EVALUATION OF VESICULOBULLOUS DISORDER

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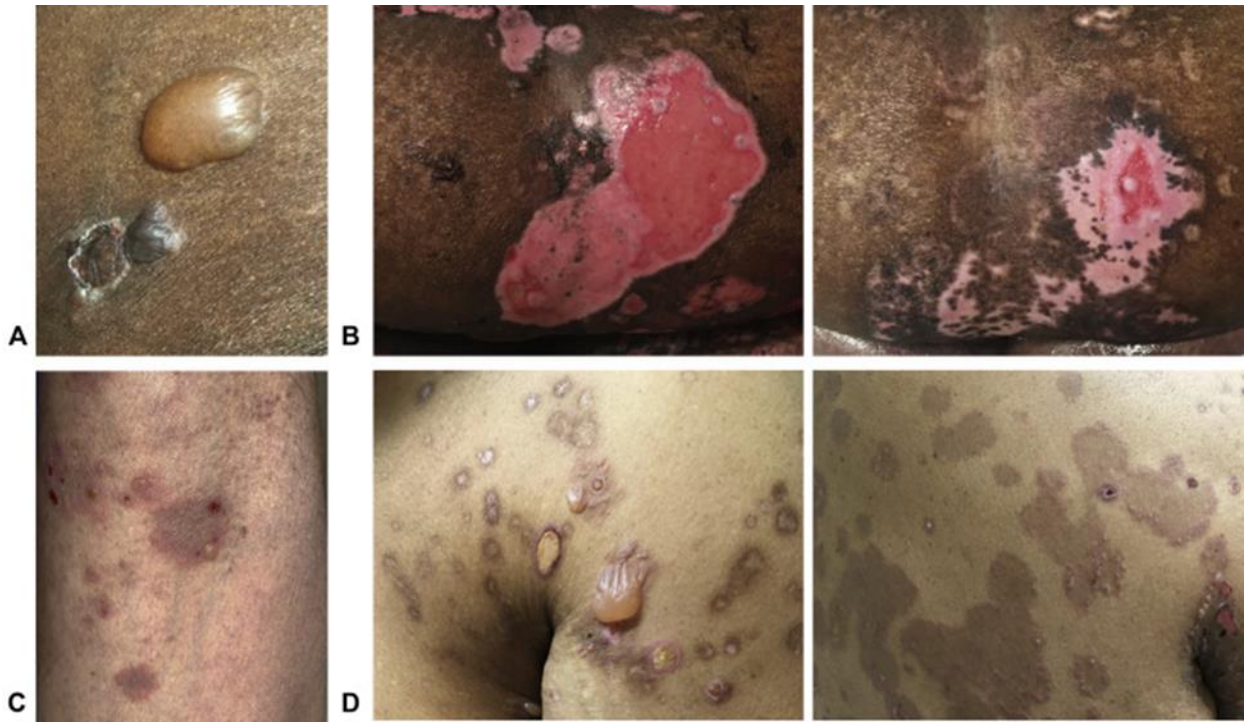
LEARNING OBJECTIVES

- At the end of lecture, a student should be able to
  - Classify vesicobullous eruptions on the basis of aetiology
  - Describe clinical features of common vesicobullous eruptions
  - Investigate a patient with vesicobullous eruption
  - Know clinical features of different immunobullous disorders

CLINICAL VIGENETTE

An 82-year-old African-American woman presented with a painful blistering eruption that began on her hands, abdomen, and extremities without mucosal erosions. Physical examination found tense bullae on the extremities and trunk lacking appreciable erythema and large abdominal erosions .



- Histopathology, immunofluorescence, and enzyme-linked immunosorbent assay (ELISA) confirmed a diagnosis of bullous pemphigoid (BP)
- The patient was treated with clobetasol cream, a tapering course of prednisone, and mycophenolate.
- After 2 months, she achieved near-total healing of erosions but had residual hypopigmentation and perifollicular repigmentation.

