



Autoimmune Dermatologic Diseases

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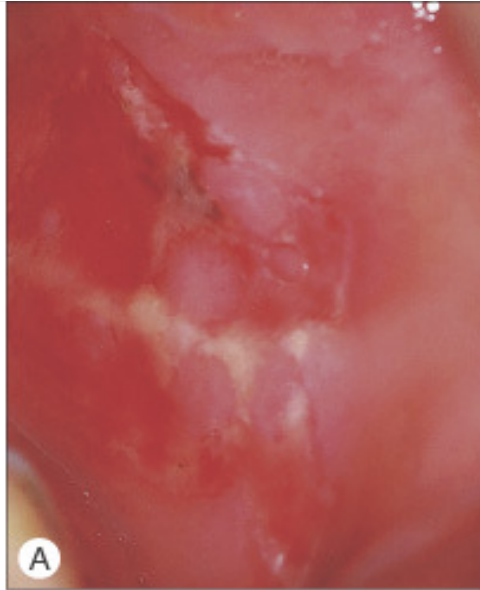
DERMATOLOGY AND PLASTIC SURGERY OF
ARIZONA

Pemphigus Vulgaris

- ▶ Epidemiology: M=F, usually 5th and 6th decades, rare in young persons
- ▶ Etiology: autoimmune blistering disease mediated by intercellular antibodies
- ▶ IgG throughout epidermis, C3 also found
- ▶ **Desmoglein-3** = mucosal involvement
- ▶ **Desmoglein-1** = cutaneous involvement
- ▶ Both = mucocutaneous involvement
- ▶ IIF uses **monkey esophagus epithelium**
- ▶ Punch Biopsy peri-lesional skin- H&E and **DIF (Michel's Media)**

Pemphigus Vulgaris

- ▶ Mouth lesions first appear 60% of the time
- ▶ Mucosa with painful erosion
- ▶ Mouth odor is offensive
- ▶ Hoarseness, difficulty swallowing with throat involvement
- ▶ Esophagus may be involved and sloughing of entire lining to form a cast (esophagitis dissecans superficialis), even with well controlled cutaneous disease



Pemphigus Vulgaris

- ▶ Fragile bullae
- ▶ Bulla is clear at first but may become hemorrhagic or even seropurulent then form erosions
- ▶ Appear first in the mouth (60%) and then commonly in the groin, scalp, face, neck, axillae, or genitals
- ▶ Positive Nikolsky, Asboe-Hansen signs

Nikolsky sign vs. Asboe-Hansen

- ▶ **Nikolsky** sign: absence of cohesion in the epidermis, upper layers are easily made to slip laterally by slight pressure or rubbing.
- ▶ **Asboe-Hansen** sign: direct pressure on intact bulla leading to bulla-spread phenomenon

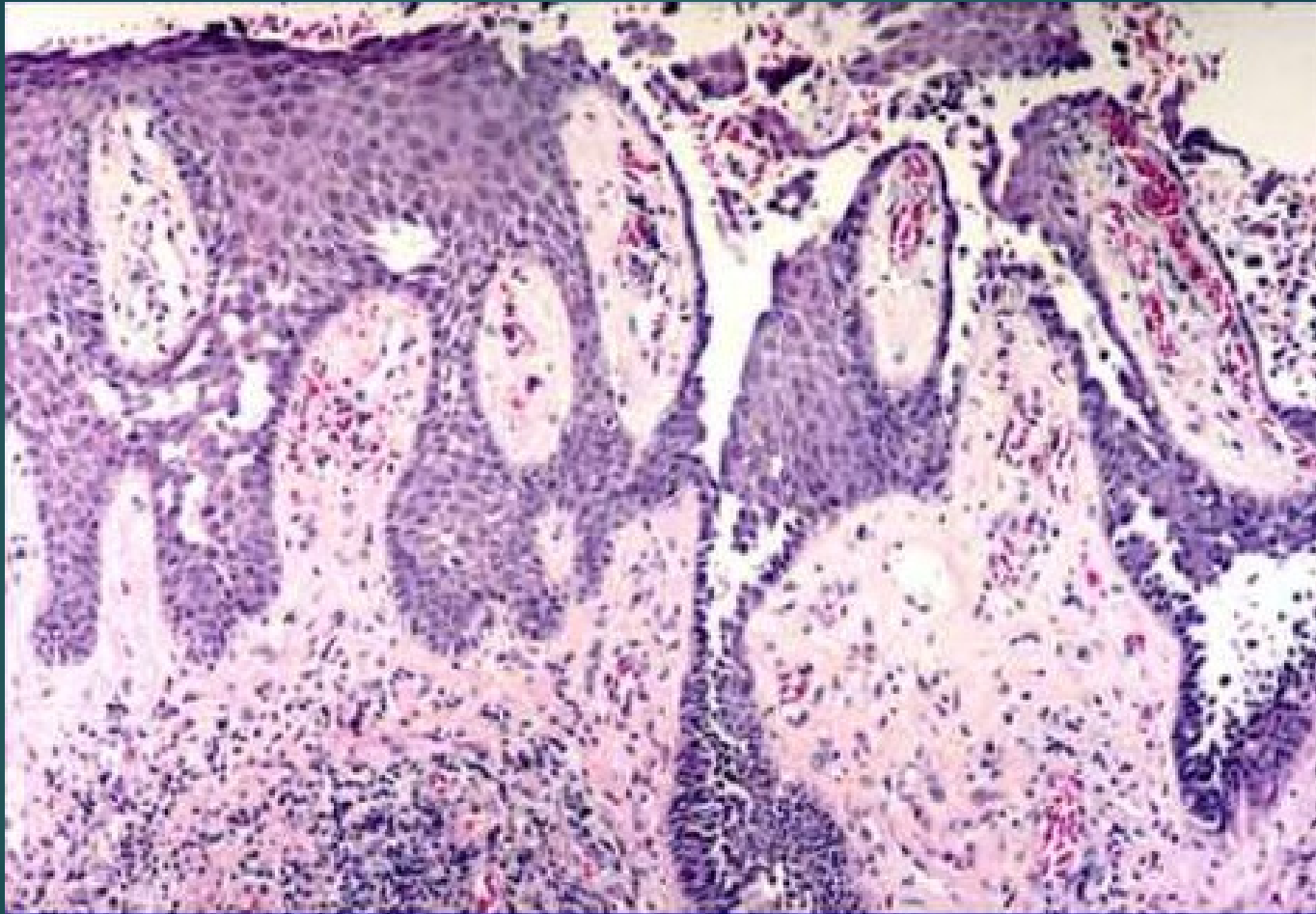
Table 31.2 Target antigens in pemphigus.

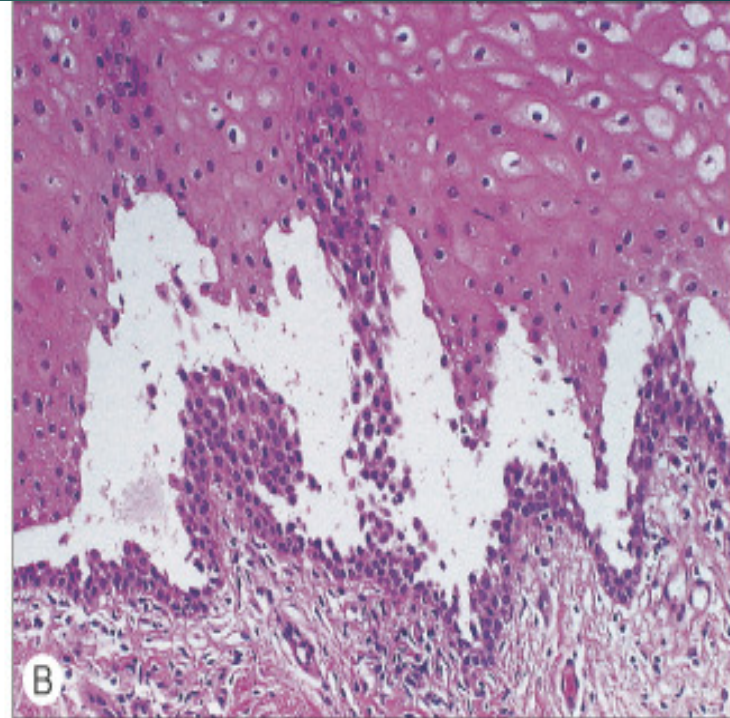
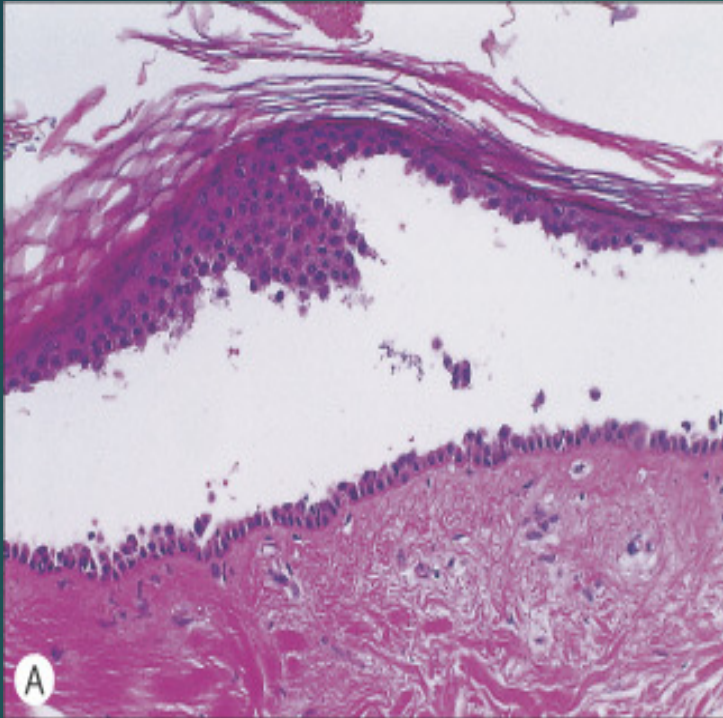
TARGET ANTIGENS IN PEMPHIGUS			
Diseases	Autoantibodies	Antigens	MW (kDa)
Pemphigus vulgaris Mucosal dominant type Mucocutaneous type	IgG	desmoglein 3	130
	IgG	desmoglein 3	130
		desmoglein 1	160
Pemphigus foliaceus	IgG	desmoglein 1	160
Paraneoplastic pemphigus	IgG	desmoglein 3	130
		desmoglein 1	160
		plectin	500
		desmoplakin I	250
		desmoplakin II	210
		BPAG1	230
		envoplakin	210
		periplakin	190
?	170		
Drug-induced pemphigus	IgG	desmoglein 3	130
		desmoglein 1	160
IgA pemphigus SPD type IEN type	IgA IgA	desmocollin 1	110/100
		?	?

