearance of the skin and no hope of a cure
- Deficient knowledge about medications and side effects