Pemphigus foliaceus

-DR.CH.VIJAY BHASKER REDDY MD

SENIOR RESIDENT

DEPARTMENT OF DVL

Pemphigus is a group of **autoimmune** blistering diseasesof the skin and mucous membranes that is characterized by:

----- histologically, **intraepidermal blisters** due to the loss of cell-cell adhesion of keratinocytes

----- immunopathologically, the finding of *in vivo* bound and circulating **IgG** autoantibodies directed against the cell surface of keratinocytes

Classification

Pemphigus vulgaris

-P.vegetans…….Hallopeau

…….Neumann

Pemphigus foliaceus

-P.erythematosus

-Fogo selvagem

Para neoplastic pemphigus

Drug induced pemphigus

Ig A pemphigus

- Patients with pemphigus vulgaris and pemphigus foliaceus have IgG autoantibodies against desmoglein 3 and desmoglein 1,respectively, while patients with para neoplastic pemphigus have IgG autoantibodies against plakin molecules in addition to autoantibodies against desmogleins

- IgA autoantibodies directed against the keratinocyte cell surface define IgA pemphigus

Pemphigus vulgaris:

CLINICAL FEATURES:

pemphigus vulgaris develop painful erosions of the oral mucosa.

More than half of the patients also develop flaccid blisters and widespread cutaneous erosions.

Pemphigus vulgaris divided into two subgroups:

(1) *mucosal dominant* with mucosal erosions but minimal skin involvement;

(2) *mucocutaneous* type with extensive skin blisters and erosions in addition to mucosal involvement

.

Mucous membrane lesions:

- may be seen anywhere in the oral cavity, scattered or extensive erosions
- most common sites are the buccal and palatine mucosa.

- painful erosions
- Intact blisters are rare and fragile and break easily

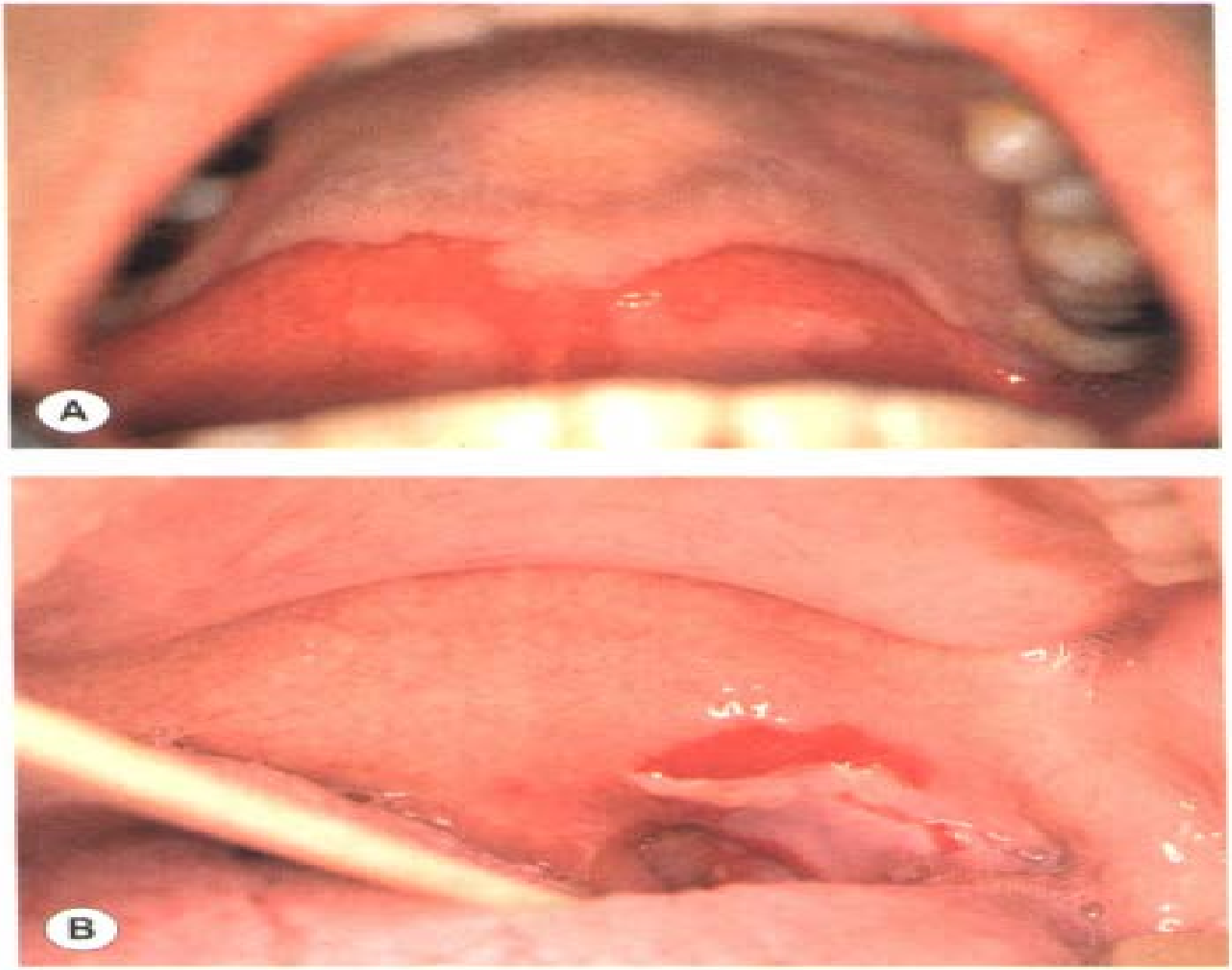
erosions are of different sizes with an irregular ill-defined border

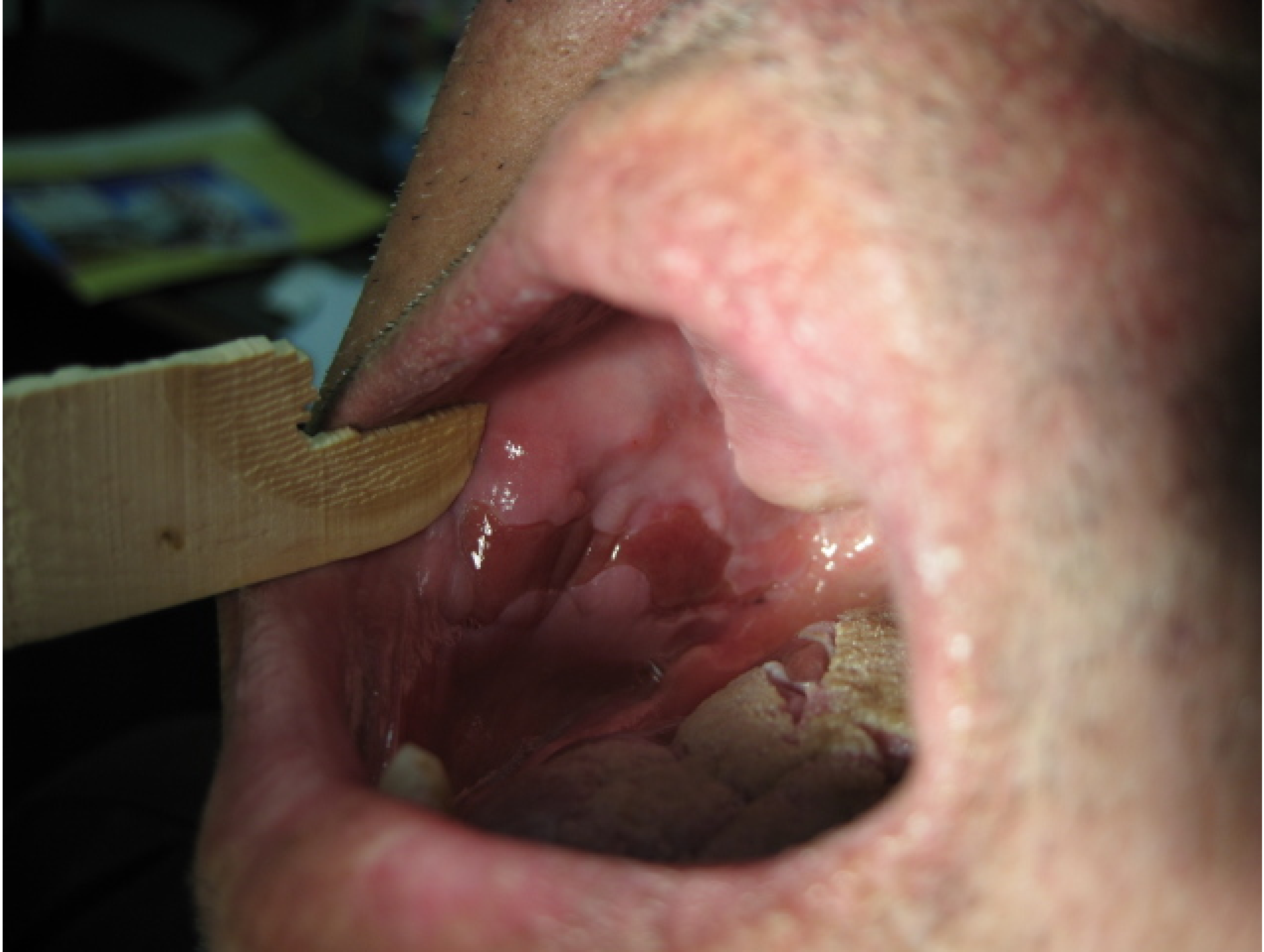
Involvement of the throat produces hoarseness and difficulty in swallowing.