BPAG1, bullous pemphigoid antigen

Pemphigus Vulgaris

- ▶ Associated conditions: **myasthenia gravis** or **thymoma**
- ▶ Drugs which induce pemphigus
 - ▶ **Penicillamine** for treatment of RA, most often seen in foliaceus type
 - ▶ Captopril, penicillin, thiopronine, interleukin-2, nifedipine, piroxicam, rifampin

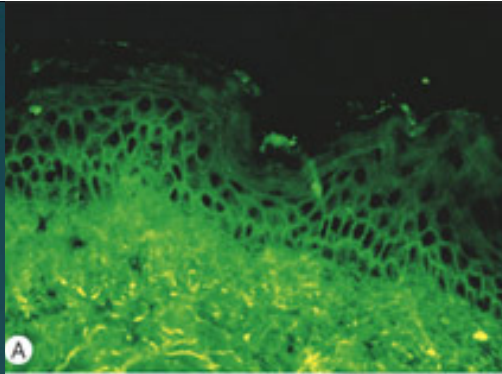




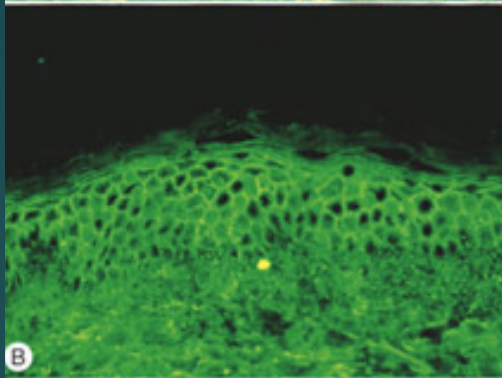
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Tombstone rows

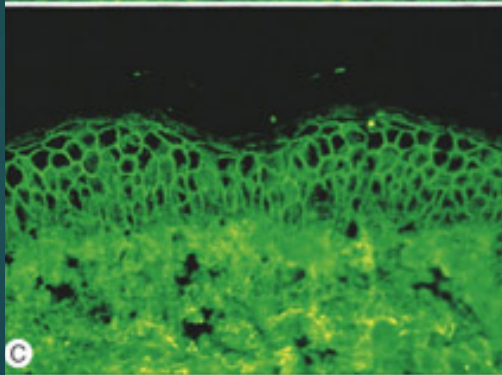
Pemphigus- IgG autoantibodies directed against the cell surface of keratinocytes.



A. Pemphigus_vulgaris: Sera containing anti-desmoglein 3 IgG alone stain the cell surfaces in the lower epidermis.



B. Pemphigus_vulgaris: Sera containing both anti-desmoglein 3 IgG and anti-desmoglein 1 IgG stain the cell surfaces throughout the epidermis.



C Pemphigus_foliaceus: Sera containing anti-desmoglein 1 IgG alone stain the cell surfaces throughout the epidermis, but more intensely in the superficial layers.

Pemphigus Vulgaris

▶ Treatment:

- ▶ Topical: Silvadene, Maalox for mouth
- ▶ Systemic: Prednisone 60mg to 100mg daily (1 mg/kg/day)
- ▶ Systemic: Azathioprine, mycophenolate mofetil, cyclophosphamide, methotrexate
- ▶ Repeat pemphigus antibody titer in 4-8 weeks after treatment. If not improving, increase prednisone up to 150mg/day
- ▶ Solu-medrol IV pulse therapy at 1g/day over 2-3 hour period, repeat for 5 days, if patient is not responding to PO meds.

Pemphigus Vulgaris

- ▶ Use of an immunosuppressant is helpful in diminishing the need for corticosteroids.
- ▶ Mycophenolate mofetil or Azathioprine are common
- ▶ Risk of death in pemphigus from side effect of oral prednisone is greater than the risk of death from the disease itself

Table 31.4 Therapeutic ladder for pemphigus vulgaris.

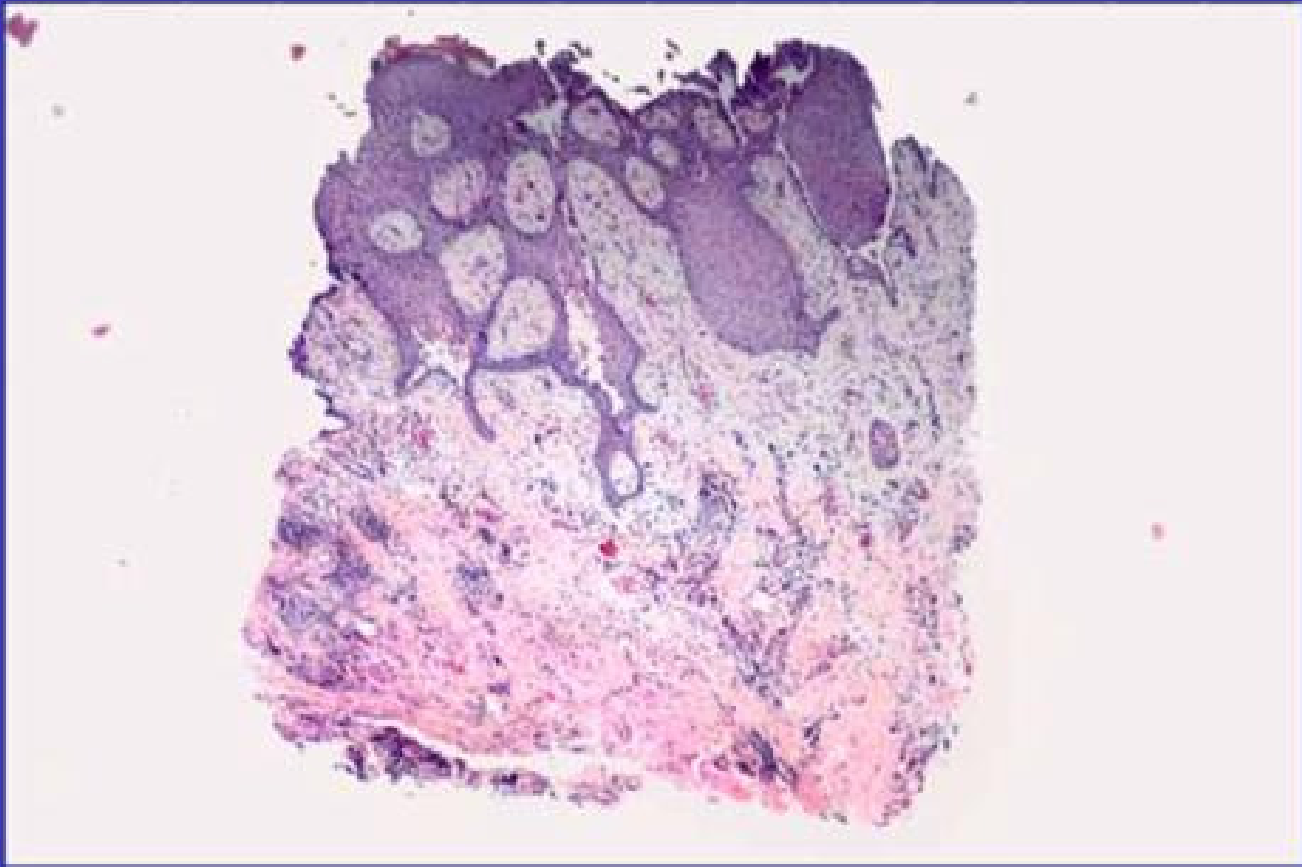
THERAPEUTIC LADDER FOR PEMPHIGUS VULGARIS	
Standard treatment	
Oral prednisone:	1.0 mg/kg/day as a initial dose (usually 60 mg/day) (1)
Aggressive treatment	
Immunosuppressive agents in combination with prednisone:	
Azathioprine:	2–4 mg/kg/day (usually 100 to 300 mg/day) (2)
Cyclophosphamide:	1–3 mg/kg/day (usually 50 to 200 mg/day) (2)
Mycophenolate mofetil:	2–3 g/day (2)
Cyclosporine:	5 mg/kg/day (3)
Pulse methyl prednisolone:	1 g/day over a period of 2–3 hours for 3–5 consecutive days (2)
Plasmapheresis (2)	
High dose intravenous immunoglobulin (2)	
<i>Key to evidence-based support: (1) good; (2) medium; (3) weak.</i>	