CORE CONCEPT

Classification of Vesicobullous Eruptions

Sr. No.	Category	Disorder
1	**Genetic**	•Epidermolysis Bullosa •Bullous Icthyosiform Erythroderma •Hailey Hailey Disease •Incontinentia Pigmenti •Acrodermatitis Enteropathica

CORE CONCEPT

Sr. No.	Category	Disorder
2	**Infection**	•**Bacterial** – Bullous Impetigo •**Viral** –Herpes Simplex,Varicella,Herpes Zooster •**Fungal** – Bullous Dermatophyte Infection
3	**Metabolic**	•Porphyrias •Bullous Diabeticorum •Pellagra

CORE CONCEPT

Sr. No.	Category	Examples
4	**Inflammatory**	•Pemphigus (vulgaris ,foliaceus) •Pemphigoid (bullous ,mucous membrane) •Linear IgA disease •Herpes Gestationis •Dermatitis Herpetiformis •Bullous SLE •Epidermolysis Bullosa Acquisita
5	**Hypersensitivity**	•Erythema Multiforme •Steven Johnson Syndrome and Toxic epidermal necrolysis •Pompholyx

CORE CONCEPT

Sr. No.	Category	Examples
6	**Exogenic**	•Bullous Papular Urticaria •Drugs •Contact Dermatitis •Friction •Radiation •Chemical Burns

CORE CONCEPT

Clinical Features

History

1.Duration

Short

Long

Recurrent

2.Onset at birth or during infancy

3.Triggers

Trauma

Drugs

Contact

Infections

CORE CONCEPT

Clinical Features

History

4. History of fever	5.Family History	6.History of Diabetes, Connective tissue disease,altered bowel habbits

CORE CONCEPT

Clinical Features (Cont…):Morphology

Sub corneal

Bulla located below the stratum corneum

- Flaccid bulla ,rupture easily so hardly seen ever.
- Scale or crust predominate with no erosions.
- Heals with no residual scarring.

Intra epidermal

Bulla in prickle cell layer of epidermis

- Flaccid bulla but take time to rupture.
- Crusted erosions on rupturing.
- Heals with pigmentary changes.

Sub epidermal

Bulla located at the dermoepidermal junction

- Tense and persistent bulla ,do not often rupture.
- Hemorrhagic crusts and ulcers on rupturing.
- Heal with milia formation and scarring.

Clinical Features (Contd…)

Distribution

- EB- sites of trauma.
- Pemphigus-scalp, face ,flexures and trunk.
- Bullous pemphigoid-trunk, limbs, flexures.
- CBDC-around body orifices.
- DH-extensors.
- Bullous papular urticaria-exposed parts.