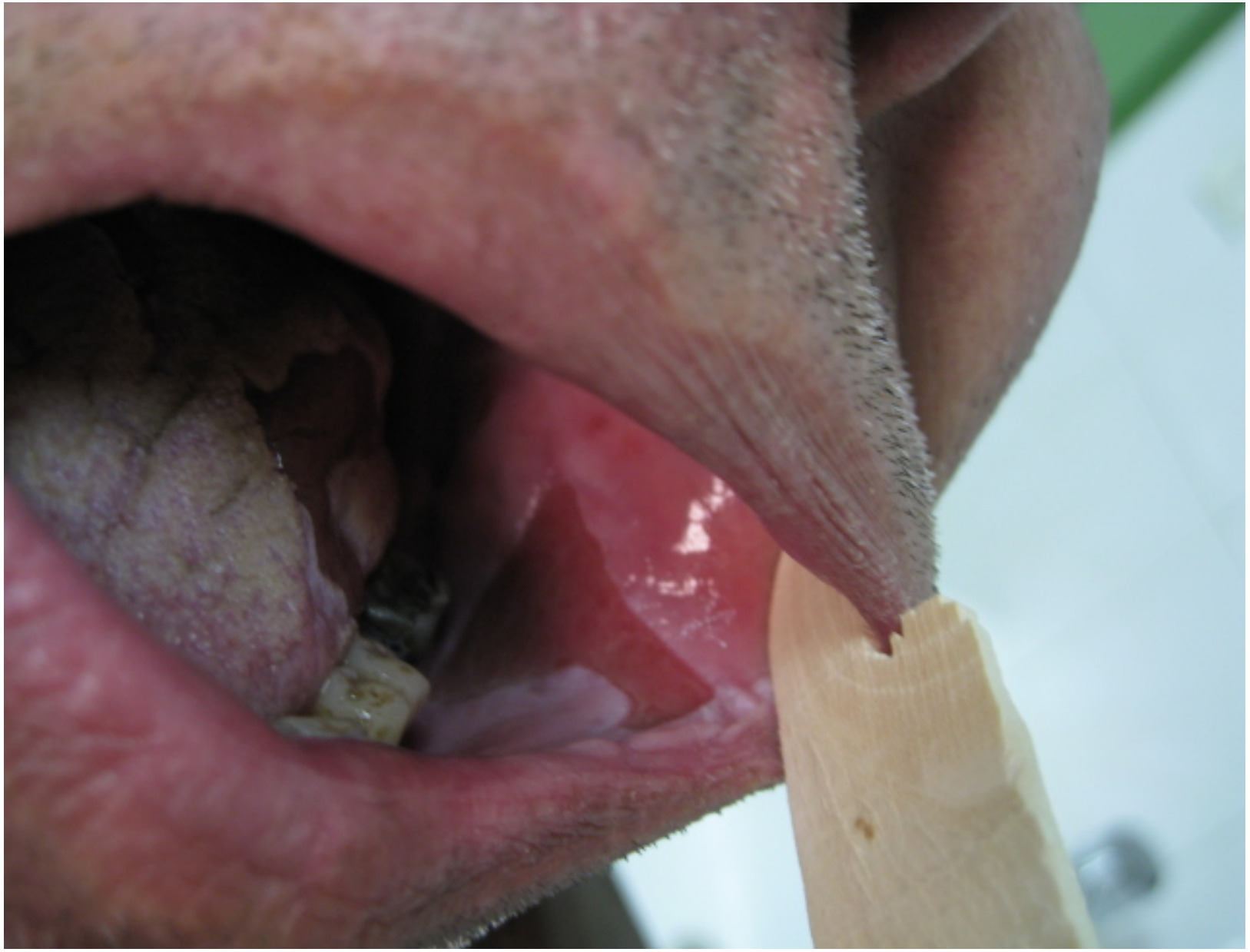
The esophagus also may be involved

The conjunctivae, nasal mucosa, vagina, penis, anus and labia can

develop lesions as well.