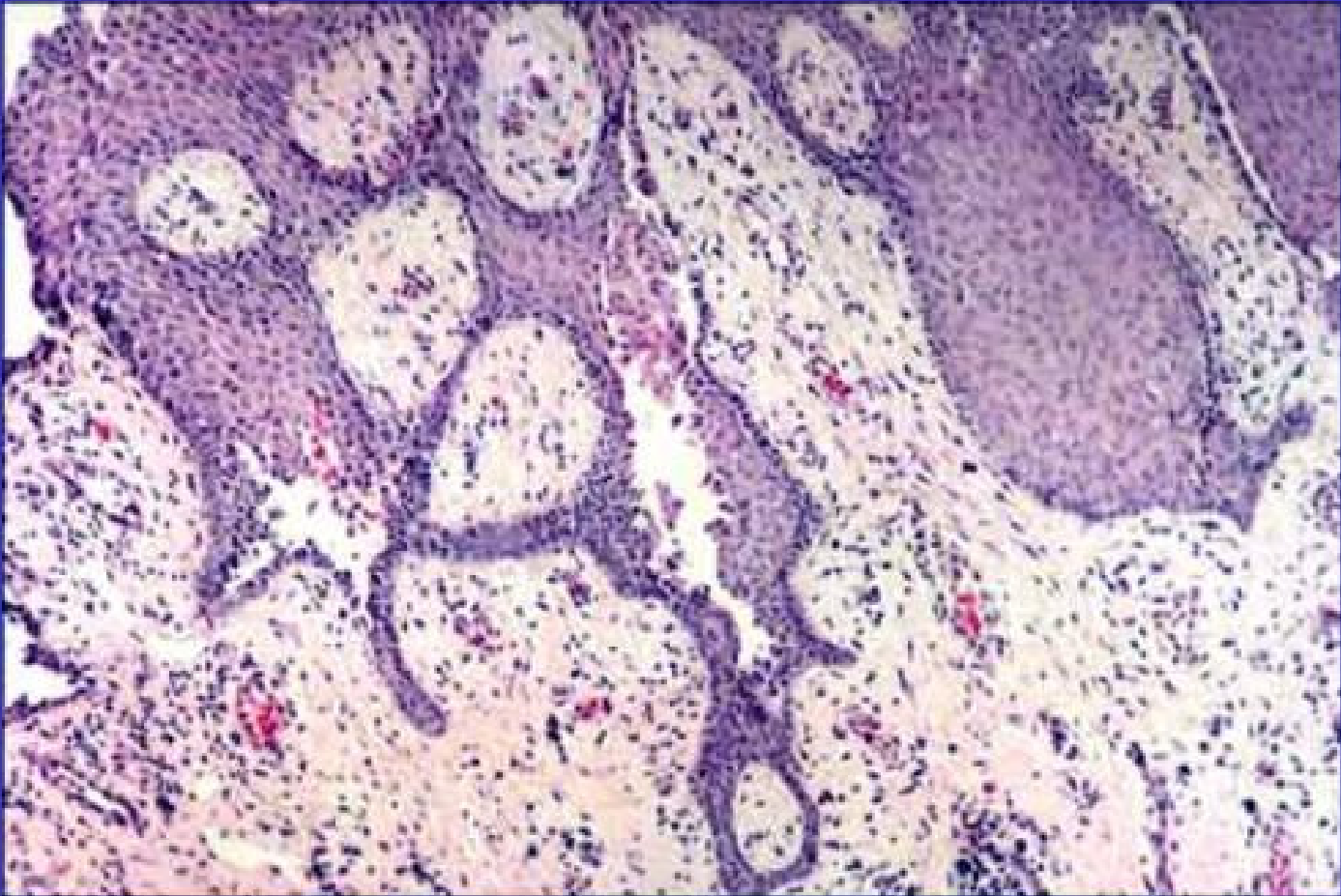
Pemphigus Vegetans

- ▶ A variant of pemphigus vulgaris
- ▶ 2 forms:
 - ▶ 1.) Neumann type
 - ▶ 2.) Hallopeau type (more localized)
 - ▶ Eosinophil microabscesses
- ▶ Clinical
 - ▶ flaccid bullae that become erosions and form fungoid vegetation or papillomatous proliferations, especially in body folds/scalp
- ▶ Histology
 - ▶ Identical to pemphigus vulgaris, but there is an increased papillary proliferation and marked epidermal hyperplasia



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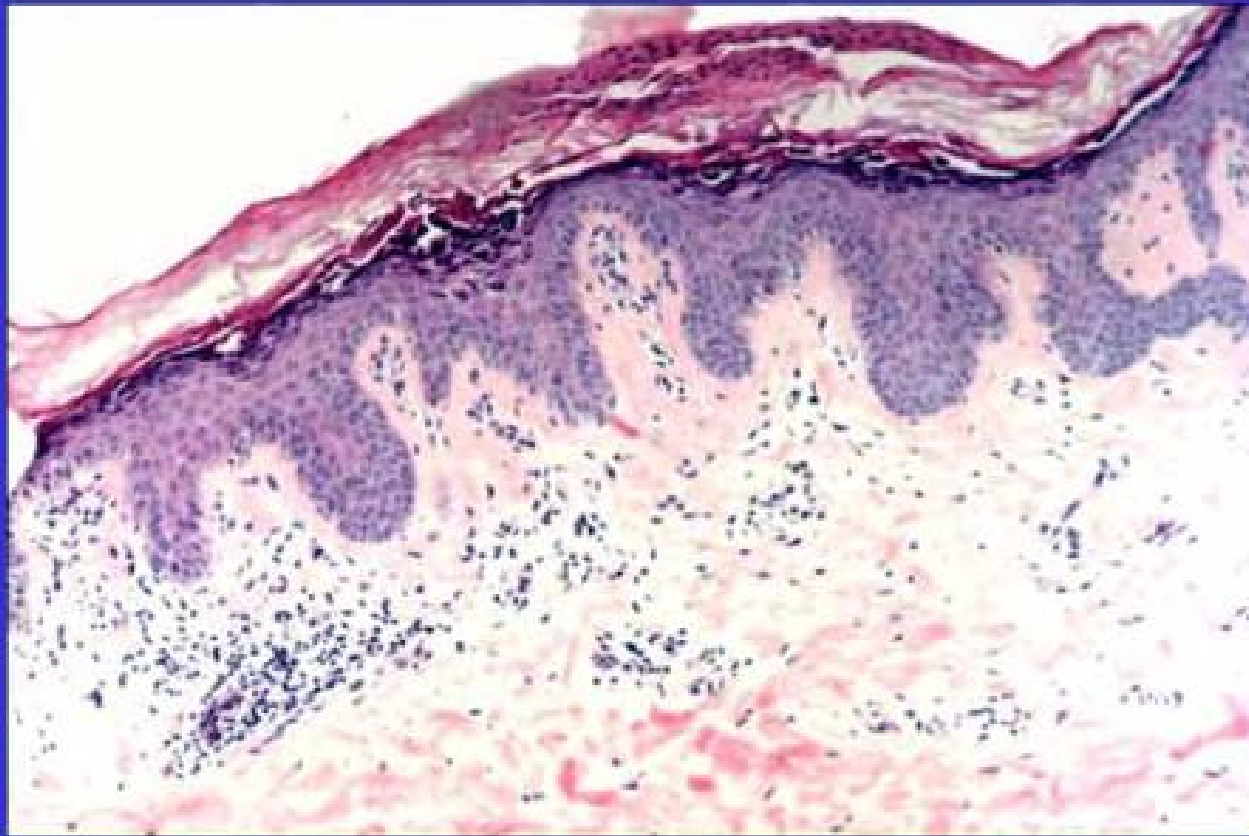
Pemphigus Foliaceus

- ▶ Mild, chronic variety of pemphigus characterized by flaccid bullae and generalized or localized exfoliation.
- ▶ Usually presents in 4th or 5th decade
- ▶ Nikolsky sign present. **Oral lesions rarely seen.**
- ▶ **Desmoglein-1** antibody.
- ▶ IIF uses **guinea pig esophagus**
- ▶ Patients are not severely ill.
- ▶ May be caused by penicillamine or captopril (and the others that cause P. vulgaris)