Configuration

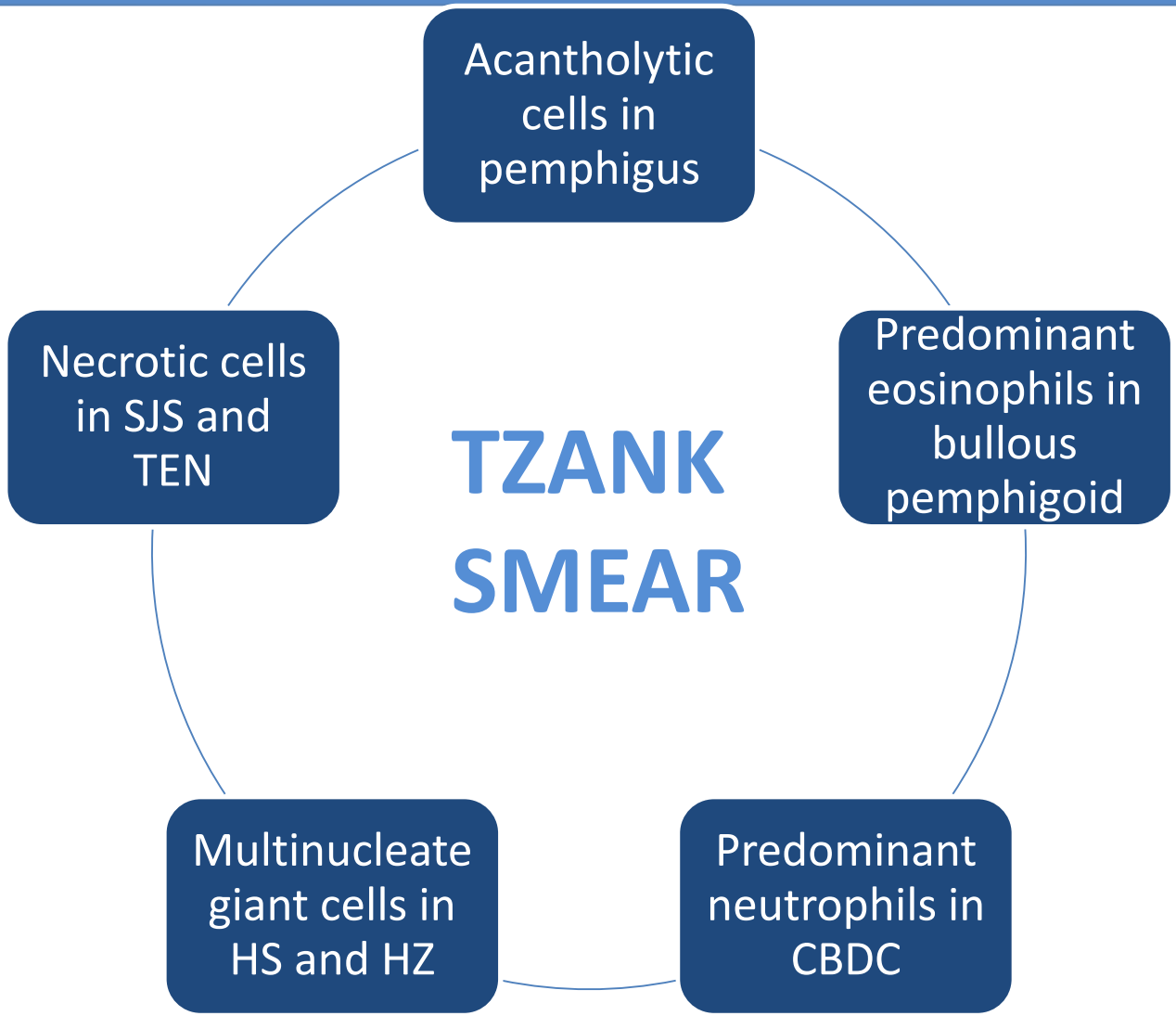
- Grouping as seen in herpes simplex and dermatitis herpetiformis.
- Segmental distribution.

Mucosal Involvement

- Some variants of EB.
- Universal in pemphigus vulgaris.
- Severe in SJS and TEN

HORIZONTAL INTEGRATION

Investigations



Investigations (Contd…) : Skin Biopsy

- **Histopathology**
- **Congenital :** Electron Microscopy
- **Metabolic:** Blood sugar levels, porphyrin levels, niacin levels
- **Infective**: Cultures,PCR, KOH preparations
- **Hypersensitivity**: Eosinophils
- I**mmunobullous**: DIF, IIF

Investigations (Contd…) : Skin Biopsy

- **Histopathology :**
- Level of the split.
  - Sub corneal.
  - Intraepidermal.
  - Dermoepidermal junction.
- Presence and type of infiltrate.
- Presence of specific cells ,e.g. acantholytic cells in pemphigus and multinucleate giant cells in herpetic infections.

Investigations (Contd…)

Subepidermal blister with dermal neutrophilic infiltration.