A
B

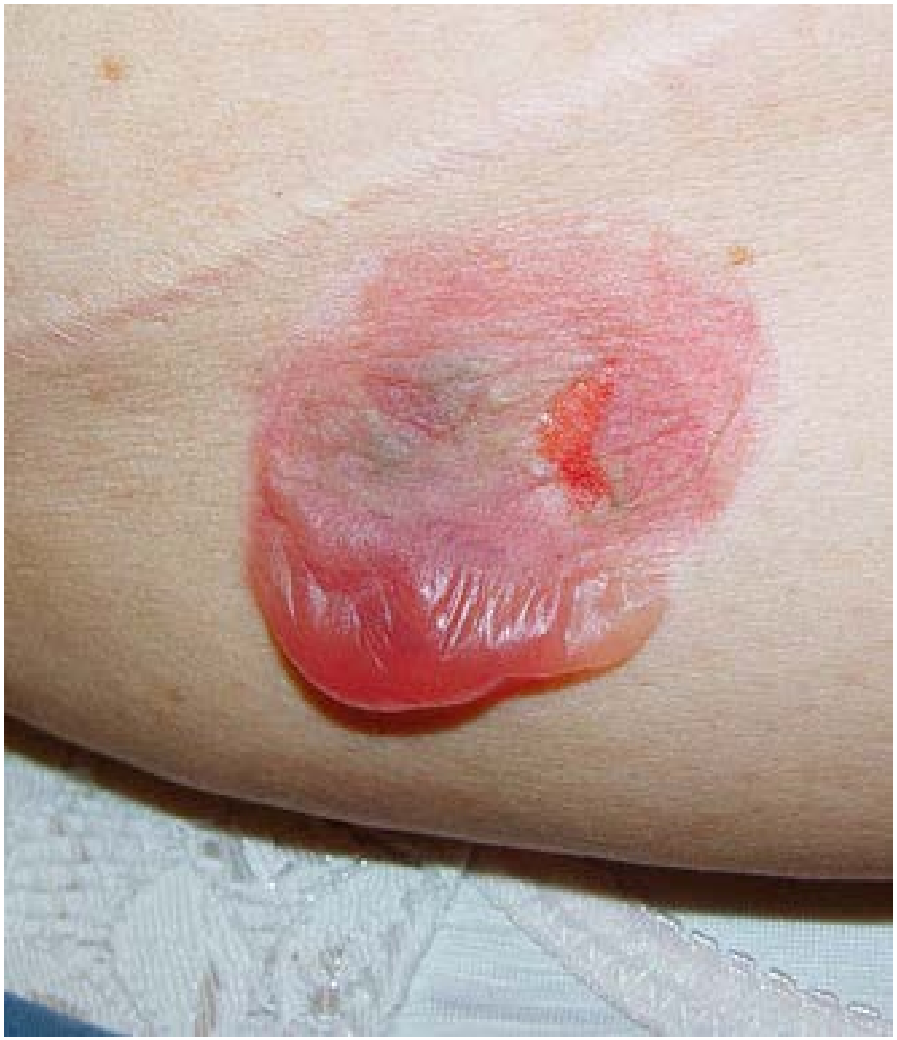


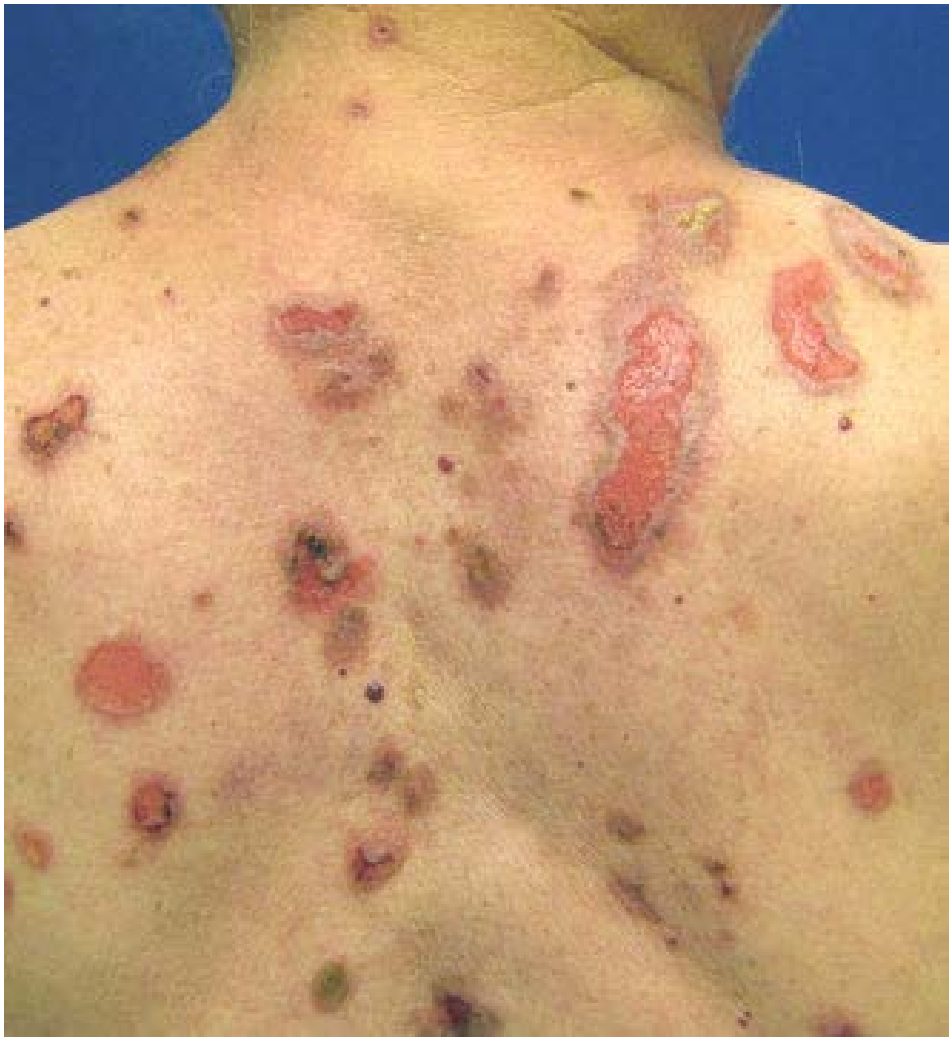


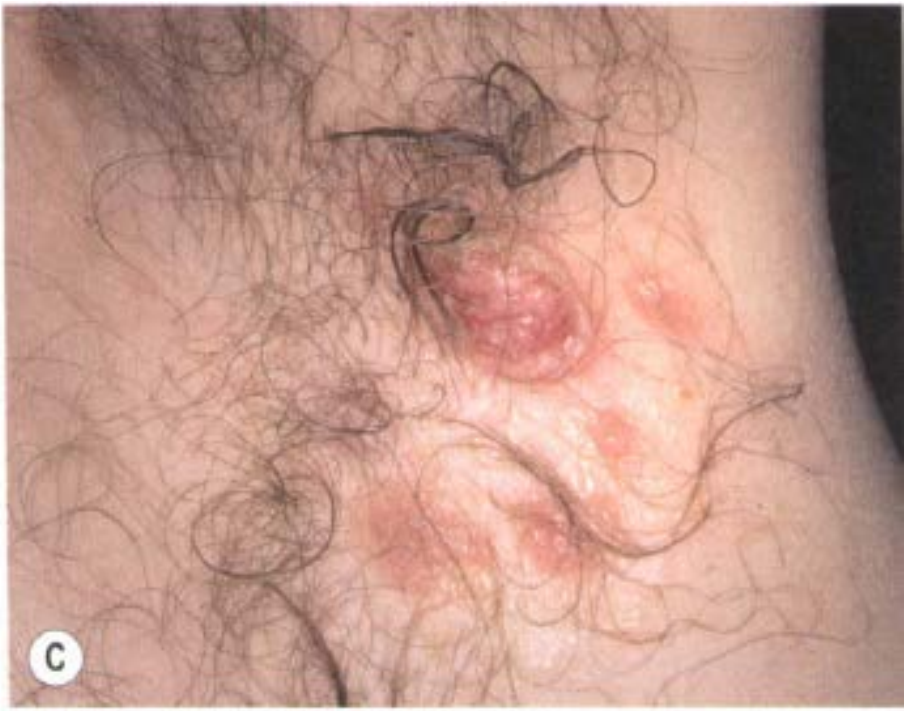
The primary skin lesions of pemphibus vulgaris are flaccid, thin walled, easily ruptured blisters

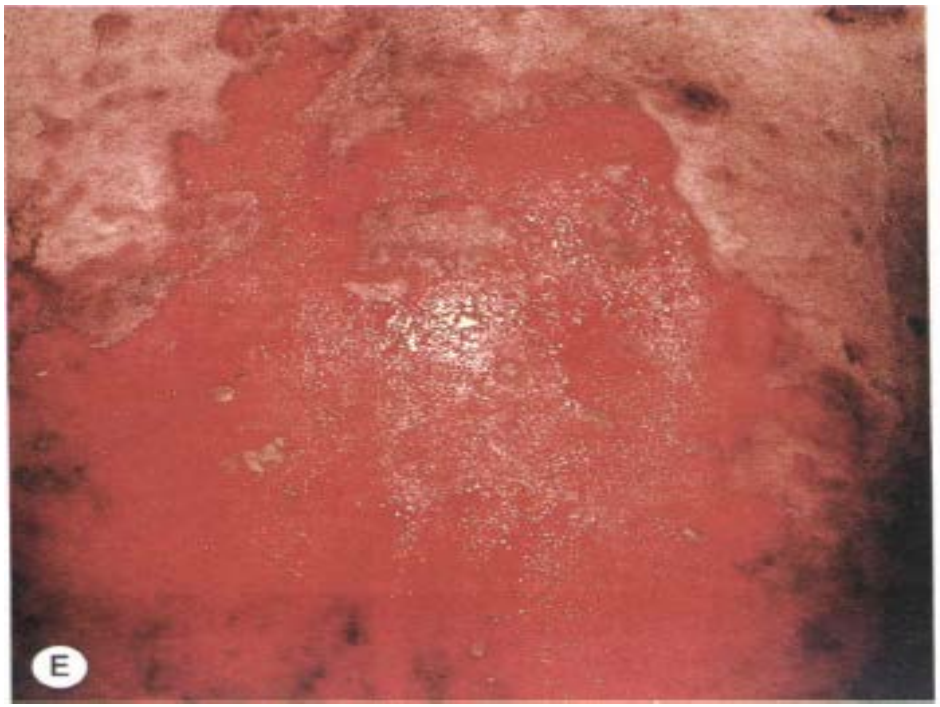
The fluid within the bullae is initially clear but may become hemorrhagic, turbid, or even seropurulent