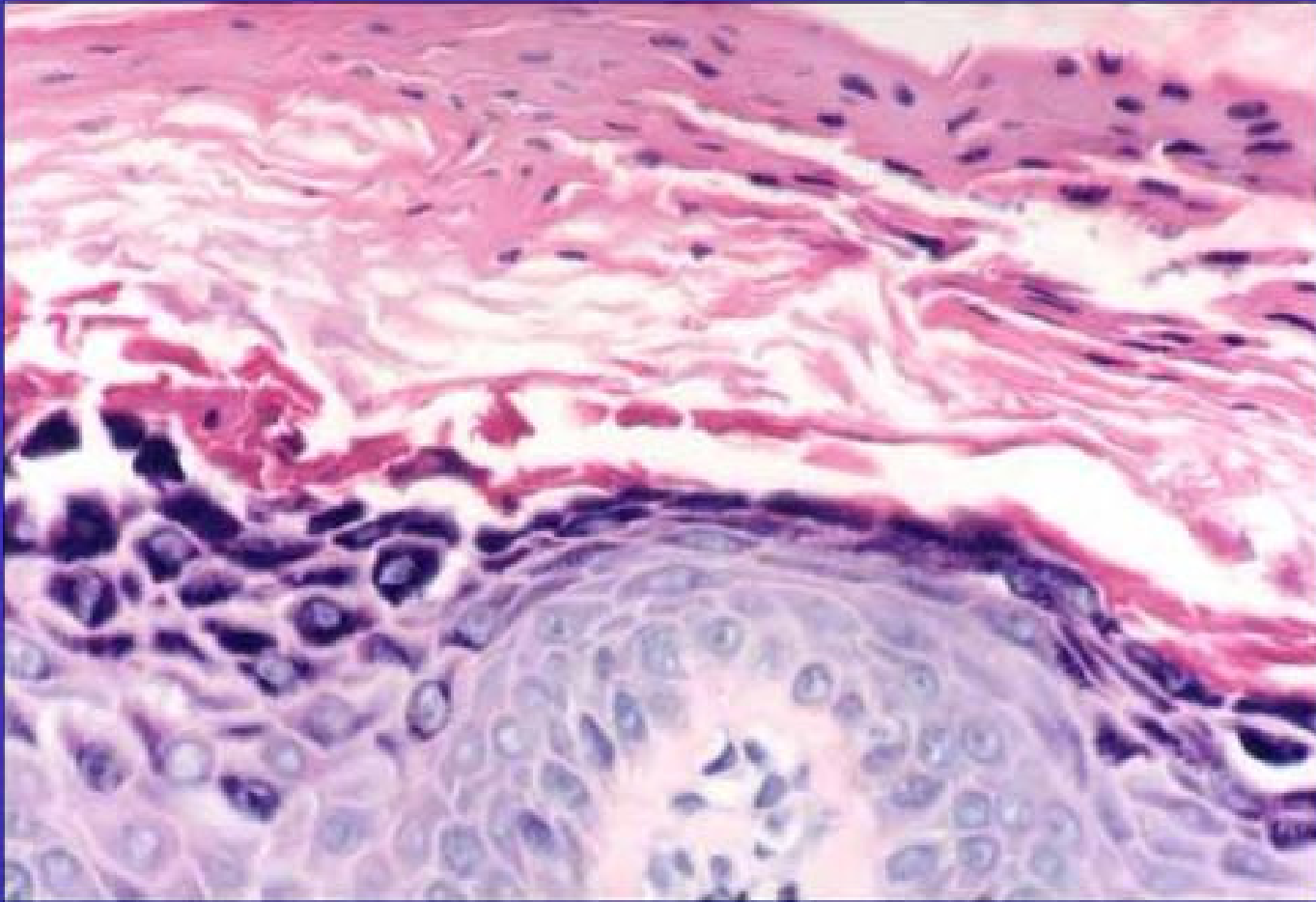


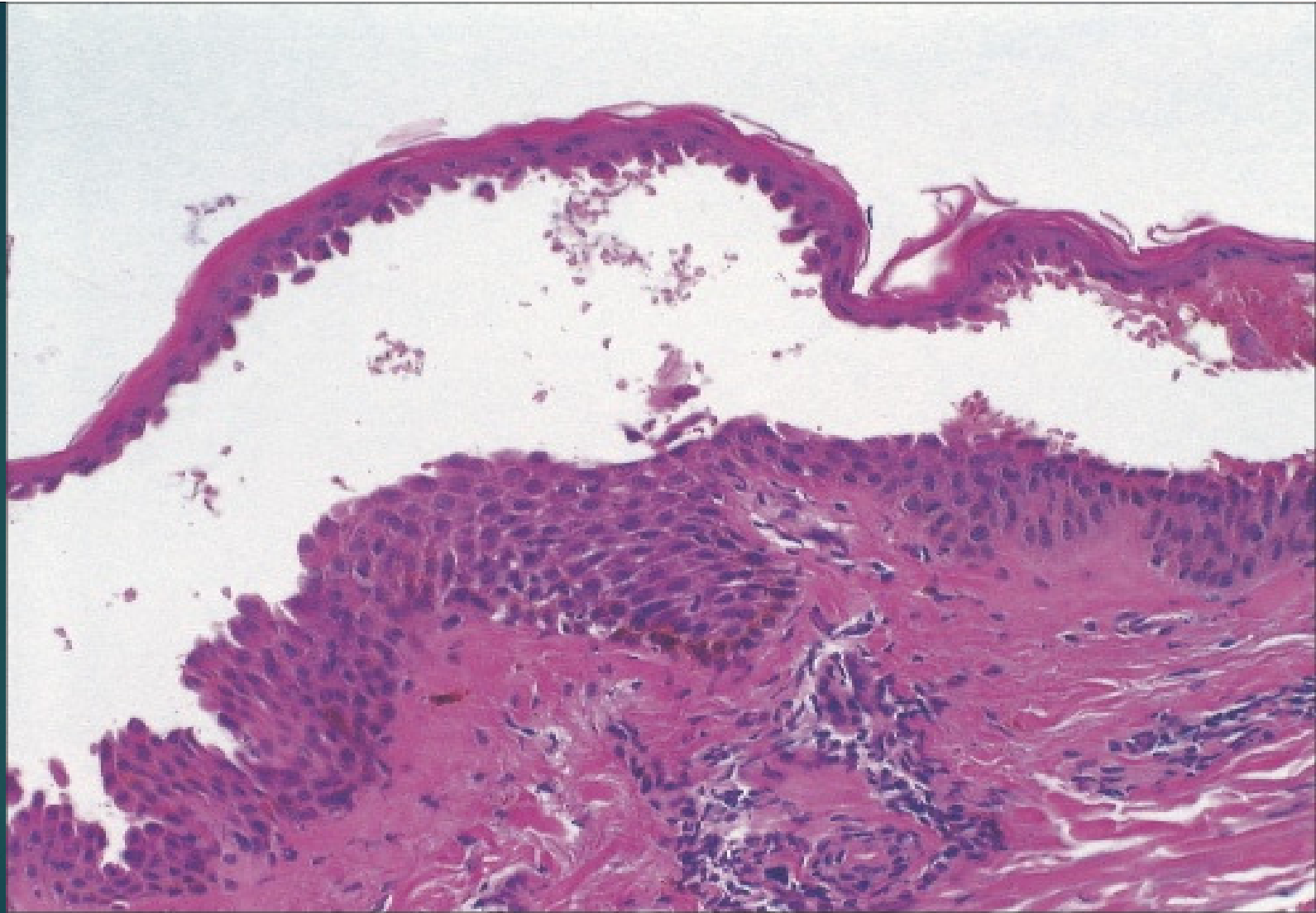
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“Corn flake” scale



Acantholysis of granular layer





Pemphigus Erythematosus

- ▶ ***Senear-Usher Syndrome***
- ▶ Resembles lupus erythematosus
- ▶ Positive for lupus band in 80% of patients
- ▶ Histologically resemble pemphigus foliaceus
 - ▶ IgG and complement - intracellular and basement membrane
- ▶ Dosage of prednisone required for control usually is much lower than that of pemphigus foliaceus, topicals too
- ▶ **Anti desmoglein-1 not desmoglein-3**



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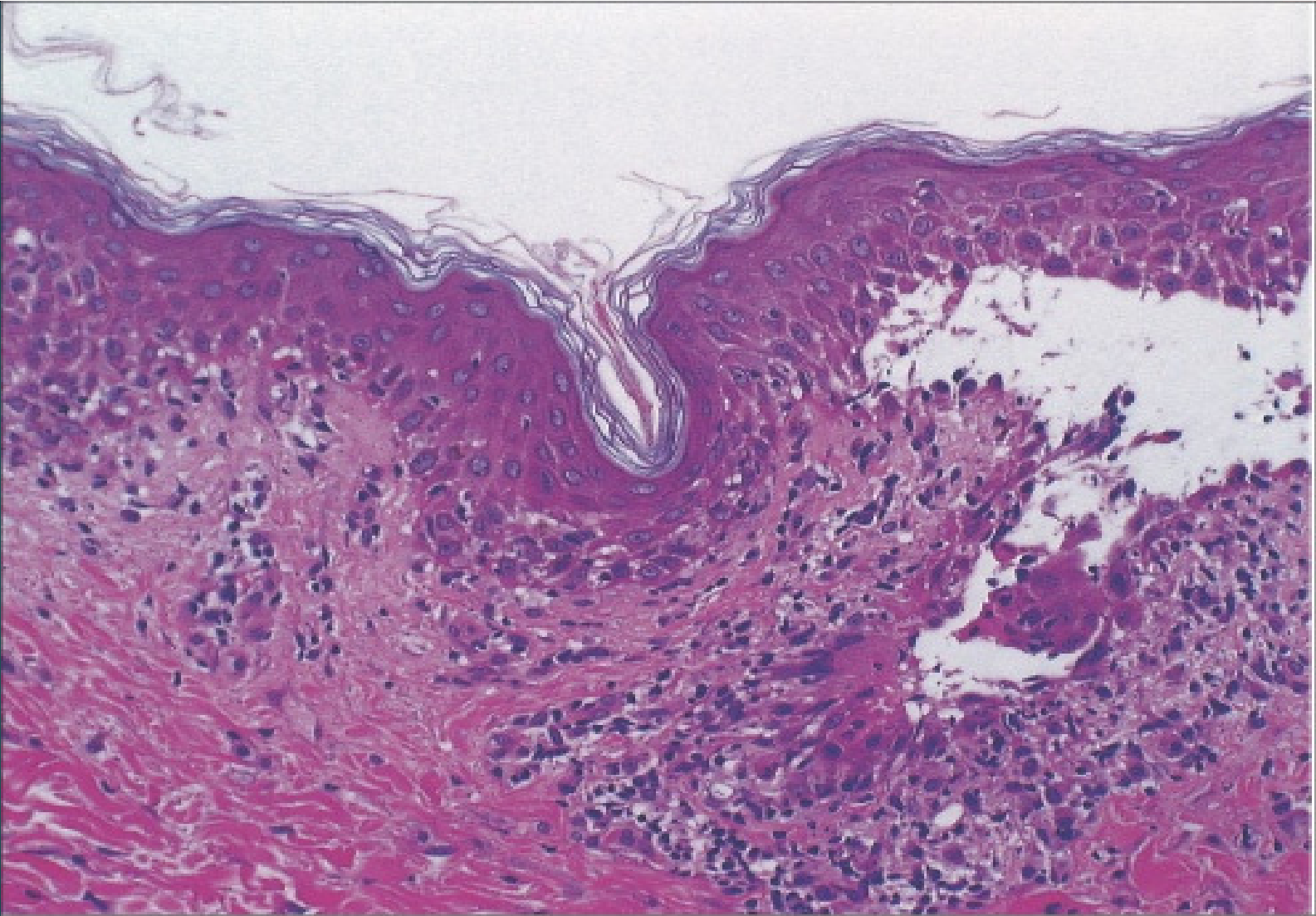
Paraneoplastic Pemphigus

- ▶ Painful mucosal lesions may present as lichenoid or Stevens-Johnson-like
- ▶ Skin lesions polymorphous - may appear as erythematous macules, lichenoid lesions, erythema multiforme-like lesions, flaccid bullae, and erosions.
- ▶ DIF reveals IgG and C3 intercellularly.
- ▶ IIF uses **rat bladder** epithelium
- ▶ Most common assoc. are **non-Hodgkin lymphoma, CLL, Castleman's disease**, sarcoma, thymoma
- ▶ Associated with HLA-DRB1*03



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The characteristic clinical feature is severe intractable stomatitis that extends onto the vermilion lip