Inflammatory Vesicobullous Disorders

CORE CONCEPT

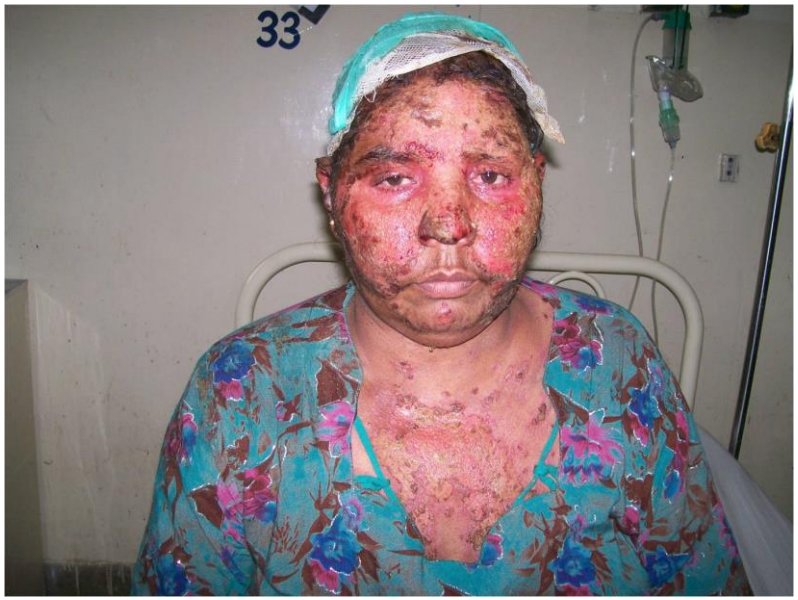
Pemphigus

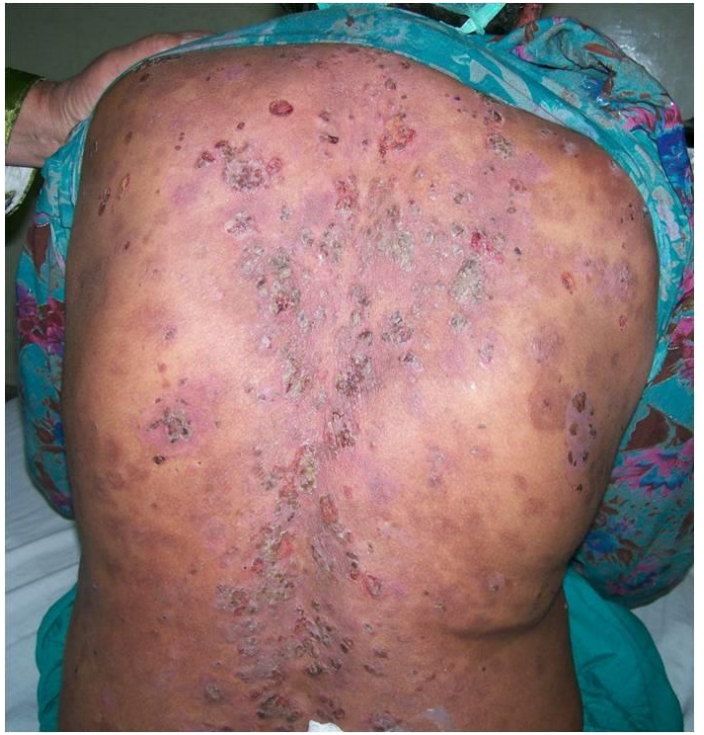
- Immunobullous disorder in which blisters form within the epidermis.
- Antibodies react with intercellular adhesion molecules, intercellular bridges disappear and keratinocytes separate from one another ,a change called acantholysis.
- Intercellular IgG deposits within the epidermis on direct immunoflourescence.
- Main types are :
    1. Pemphigus vulgaris.
    2. Pemphigus foliaceus.

CORE CONCEPT

Pemphigus vulgaris

- **Pemphigus vulgaris** can present with mucosal lesions alone or with mucocutaneous lesions and is characterized by generalized flaccid blisters which rupture rapidly to leave behind painful erosions.