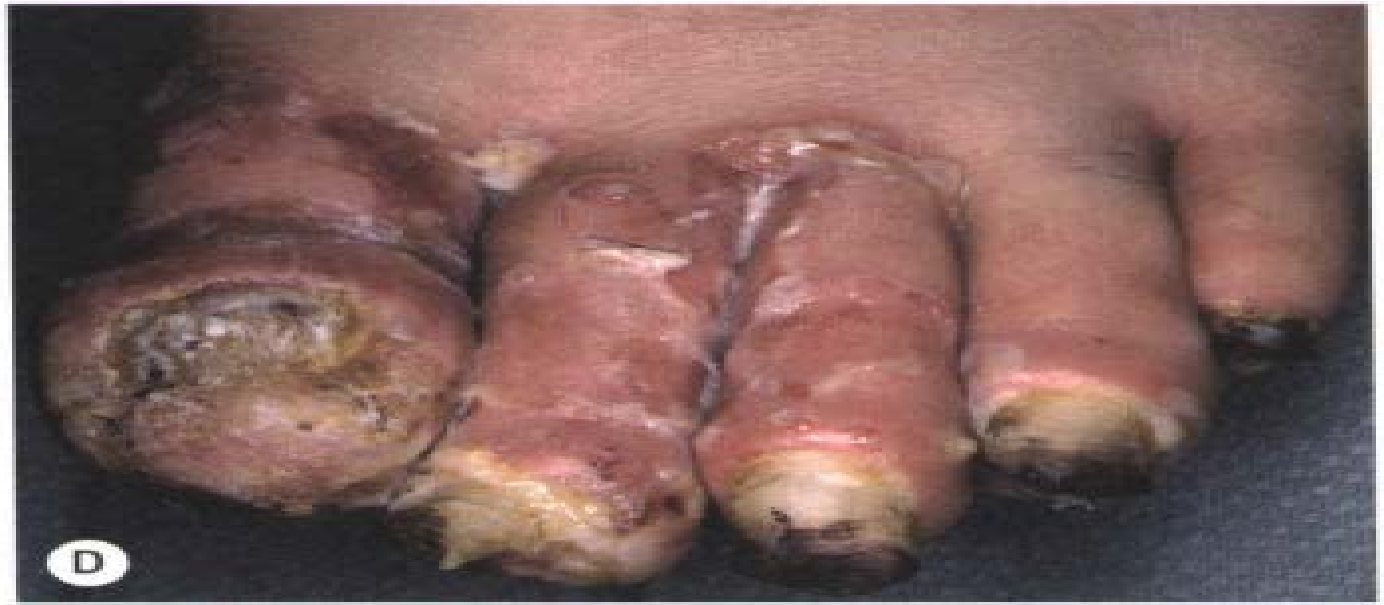
These erosions often attain a large size and can become generalized.






C


E


D

Those lesions that do heal often leave hyperpigmented patches with no scarring.

Associated pruritus is uncommon.

Pemphigus Vegetans:

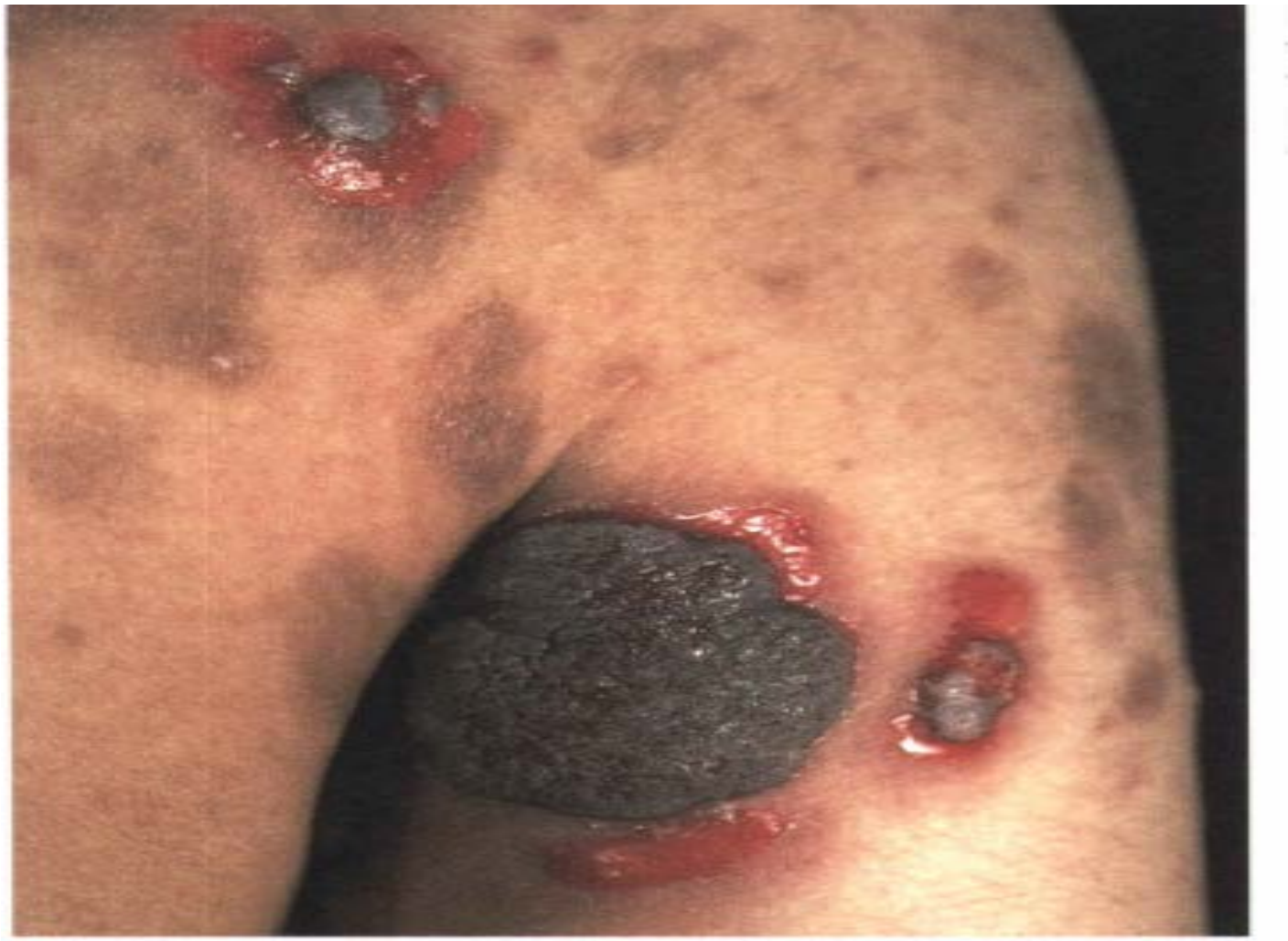
is a rare variant of pemphigus vulgaris

represent a reactive pattern

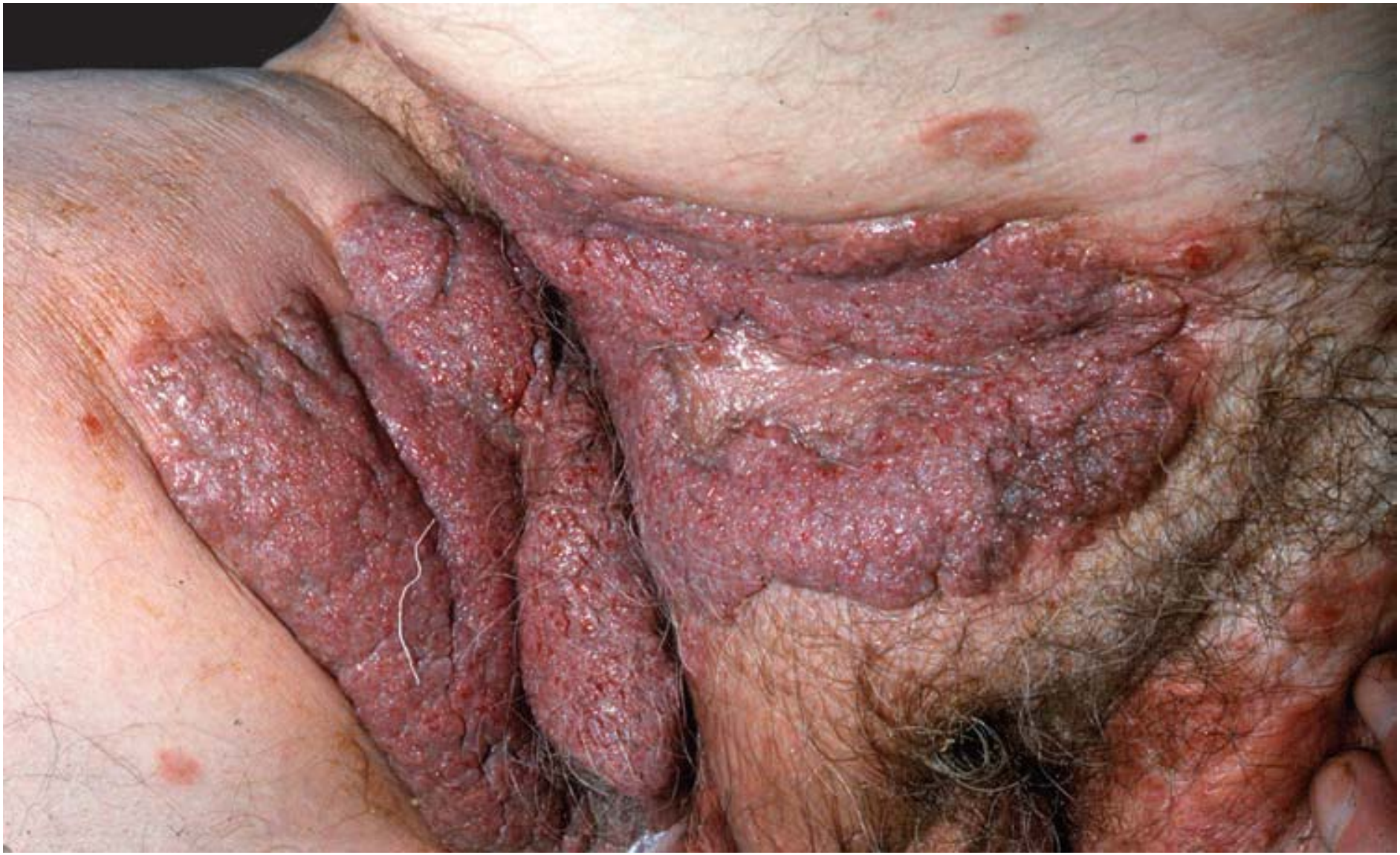
characterized by flaccid blisters that become erosions and then form fungoid vegetations or papillomatous proliferations, soon progress to vegetative plaques.

especially in intertriginous areas and on the scalp or face

especially in intertriginous areas and on the scalp or face



Papillomatous, cauliflower-like, oozing growths in the groin and pubis



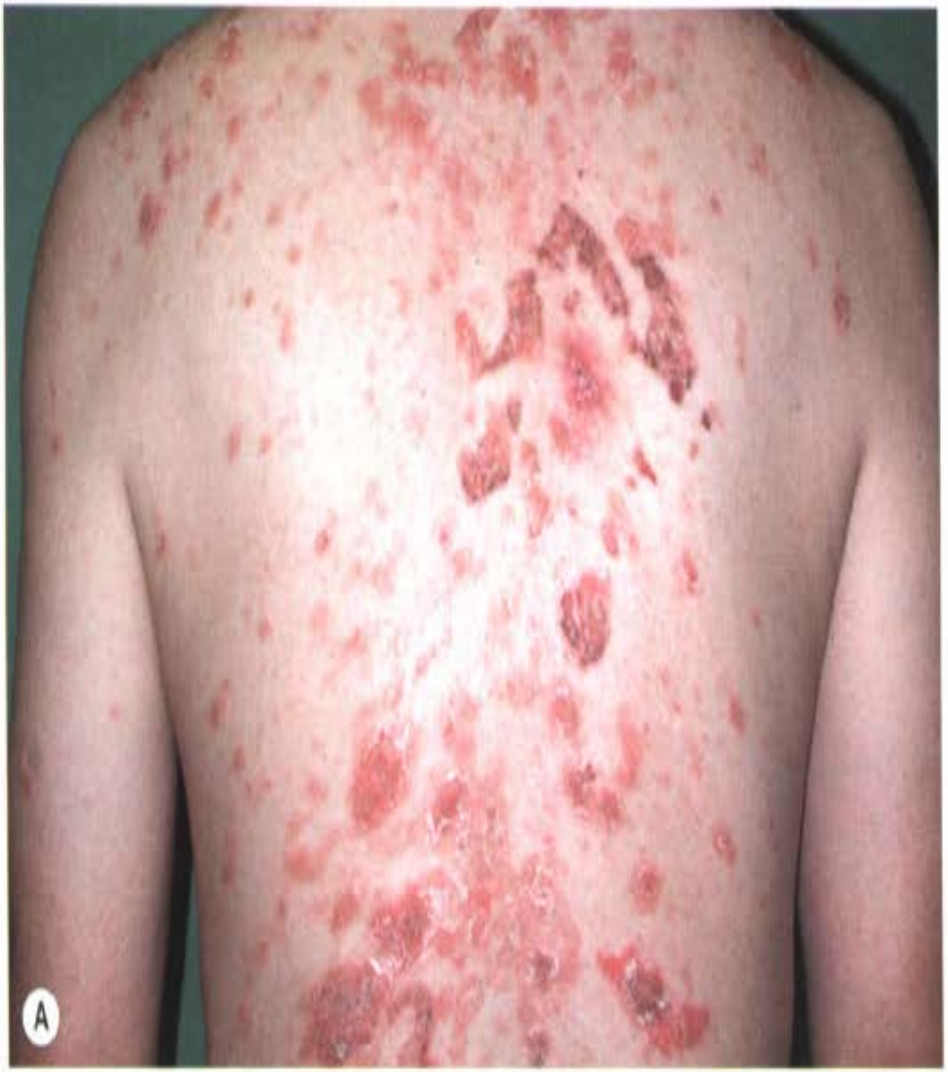
Pemphigus Foliaceus

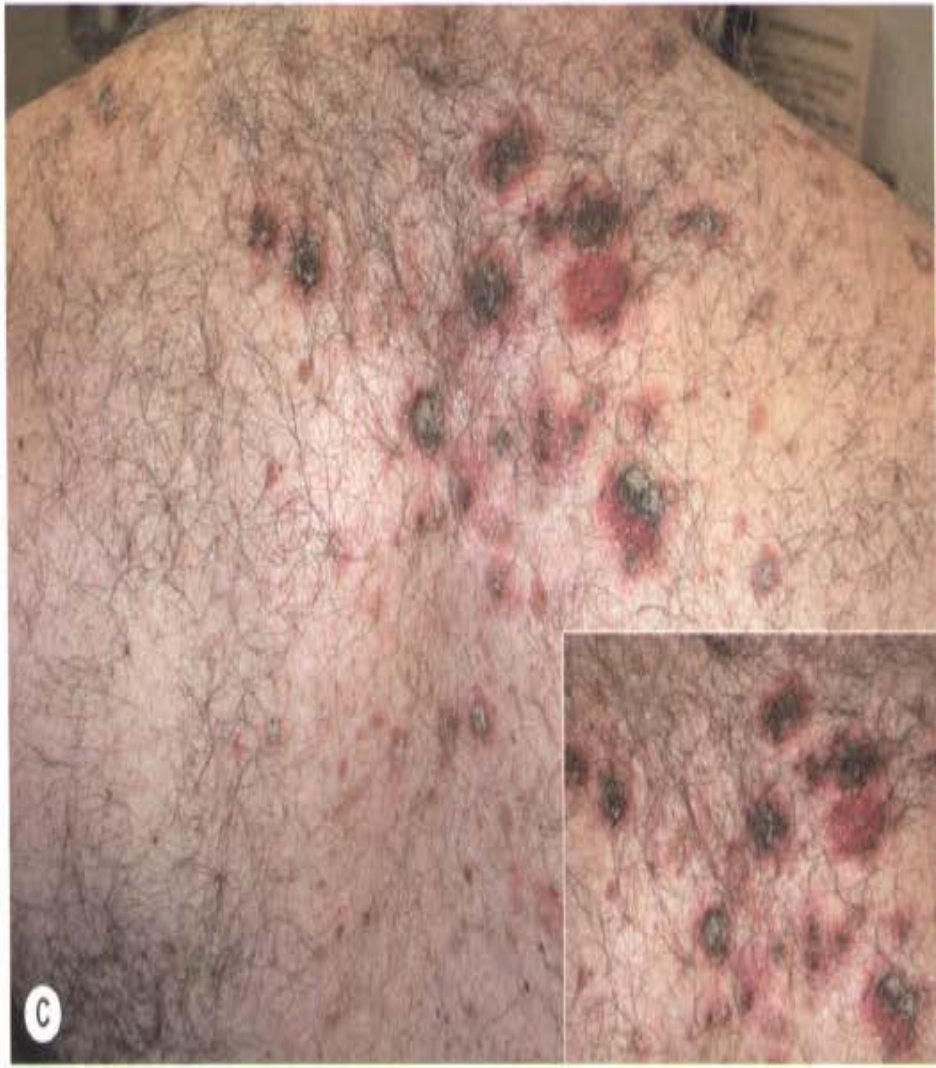
-They complain of burning and pain in association with the skin lesions.