Bullous Pemphigoid

▶ Demographics

- ▶ Elderly
- ▶ Average age of onset: 65 to 75

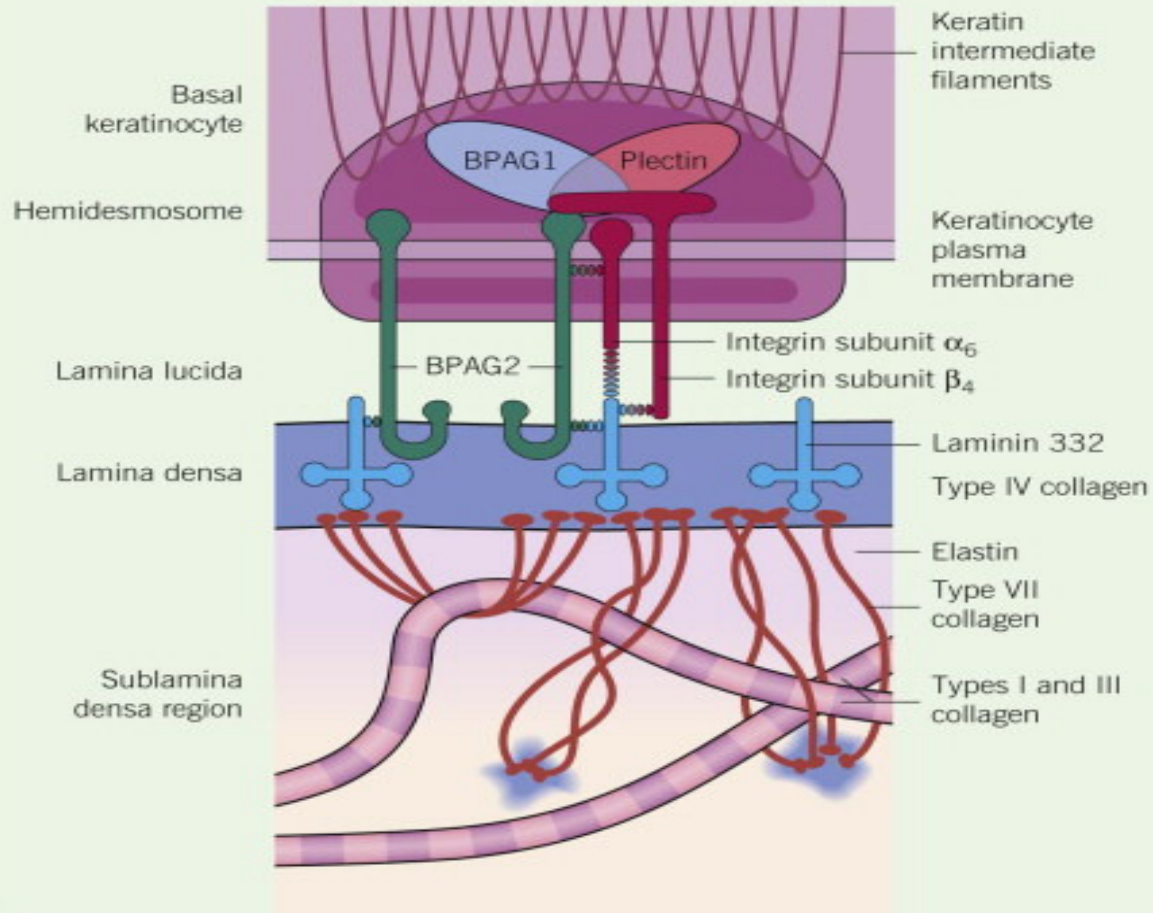
▶ Etiology

- ▶ Circulating basement membrane zone antibodies of the IgG class present 70%
- ▶ Site of IgG binding has been localized to the lamina lucida, with accentuation near hemidesmosome
- ▶ Bullous pemphigoid antigen 1 (BPAg1 – 230kD) and 2 (BPAg2 – 180kD) identified in 90% of patients

Bullous Pemphigoid

- ▶ Large bullae
- ▶ When rupture, shows large denuded area and do not generally increase in size
- ▶ Denuded areas show a tendency to heal spontaneously
- ▶ Begins at a localized site, frequently the shin
- ▶ Young girls may be initially seen with localized vulvar erosions and ulcers that resemble signs of child abuse (rare, as is MC in elderly)

INTERACTIONS OF SELECTED MOLECULES WITHIN THE EPIDERMAL BASEMENT MEMBRANE



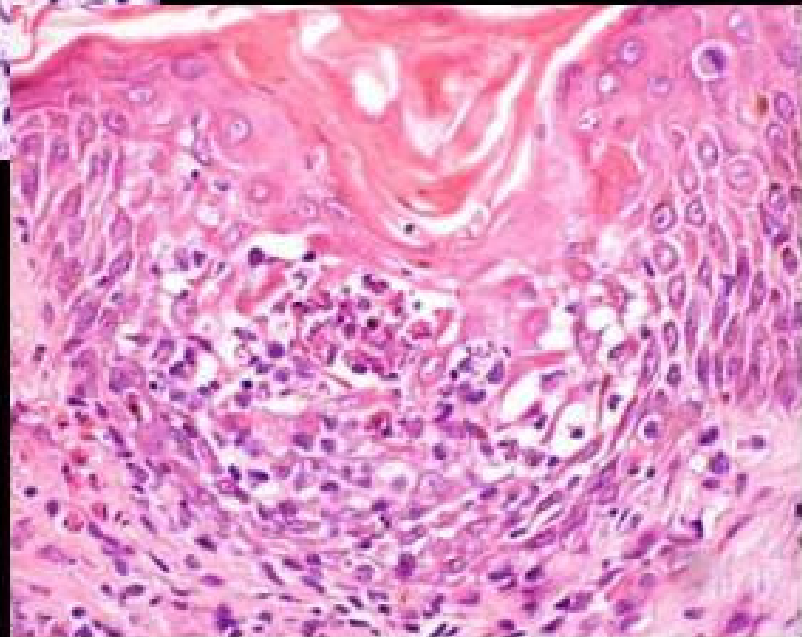
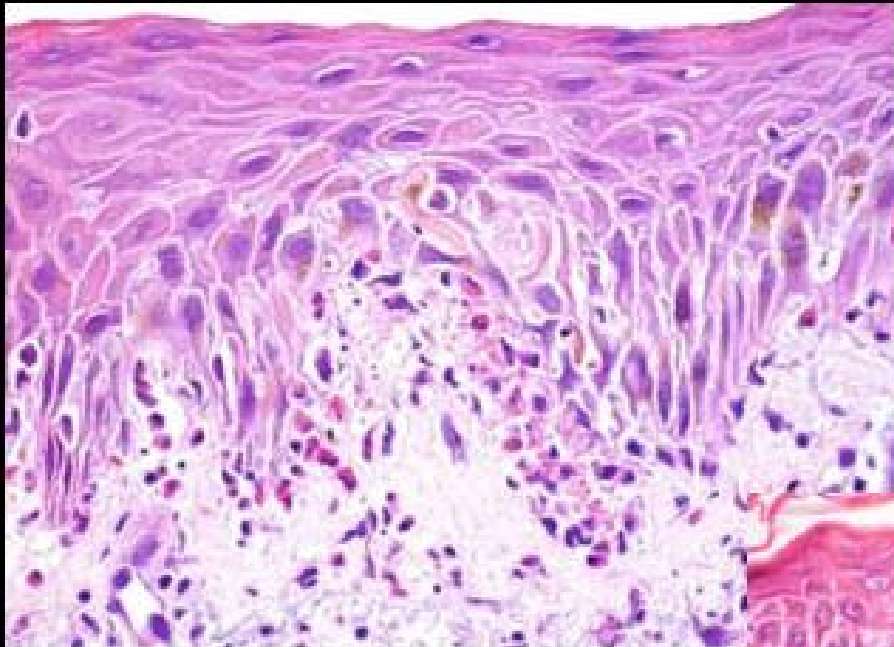




Pemphigoid Histology

- ▶ Eosinophils predominate
 - ▶ Subepidermal bulla with intact roof
 - ▶ Vacuolization along the DEJ adjacent to bulla
 - ▶ Edematous papillae often jut into bulla cavity
 - ▶ Eosinophils in superficial dermis and bulla cavity
 - ▶ Eosinophils migrate into epidermis in early lesions (eosinophilic spongiosis)

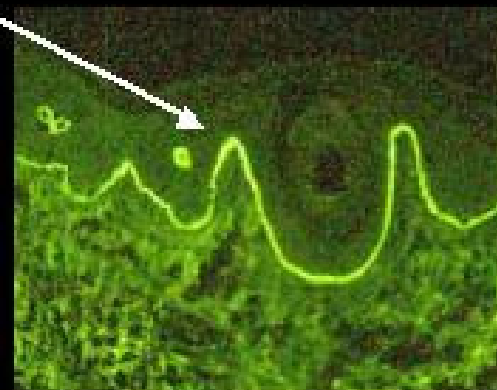
Pemphigoid





Direct Immunofluorescence

- Linear IgG (90%)
and C₃ (100%)
along the DEJ



Treatment

- ▶ Same treatment for pemphigus, with the expectation that disease will respond readily with lower dose of corticosteroid.
- ▶ Even with extensive dz, topical steroids (class 1) should be attempted
- ▶ Tetracycline class with nicotinamide TID
- ▶ In severe case, pulse therapy with methylprednisolone giving 15mg/kg in 16 ml bacteriostatic water over period of 30 to 60 minutes daily for 3 doses.
- ▶ Azathioprine is commonly used in resistant cases
- ▶ Dapsone, Mycophenolate mofetil, methotrexate, IVIG, cyclosporine, others used

Cicatricial Pemphigoid (Benign Mucosal Pemphigoid)

- ▶ Vesicles which quickly rupture, leaving erosions and ulcers with scarring
- ▶ Primarily occurs on mucous membranes, conjunctiva (66%) and oral mucosa (90%)
- ▶ Oral mucosa may be the only affected site for years; desquamative gingivitis of buccal mucosa
- ▶ Ocular complications: bilateral, flaccid vesicles on conjunctiva, xerosis, symblepharon, and blindness may result
- ▶ Generally confined to head and neck
- ▶ 25% have cutaneous lesions as well