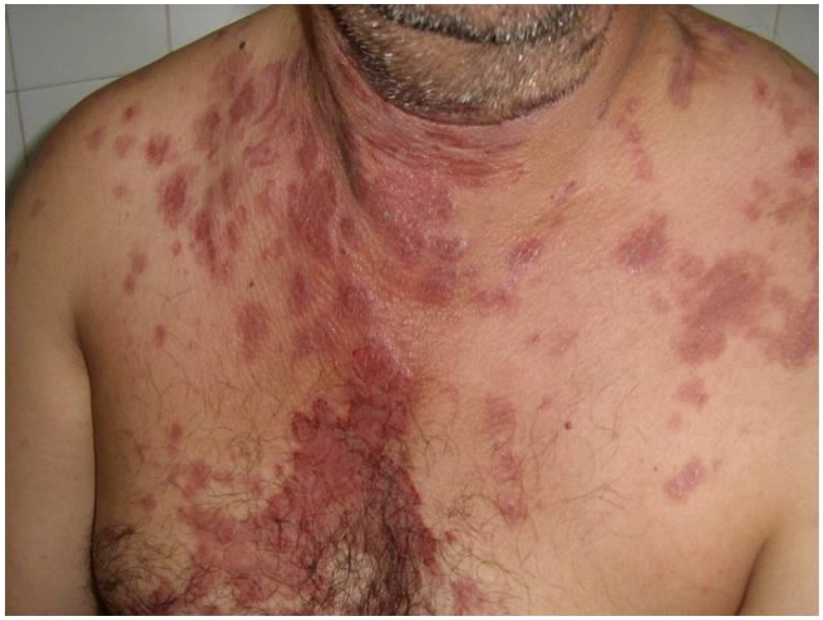


Widespread crusted erosions on face and trunk in a patient with pemphigus vulgaris.

CORE CONCEPT

Pemphigus foliaceus

- **Pemphigus foliaceus** is amore superficial form .Patients develop very superficial blisters which rupture very rapidly, thus secondary changes of scale ,crust and erosions may be the only findings present. Mucosal lesions are uncommon.



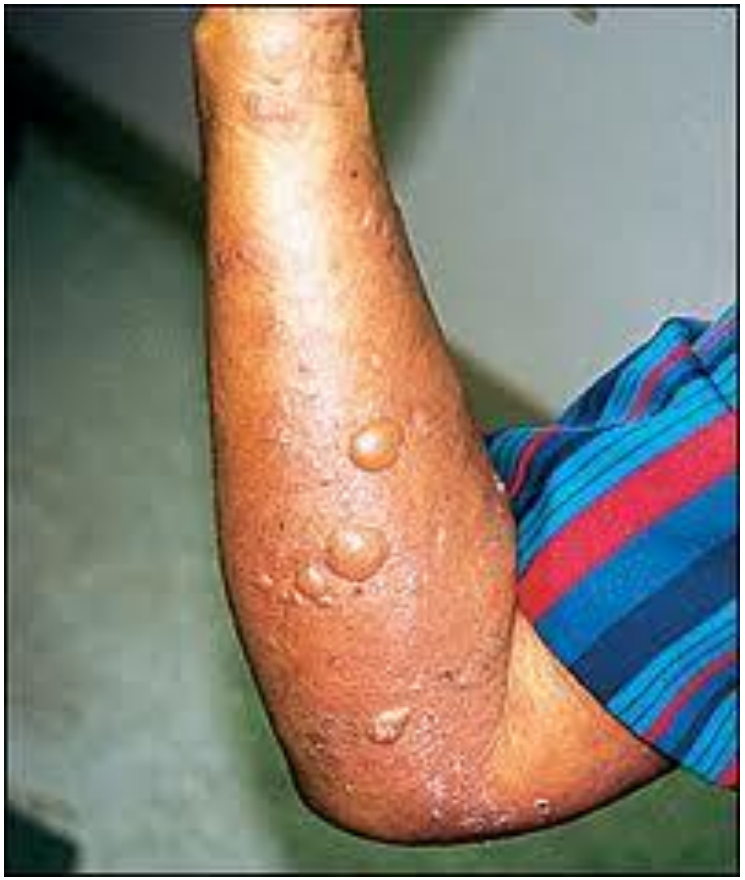
Well demarcated scaly ,crusted lesions scattered over the chest in a patient with pemphigus foliaceus.

CORE CONCEPT

Bullous Pemphigoid

- A chronic ,autoimmune ,blistering disease of elderly people caused by auto antibodies against the hemidesmosomes in the basement membrane.
- The blisters are sub epidermal and intact epidermis forms the roof.
- Presents with pruritis and urticated erythematous lesions → large tense blisters → rupture in a few days and heal with pigmentary changes.
- Mucous membrane can be involved.
- Linear IgG deposits at basement membrane zone on direct immunoflourescence.