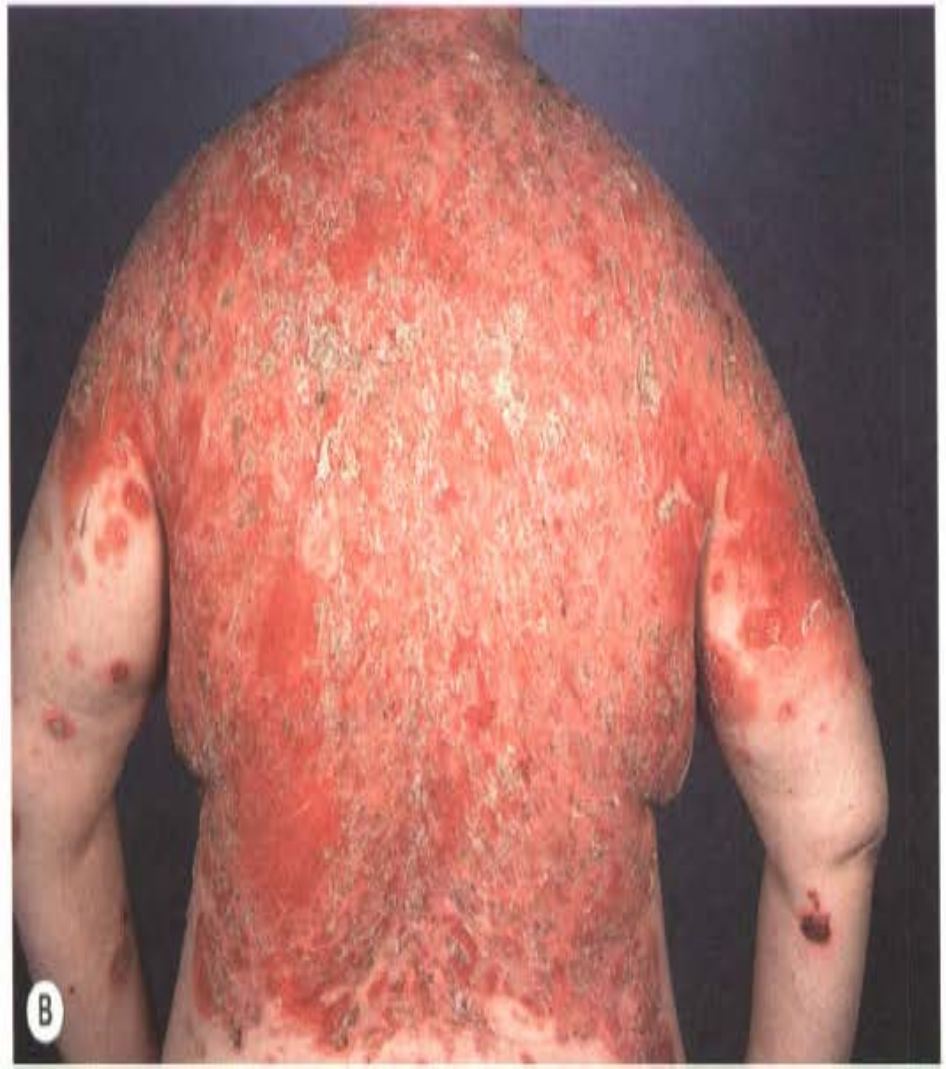
-develop scaly, crusted cutaneous erosions, often on an erythematous base,

- do not have clinically apparent mucosal involvement even with widespread disease

- The onset of disease is often subtle, with a few scattered crusted lesions that are transient


A


C


B


D

They are usually well demarcated and have a seborrheic distribution,

they favor the face, scalp and upper trunk

- the vesicle is so superficial and fragile, often only the resultant crust and scale are seen

The disease may

- *stay localized for years*

  or

- *it may rapidly progress, in some cases to generalized involvcment and an exfoliative erythroderma*

- oral lesions is extremely rare

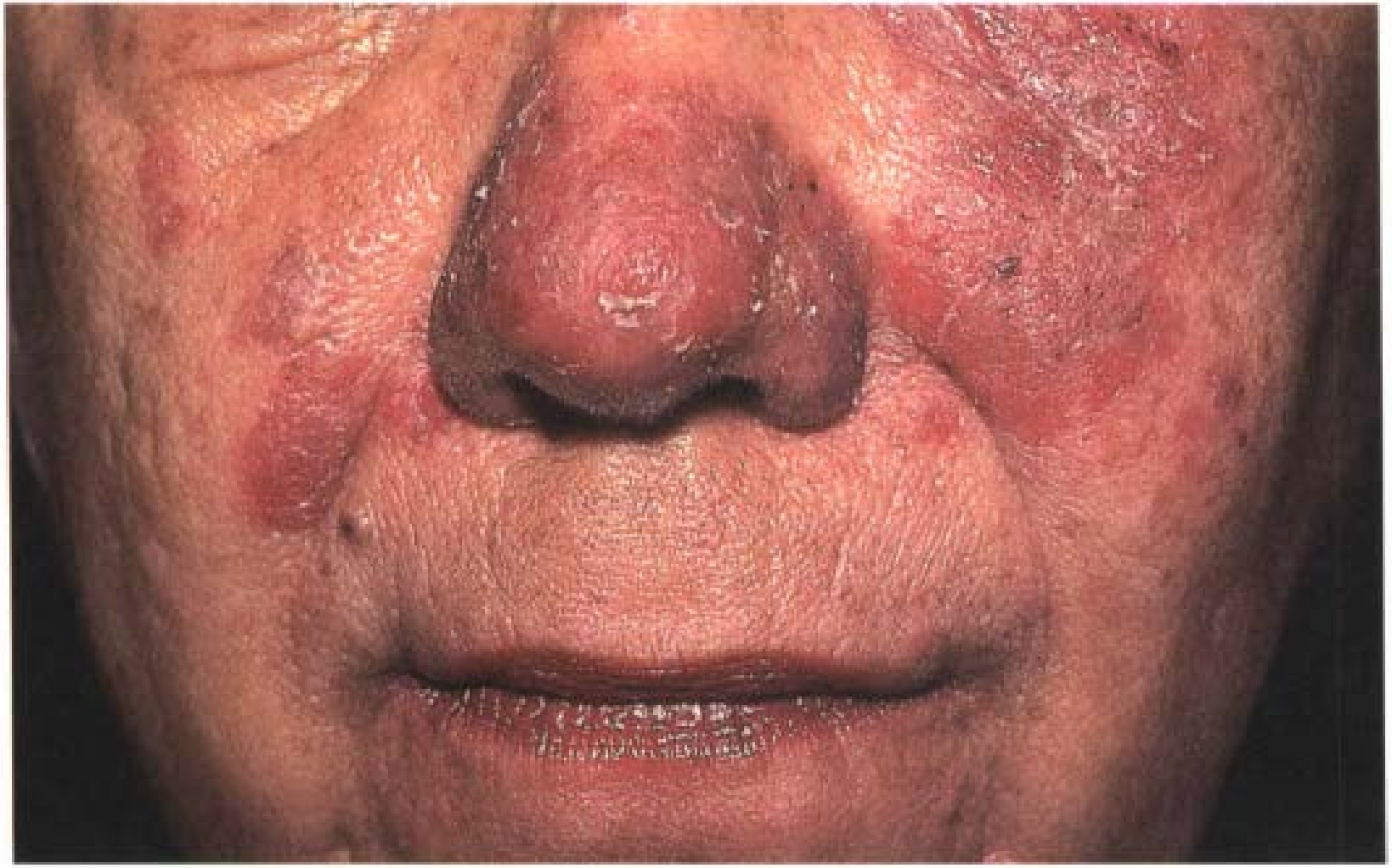
Pemphigus Erythematosus
(Senear-Usher Syndrome)