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Cicatricial Pemphigoid

- ▶ Tends to affect middle-aged to elderly (avg age 58)
- ▶ 2:1 female:male ratio
- ▶ Chronic disease that may lead to slowly progressive shrinkage of the ocular mucous membranes and blindness
- ▶ Also occurs in pharynx, esophagus, larynx, nose, penis, vagina, anal mucosa



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Cicatricial Pemphigoid

- ▶ Cutaneous lesions in 25%; tense bullae
- ▶ Bullae heal with or without scarring, occur on the face, scalp, neck, inguinal region and extremities
- ▶ Some pts may have antibodies targeted against classic bullous pemphigoid antigens and should be classified as “mucosal predominate bullous pemphigoid”
- ▶ Chronic course, pts health not usually affected

Cicatricial Pemphigoid

- ▶ Direct IF: C3 and IgG at the lamina lucida in 80-95%
- ▶ Tx: Mild cases topical steroids, or intralesional triamcinolone every 2-4 weeks
- ▶ Tx: Dapsone, prednisone, azathioprine, cyclophosphamide, cyclosporine, TCN, mycophenolate mofetil

Dermatitis Herpetiformis (Duhring)

- ▶ Chronic, relapsing, severely pruritic
- ▶ Clinical
 - ▶ Grouped symmetrical, polymorphous, erythematous-based lesions (often extensor)
 - ▶ May be papular, papulovesicular, vesiculobullous, bullous, or urticarial
 - ▶ Itching and burning are intense
 - ▶ Spontaneous remissions





Dermatitis Herpetiformis

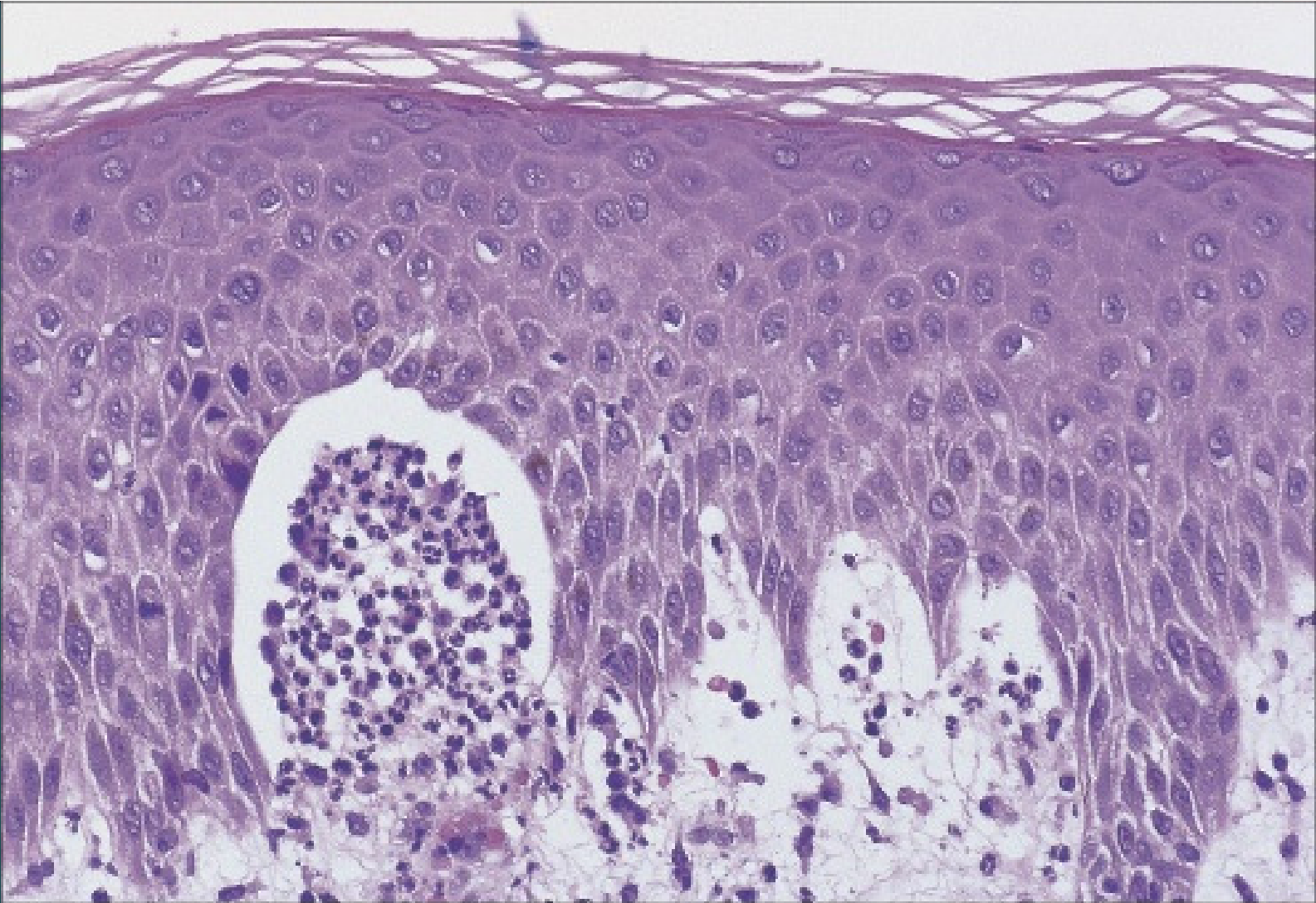
- ▶ Very few patients with DH ever have diarrhea although DH is associated with Gluten-sensitive-enteropathy (GSE)
- ▶ 77-90% of pts with DH and IgA deposits in the skin are HLA-B8 positive (like GSE)
- ▶ HLA-DR3 and DQw2 increased as well
- ▶ Gluten is a protein found in cereals except for rice, oats, and corn (ROC diet)
- ▶ IgA antibodies are formed in the jejunum, may deposit in the skin

Dermatitis Herpetiformis

- ▶ Associated with thyroid disorders, small bowel lymphoma, non-Hodgkins lymphoma
- ▶ 70-100% of pts have abnormalities of the jejunal mucosa
- ▶ Gluten-free diet decreases Dapsone dose requirements after 3-4 months

Dermatitis Herpetiformis

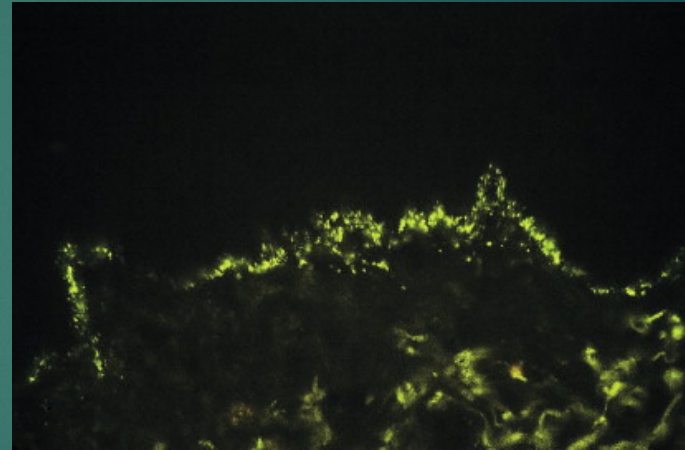
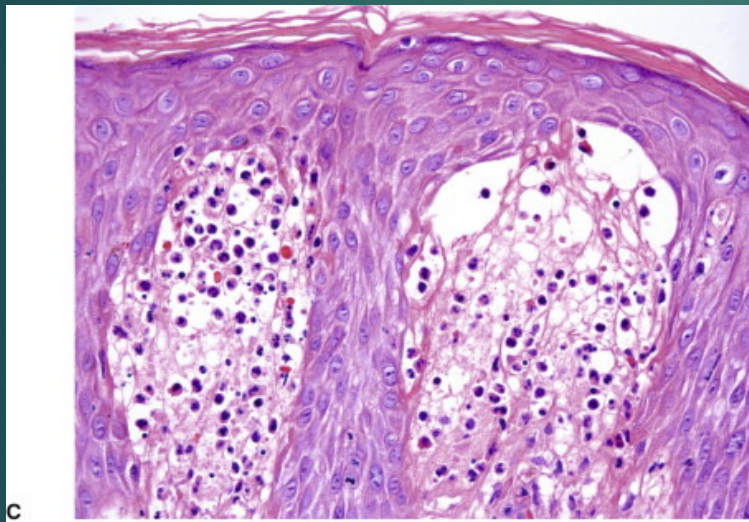
- ▶ Ddx: Pemphigoid, EM, scabies, contact dermatitis, atopic dermatitis, eczema, insect bites, prurigo nodularis
- ▶ **DIF: IgA in a granular pattern in the dermal papillae in normal skin is specific and pathognomonic for DH**



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Dermatitis herpetiformis

- ▶ DIF: IgA in a granular pattern in the dermal papillae in normal skin is specific and pathognomonic for DH



Dermatitis Herpetiformis

- ▶ 70% Circulating IgA antibodies to smooth muscle cell endomysium (anti-endomysial antibodies)
- ▶ Transglutaminase 3 (TTG-3), major autoantigen in gluten-sensitive enteropathy
- ▶ IgA deposits may be focal, so multiple biopsies may be needed.
- ▶ Deposits of the antibody are more often seen in previously involved skin or normal appearing skin adjacent to involved skin

Dermatitis Herpetiformis

- ▶ Equal male:female
- ▶ Onset between 20 to 40 years
- ▶ Treatment
 - ▶ Dapsone 50-300mg daily (hemolytic anemia, methemoglobinemia; check G6PD prior to tx) monitor Hct, WBCs, LFTs
 - ▶ Gluten-free diet will decrease need for medication

Connective Tissue Diseases

- ▶ Lupus Erythematosus Variants
- ▶ Dermatomyositis
- ▶ Scleroderma
- ▶ Eosinophilic Fasciitis
- ▶ Mixed Connective Tissue Disease
- ▶ Nephrogenic Systemic Fibrosis
- ▶ Sjögren Syndrome (Sicca Syndrome)
- ▶ Rheumatoid Arthritis
- ▶ Relapsing Polychondritis