Bullous Pemphigoid (Contd…)



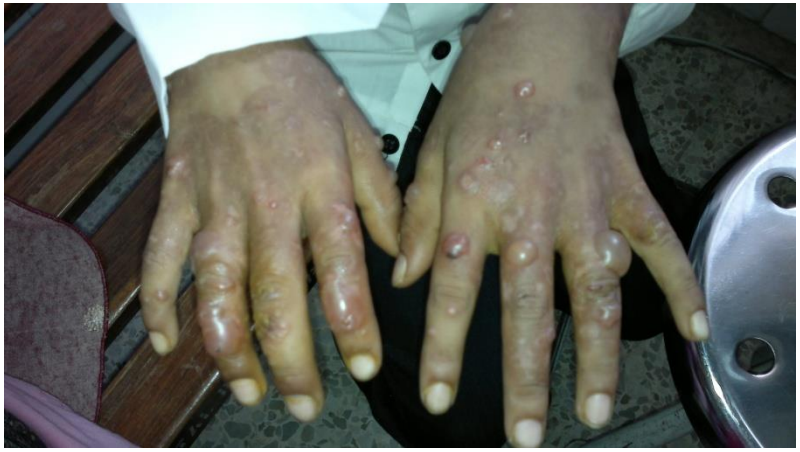
Large tense blisters on an erythematous base.

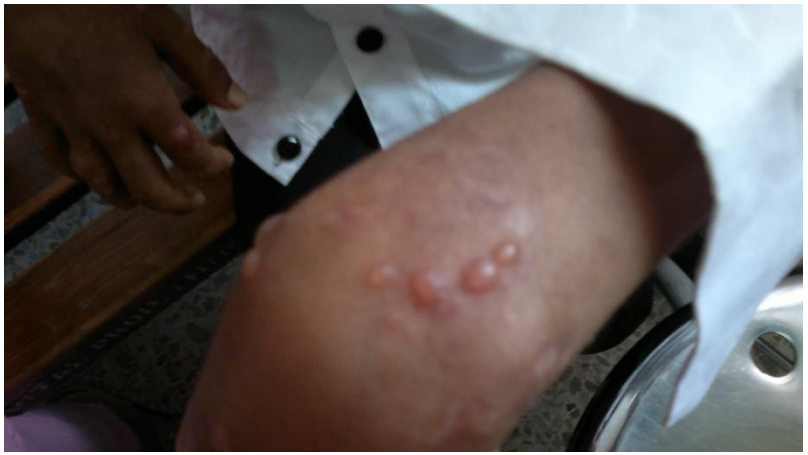
CORE CONCEPT

Linear IgA Disease

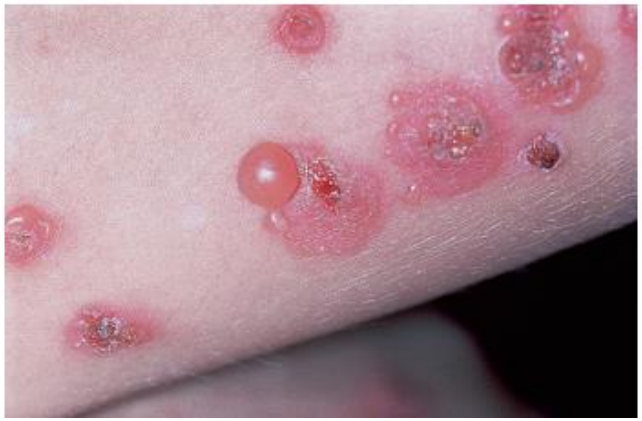
- Chronic ,acquired ,sub epidermal disease of children and adults.
- Cutaneous and mucosal involvement.
- IgA basement antibodies.
- Intensely itchy.
- Presents as urticated plaques ,papules, annular or polycyclic lesions with tense blistering around the edge ,the string of pearl sign.
- Linear IgA deposits at BMZ on direct IMF.