- localized variant of pemphigus foliaceus
- In the malar region of the face
- typical scaly and crusted lesions of pemphigus foliaceus

patients with immunologic features of:

- **both lupus erythematosus and pemphigus**
- **IgG and C3 deposition on cell surfaces of keratinocytes as well as the basement membrane zone**
- **circulating antinuclear antibodies'**

Senear-Usher Syndrome



Paraneoplastic Pemphigus

with underlying neoplasms,both malignant and benign.

The most commonly associated neoplasms are:

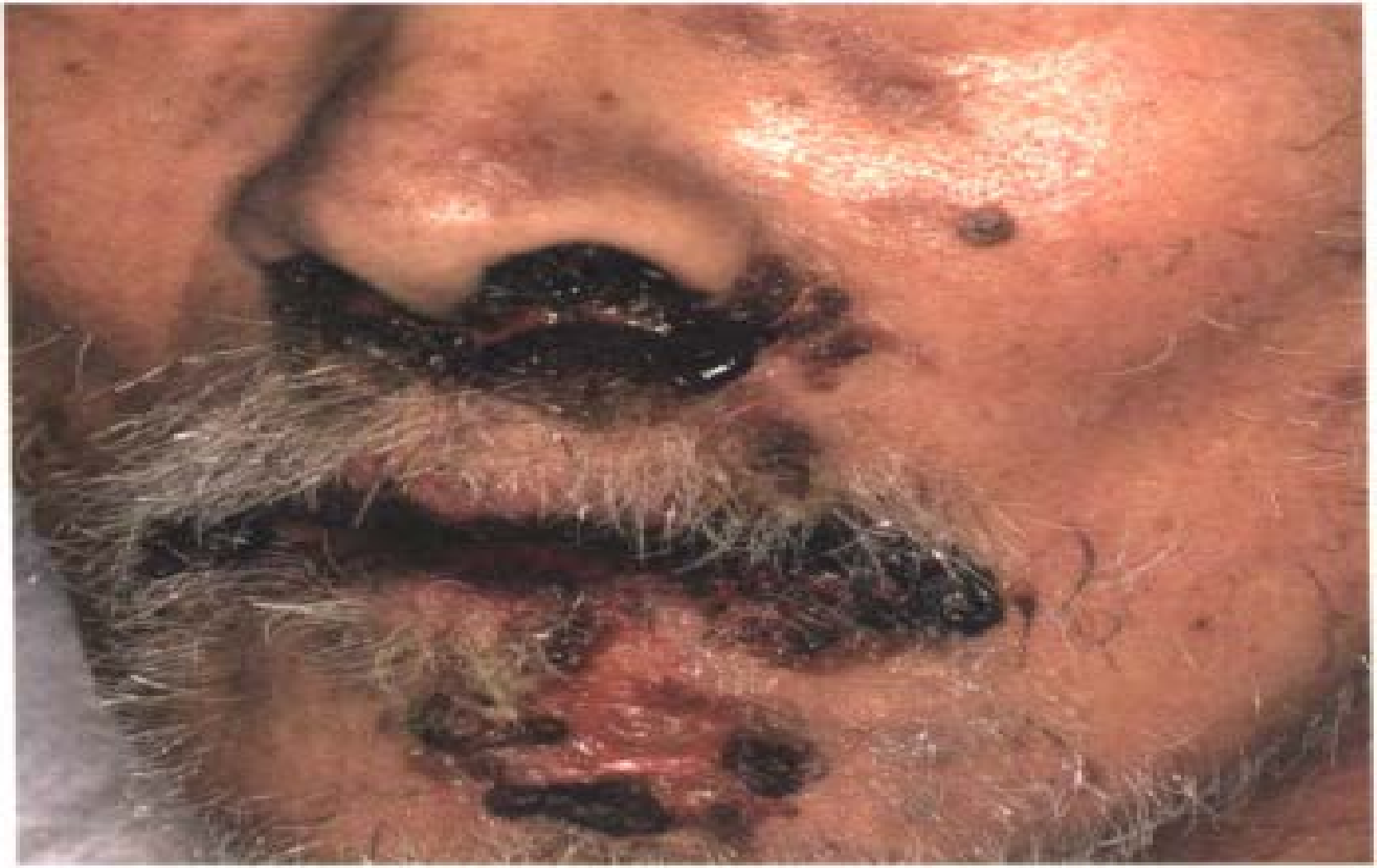
- non-Hodgkin lymphoma (40)
- chronic lymphocytic leukemia(30%),
- Castleman's disease (10%),
- malignant and benign thymomas(6%), sarcomas (6%) and Waldenstrom's macroglobulinemia (6%)

The most constant clinical feature of paraneoplastic pemphigus is the presence of intractable stomatitis.

This stomatitis consists of erosions and ulcerations that affect all surfaces of the oropharynx and characteristically extend onto the vermilion lip

Cutaneous findings are polymorphic:

- may present as erythematous macules,
- flaccid blisters and erosions resembling pemphigus vulgaris,
- tense blisters resembling bullous pemphigoid,
- erythema multiforme-like lesions
- lichenoid eruptions