Systemic Lupus Erythematosus

- ▶ Young to middle aged **women 6:1**
- ▶ **African american 4:1**
- ▶ Wide range of symptoms/signs
- ▶ **80% SLE patients have skin involvement**
- ▶ Many have autoantibodies, arthralgia, and other signs without meeting ACR criteria early, evolve to meet them over time
- ▶ ACR has 11 criteria for diagnosis of SLE (4 are mucocutaneous findings)

SLE

Diagnostic criteria

- ▶ **D**- discoid rash
- ▶ **O**- oral ulcers (21%)
- ▶ **P**- pleuritis, pericarditis
- ▶ **A**- arthritis
- ▶ **M**- malar rash
- ▶ **I**- immunologic disorders: anti-dsDNA antibodies, anti-Sm, anti-phospholipid antibodies
- ▶ **N**- neurologic disorders : seizure, psychosis (in absence of other causes)
- ▶ **R**- renal abnormalities: proteinuria >0.5g/day or casts
- ▶ **A**- ANA+
- ▶ **S**- sun sensitivity
- ▶ **H**- heme abnormalities: hemolytic anemia, thrombocytopenia, leukopenia

- ▶ **SLE= 4 or more of above criteria are satisfied**

Cutaneous Manifestations of SLE

- ▶ **Butterfly facial erythema**
(acute disease)
 - ▶ Begins on malar area & bridge of nose, may have associated edema
 - ▶ Eruption may last days to weeks and resolve w/o scarring
- ▶ Ears & chest common early sites → mild erythema to intense edema
- ▶ Persistent erythema of palms, soles, elbows, knees
- ▶ Leg ulcers, deeply punched out with little inflammation, pretibial or malar areas
- ▶ Calcinosis cutis (uncommon but dramatic when present)



Cutaneous SLE

- ▶ Mucous membrane involvement 20-30%
 - ▶ Erythema, petechiae, ulcers
 - ▶ Eyes, mouth, gums, hard palate

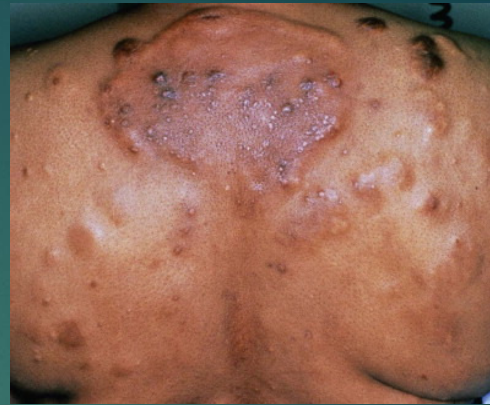


Cutaneous SLE

- ▶ Vascular lesions (occur in 50%)
 - ▶ Erythema, edema, tenangiectasia of fingers & toes
 - ▶ Nailfold capillaries (wandering loops)
 - ▶ Periungual telangiectasia, red/spotted lunula
 - ▶ Rowell syndrome
 - ▶ Erythema multiforme-like lesions predominate
- ▶ Diffuse non-scarring hair loss is common
 - ▶ Lupus hairs
 - ▶ Short hairs in frontal region from **chronic telogen effluvium** and increased **hair fragility**

Cutaneous SLE

- Papulonodular mucin deposition on trunk, arms, head or neck
- Palisaded neutrophilic granulomatous dermatitis
 - Also seen in RA or other immune complex mediated disorders



Papulonodular
mucinosis



Palisaded neutrophilic
granulomatous dermatitis

Still More Cutaneous Manifestations of SLE

▶ Bullous LE

- ▶ Single or grouped vesicles/bullae
- ▶ Widespread, sun-exposed areas
- ▶ Histology
 - ▶ Neutrophils at DE junction & in dermal papillae
 - ▶ **Subepidermal bulla containing neutrophils**
 - ▶ DIF = IgG, IgM, IgA or C3, in continuous granular fluorescence at BM zone, in or below lamina densa
 - ▶ **(antibodies to type VII collagen)**



- ▶ HLA-DR2 +
- ▶ Treatment
 - ▶ **Dramatic response to Dapsone** (unlike epidermis bullosa aquisita and TEN)

Childhood SLE

- ▶ Ages 3-15
- ▶ Girls 4:1
- ▶ Typical butterfly eruption, photosensitivity
- ▶ May be morbilliform, ulcerating, bullous, purpuric or ulcerating lesions
- ▶ **Oral mucosa** frequently involved
- ▶ May also see joint, renal, neurologic, & GI disease
- ▶ Wt. loss, fatigue, HSM, lymphadenopathy, & fever
- ▶ SLE + antiphospholipid antibodies in pediatric pts = high risk of developing thromboembolic events

Childhood SLE



Systemic Lupus Erythematosus

▶ Labs

- ▶ Hemolytic anemia, thrombocytopenia, lymphopenia, leukopenia, ↑ ↑
↑ESR (active disease), false + for syphilis, RF+, ↑serum globulin
- ▶ Albumin, RBCs, and casts are frequently found in urine
- ▶ Immunologic
 - ▶ ANA (+ in 95% SLE)
 - ▶ Anti-dsDNA (**specific**, ↑**risk of renal disease**, correlates with disease activity)
 - ▶ Anti-Sm antibody (sensitivity 10%, high **specificity**)
 - ▶ Anti-nRNP (**MCTD**)
 - ▶ Anti-La antibodies (**SCLE & Sjögren**)
 - ▶ Anti-Ro antibodies (25% SLE, 40% Sjogren, **70% SCLE, 95% Neonatal LE, C2-C4 deficient LE 50-75%**), striking photosensitivity
 - ▶ Lupus band test/DIF = continuous granular deposits of immunoglobulins & complement along DE junction in 75% of DLE
 - ▶ Serum complement (low=active disease)
 - ▶ Anti-ssDNA (**sensitive**, IgM in DLE)
 - ▶ Antiphospholipid antibody

SLE Treatment

- Avoid sun exposure, high SPF sunscreen daily
- Avoid exposure to excessive cold, heat, or trauma
- Local Treatment:
 - Topical steroids, potent or superpotent +/- occlusion
 - ILS, 2.5-10mg/ml with 30 gauge needle every 4-6 wks
 - Topical calcineurin inhibitors, (2nd line)

SLE Systemic Treatment

➤ Antimalarials

- **Plaquenil**- first line, dose of 6.5 mg/kg/day or less
 - Ocular toxicity rare at this dose, ophthalmologic consult before and at 4-6 months intervals during treatment
- If no response after 3 months try chloroquine (250mg/day) or add quinacrine (100mg/day)
 - May exacerbate skin disease in PCT patients
- **Quinacrine** = yellow discoloration of skin and conjunctivae or blue-black pigmentation of hard palate, nailbed, ear cartilage, & sclera

SLE Systemic Treatments

- Corticosteroids
 - to treat flares or obtain initial control while antimalarials are being initiated (< 3 mos)
 - Optimize to lowest possible dose to control symptoms and lab abnormalities
- Immunosuppressive agents

Table 43.4 Therapeutic ladder for lupus erythematosus

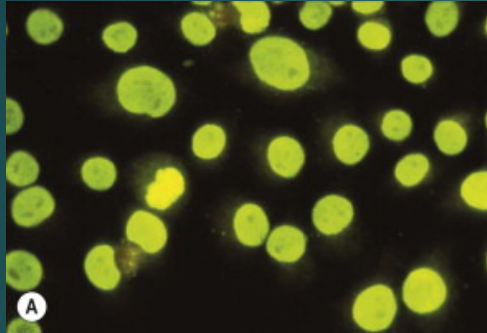
THERAPEUTIC LADDER FOR LUPUS ERYTHEMATOSUS	
Mild and/or localized disease	
<ul style="list-style-type: none"> • Sunscreens (high SPF with UVA protection) (2) • Topical corticosteroids (2) • Superpotent topical corticosteroids (2) Highly effective local treatment • Topical immunomodulators (e.g. tacrolimus) (3) • Intralesional corticosteroids (3) • Hydroxychloroquine sulfate (200 mg twice daily) (1) first line, safest systemic treatment, ocular toxicity • Above plus quinacrine (mepacrine) (100 mg/day) (2) toxicity 	
Extensive/persistent cutaneous disease	
<ul style="list-style-type: none"> • Oral retinoids (2) Second line • Dapsone/sulfapyridine (3) • Clofazimine (3) • Methotrexate (3) • Thalidomide (2) • Auranofin (3) • Azathioprine (2) 	
Systemic disease	
<ul style="list-style-type: none"> • Prednisone (1) • Azathioprine (1) • Mycophenolate (2) • Cyclophosphamide (2) • (Remicade®) (2) • Cyclosporin A (ciclosporin) (2) • Interferon (1) 	<ul style="list-style-type: none"> • CD4 monoclonal antibody (3) • IVIG 2 • Bone marrow transplant (2) • TNF-α inhibitors, e.g. Enbrel, infliximab • Leflunomide (Arava®) (2) • UVA-1 (340–400 nm) (3)
<p><i>Key to evidence-based support: (1) Double blind; (2) Case series; (3) Anecdote.</i></p>	

Connective Tissue Diseases

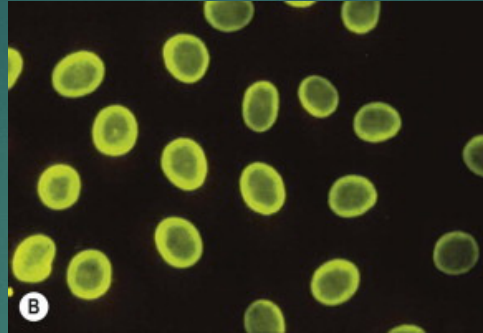
ANA Assay

- ▶ ANA immunofluorescence is considered the first line screening test for Autoimmune Connective Tissue Diseases (AI-CTD)
- ▶ Titer <1 60 using human tumor cell line substrate has little clinical utility