Linear IgA Disease (Contd…)





Chronic bullous disease of childhood ,with tense blisters as a string of pearl around the older lesions.



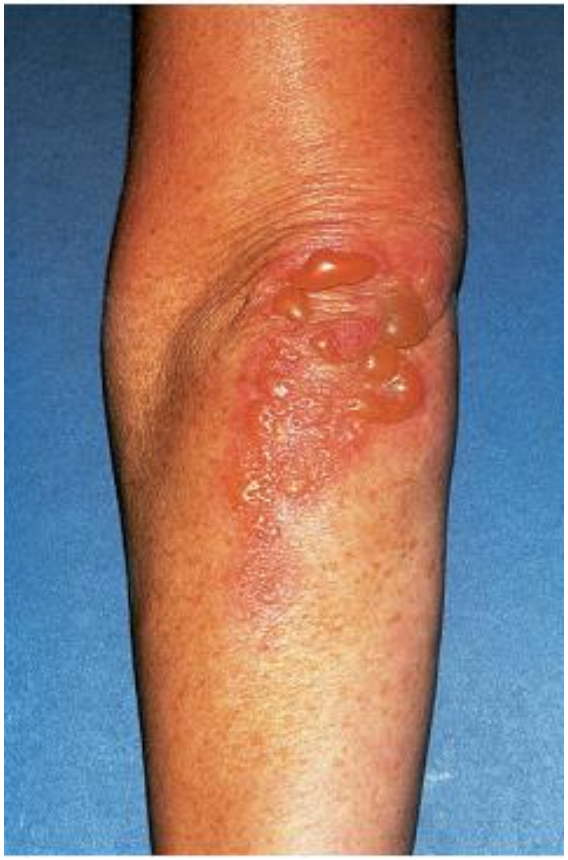
Linear IgA disease in an adult with intact tense bullae and annular lesions on the thigh.

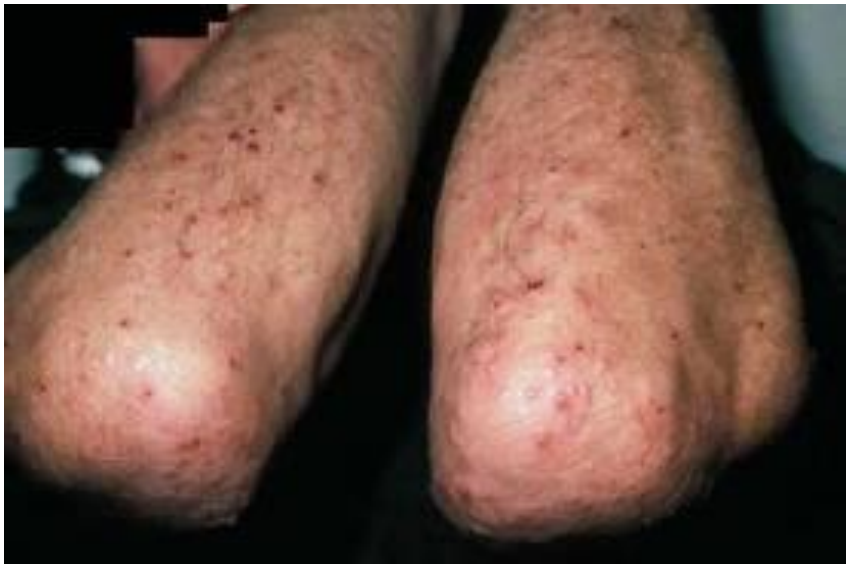
CORE CONCEPT

Dermatitis Herpetiformis

- Chronic ,recurrent ,papulovesicular disease.
- 2nd and 4th decade of life.
- Underlying gluten sensitive enteropathy.
- Blister is sub epidermal.
- Intnsely itchy.
- Grouped ,b/l symmetrical ,papulovesicular eruption distributed on the extensors.
- Presents with multiple erosions secondary to sratching.
- Diagnosed by classical skin presentation.
- Confirmed by immunoflourescence ,showing granular IgA deposits at dermoepidermal junction.