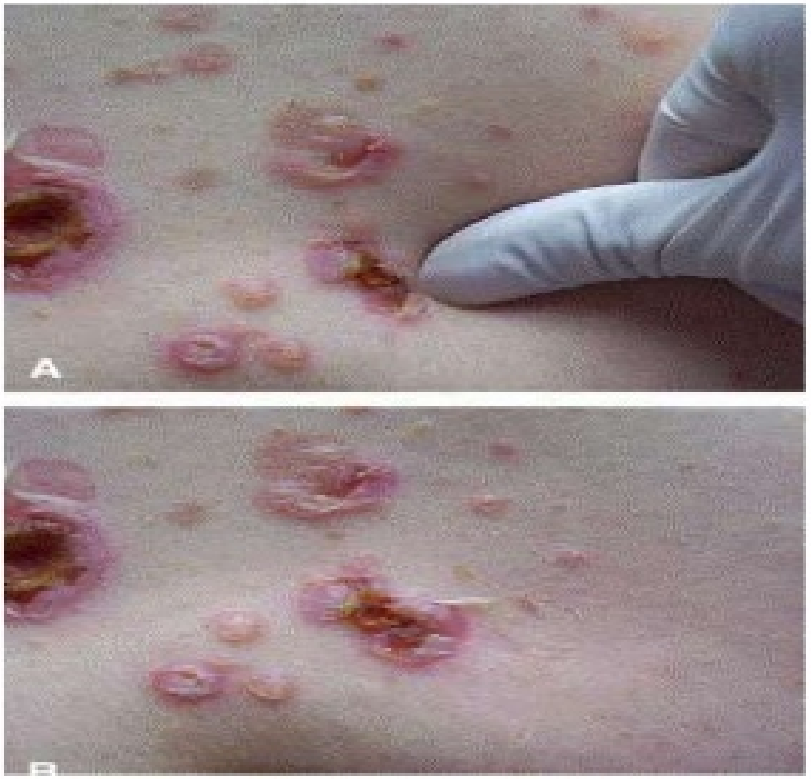


Diagnosis

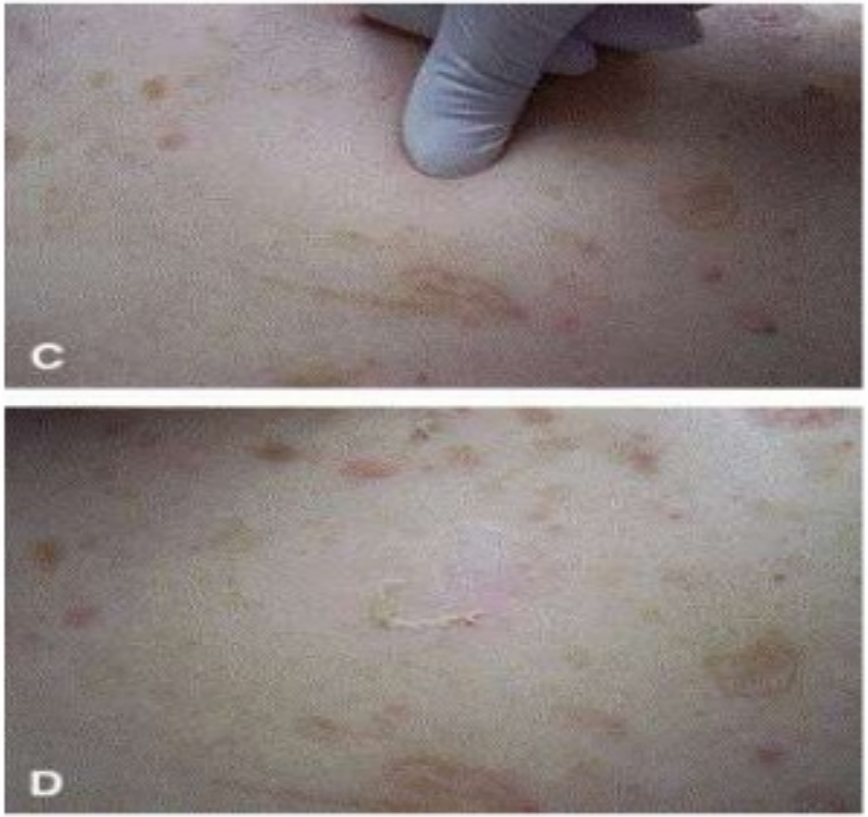
- h/o and clinical examination
- Tzanck test
- Skin biopsy
- **Direct immunofluorescence……..gold standard**

Variants of nikolsky's sign

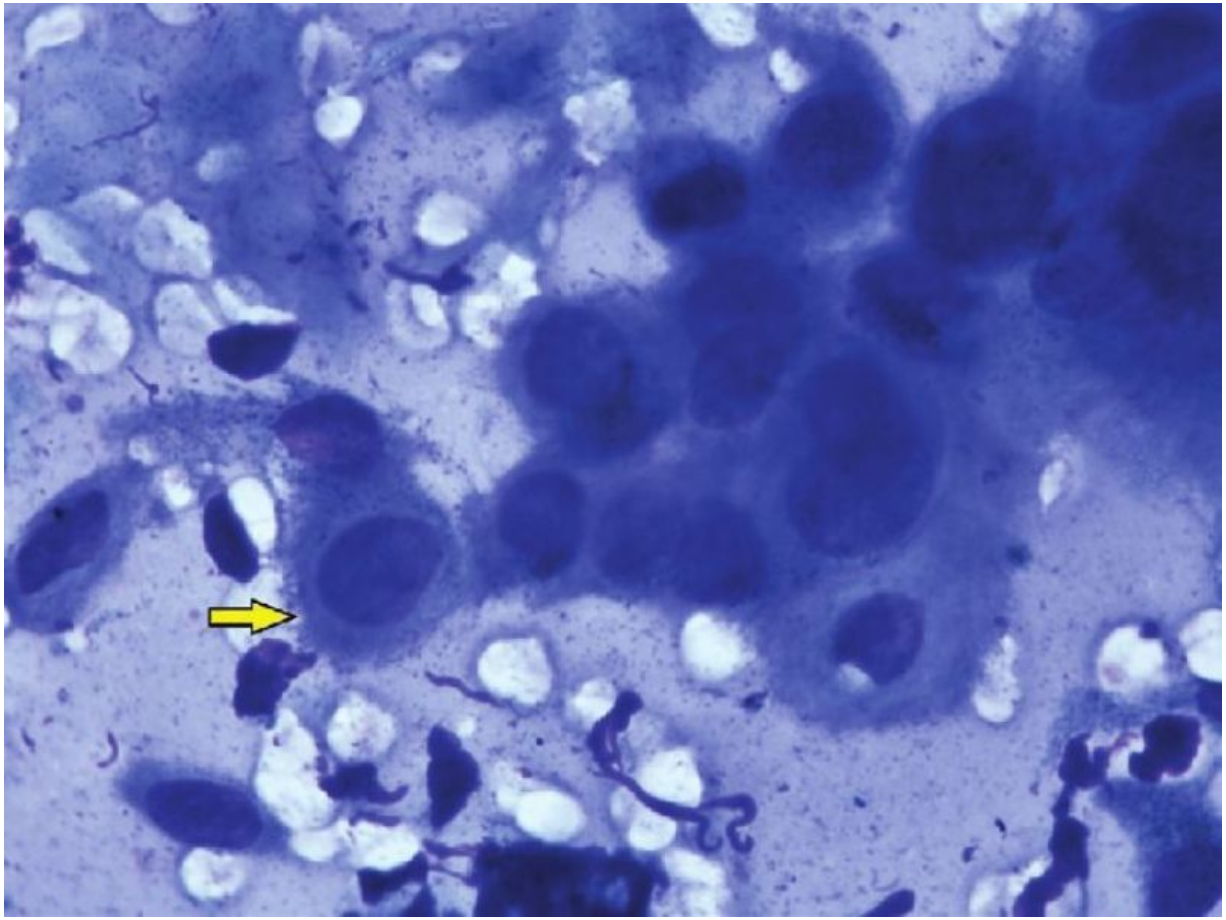
- **Marginal nikolsky:** nikolsky sign elicited over the normal looking skin of a pemphigus patient close to existing lesions.



- **Direct nikolsky sign:** nikolsky sign elicited over the normal looking skin at a distant site.
- Positive direct nikolsky sign indicates severe activity of the disease in pemphigus.



Tzanck test



Skin biopsy

DIF

Indirect IF

TREATMENT

- **STEROIDS**…..topical/intralesional
  ……prednisolone 1-2 mg /kg body wt
- **STEROID SPARING AGENTS**

CYCLOPHOSPHAMIDE

AZATHIOPRINE

HCQS

NICOTINAMIDE/TETRACYCLINE/MINOCYCLINE

DAPSONE

RETINOIDS

MMF

IVIG

- The use of a steroid sparing agent reduces the side effects encountered with systemic corticosteroids but care needs to be taken to monitor for systemic toxicity of the steroid sparing agents themselves including the increased risks of infection and malignancy.
- Probably the lease toxic steroid sparing regimen is the use of tetracycline 2g/day and nicotinamide 2g/day.

- Cyclophosphamide and azathioprine are the drugs most commonly used as a steroid sparing agents in pemphigus subtypes. Cyclophosphamide has significant potential systemic toxicity and patients must be carefully monitored during treatment which usually is a single daily dose of 1-2 mg/kg.
- Azathioprine in general has less systemic toxicity than cyclophosphamide but it works more slowly. It is given at a dose of 1-3 mg/kg/day.
- Other steroid sparing modalities for pemphigus which are sometimes effective include cyclosporin, chlorambucil and plasmapheresis.

Plasmapharesis:

in conjunction with glucocorticoids and immunosuppressive agents in poorly controlled patients, in the initial phases of treatment to reduce antibody titers

PROGNOSIS

- Benign
- Rarely, chronic course with repeated relapses like p. vulgaris
- Clinical and immunopathological evolution into p.vulgaris

- **THANK YOU**