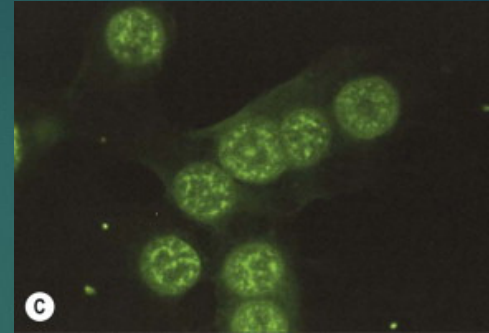
ANA Immunofluorescence Patterns



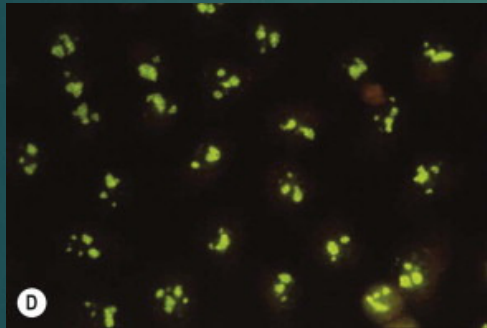
a) Homogenous (seen with antihistone antibodies)



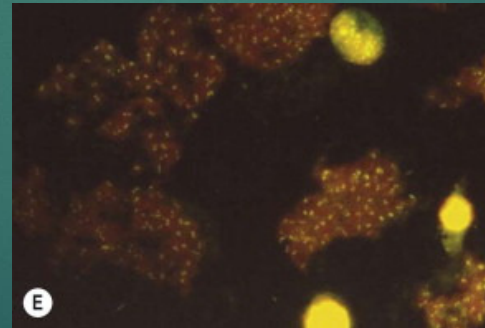
b) Peripheral (suggestive of SLE w/ renal disease)



c) Speckled (suggestive of MCTD)

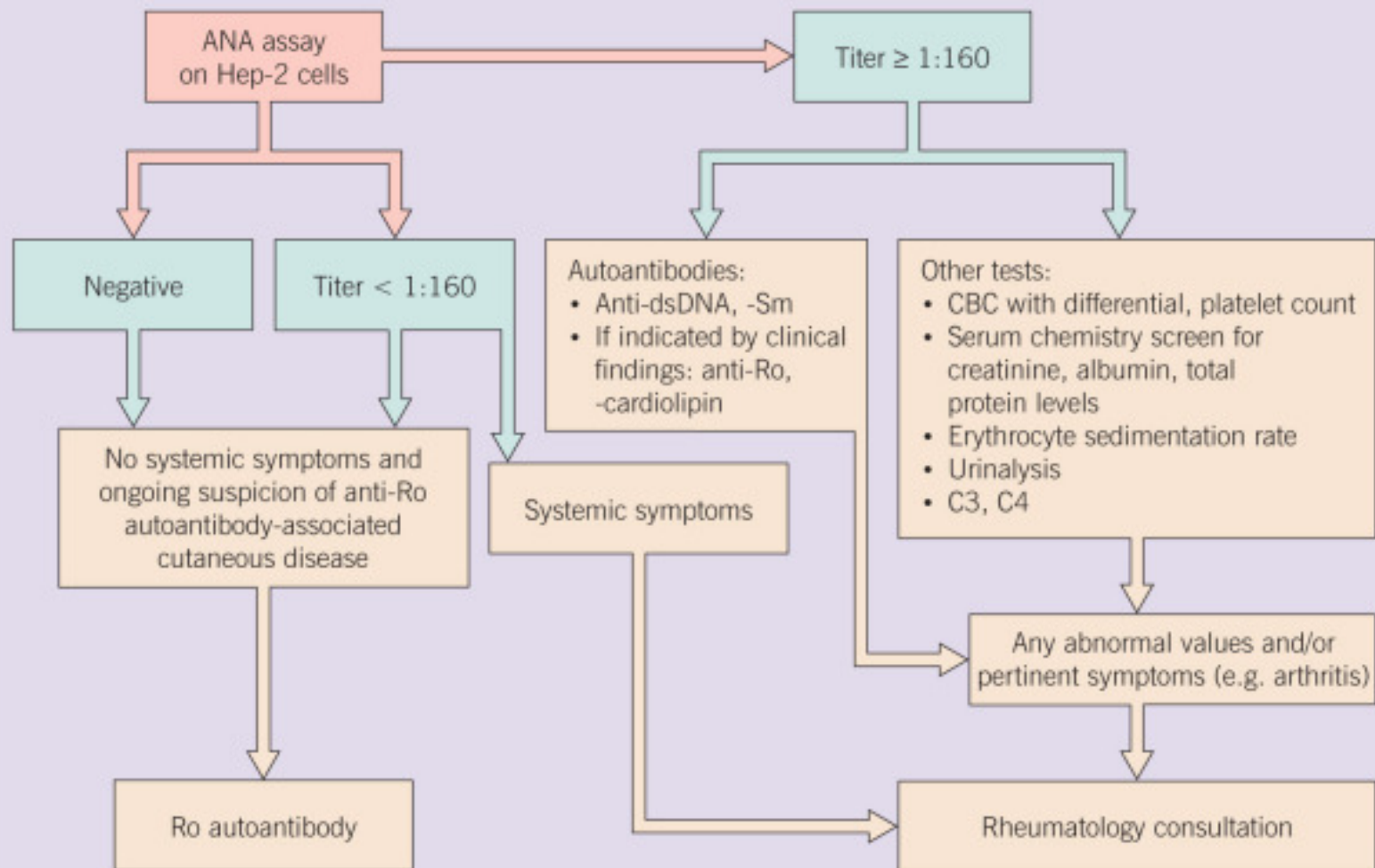


d) Nucleolar (suggestive of SSc)



e) Centromeric = **specific for CREST**

APPROACH TO THE PATIENT WITH SUSPECTED CUTANEOUS LUPUS ERYTHEMATOSUS



Lupus Erythematosus (LE)

I. **Chronic Cutaneous LE**

- A. Discoid LE
 - 1. Localized
 - 2. Disseminated
- B. Hypertrophic (verrucous) LE
- C. Lupus Erythematosus - Lichen Planus Overlap
- D. Chilblain LE
- E. Tumid Lupus - rare
- F. Lupus Panniculitis (LE Profundus)
 - 1. With No Other Involvement
 - 2. With Overlying Discoid LE
 - 3. With Systemic LE

II. **Subacute Cutaneous LE**

- A. Papulosquamous
- B. Annular
- C. Syndromes Commonly Exhibiting Similar Morphology
 - 1. Neonatal LE
 - 2. Complement Deficiency Syndromes
 - 3. Drug Induced

III. **Systemic LE**

- A. Cutaneous
- B. Systemic

Discoid LE

- Young adults, F:M=2:1
- Dull red macules or indurated plaques with adherent scale
- Lesions heal centrally first with atrophy, scarring, and dyspigmentation
- Carpet-tack like spines on undersurface of scale (Langue au chat or Cat's tongue)
- Up to 24% will have mucosal involvement.
- 95% of cases confined to the skin at the onset will remain so



Discoid Lupus Erythematosus



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Carpet tack scale (langue du chat)

Localized Discoid LE

- ▶ Lesions typically above the neck
- ▶ Scalp, bridge of nose, malar areas, lower lip, concha & external canal
- ▶ Scarring, itching, tenderness
- ▶ In scalp, perifollicular erythema & easily extractable anagen hairs = active disease
- ▶ Monitor for BCC or aggressive SCC, esp. in scars
- ▶ Progression to SLE is rare, 95% of DLE will remain DLE
- ▶ However SLE patients frequently have DLE



Generalized Discoid LE

- ▶ Less common than localized DLE
- ▶ Thorax, upper extremities + head and neck
- ▶ Scalp = balding w/ hyper and hypopigmentation
- ▶ Diffuse scarring of face & upper extremities
- ▶ Lab abnormalities more common than in localized
 - ▶ ESR, ANA, ssDNA antibodies all elevated
 - ▶ Leukopenia



Discoid LE Treatment

- ▶ SUNSCREEN!!!! Avoid heat, cold, trauma exposure
- ▶ Topical steroid, high potency with occlusion if needed
- ▶ Intralesional Injection with Kenalog
- ▶ Antimalarials: safest and most beneficial system therapy.
 - ▶ Plaquenil (hydroxychloroquine) for 3 months, if no response switch to Aralen (chloroquine); must get regular ophthalmologic exams
 - ▶ If response is still incomplete, add quinacrine, since this won't increase retinal toxicity
 - ▶ May exacerbate PCT, bleach hair/conjunctivae (quinacrine)

Neonatal LE

- ▶ Most infants are born to mothers who carry **Ro/SSA antibody**
- ▶ No lesions at birth, develop in first few weeks of life
- ▶ Annular scaling erythematous macules & plaques on head and extremities
- ▶ Raccoon eyes (periocular involvement)
- ▶ Telangiectatic macules or angiomatous papules may occur in sun-protected areas; may be persistent



- ▶ 50% of mothers asymptomatic at delivery
- ▶ 75% are of neonatal LE infants are girls

Neonatal LE

- ▶ Skin lesions usually resolve spontaneously by 6 months of age
- ▶ Usually heal without significant scarring, may see dyspigmentation or atrophy
- ▶ ½ infants half have a congenital 3rd degree heart block that is permanent
- ▶ Thrombocytopenia or hepatic disease as frequent as heart disease





NEPHROGENIC SYSTEMIC FIBROSIS

Nephrogenic Systemic Fibrosis

- ▶ Recently recognized fibrosing skin condition resembling scleromyxedema histologically
- ▶ Pts with renal insufficiency on hemodialysis who have had MRIs w/ **gadolinium-containing** contrast agents
 - ▶ Also concurrent infection, increased serum phosphate and calcium levels, and acidosis may be contributory
- ▶ Thickened, sclerotic papules/plaques on extremities and trunk (**spares face**); systemic involvement usually not present
- ▶ Different from scleromyxedema by lack of facial involvement, absence of plasma cells & lack of paraproteinemia
- ▶ No effective therapy, transplant may benefit, optimize renal function

Nephrogenic Fibrosing Dermopathy