Dermatitis Herpetiformis (Contd…)





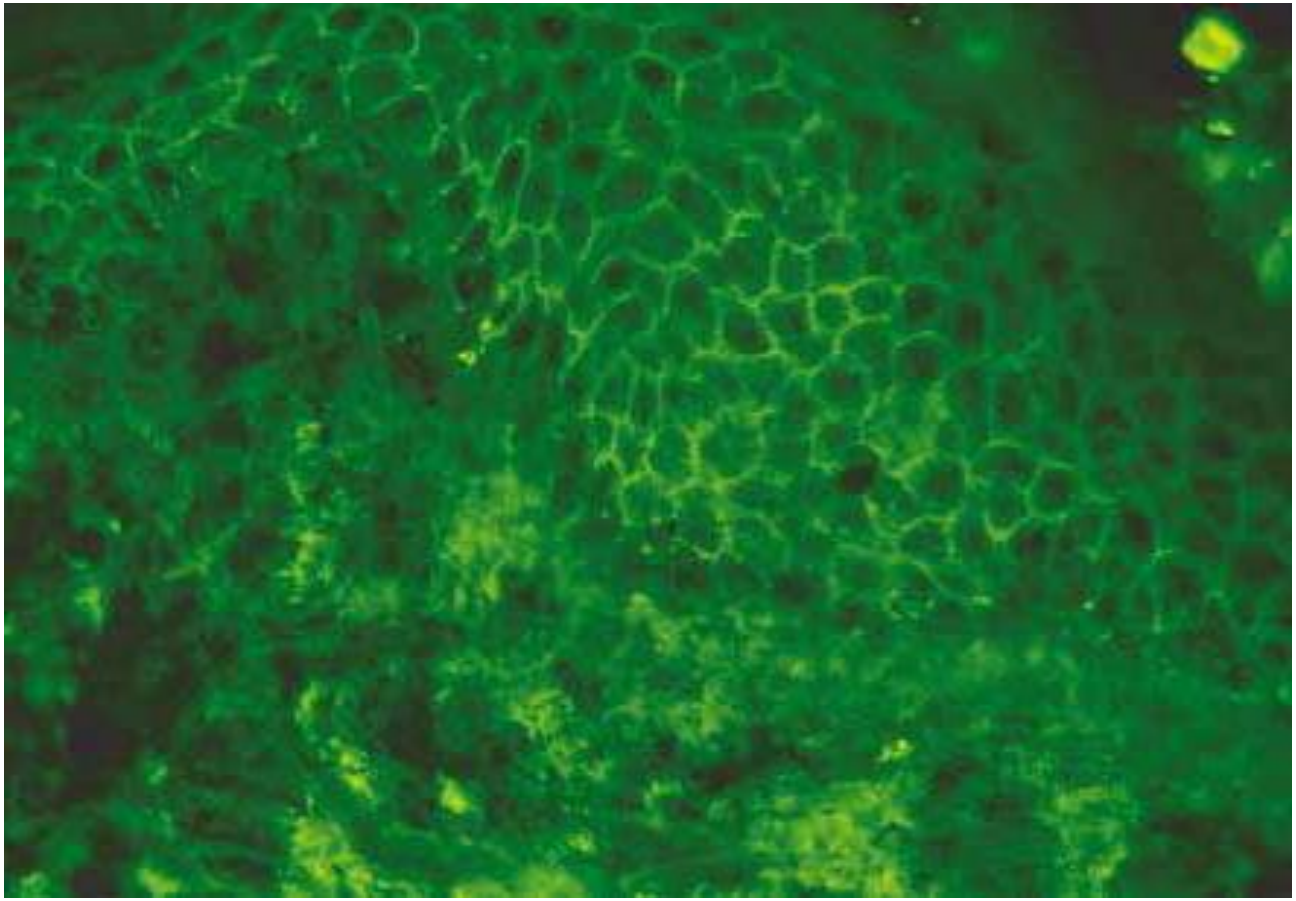
Secondary excoriations due to scratching ,as DH is intensely itchy.

Intact tense bullae on the elbow.

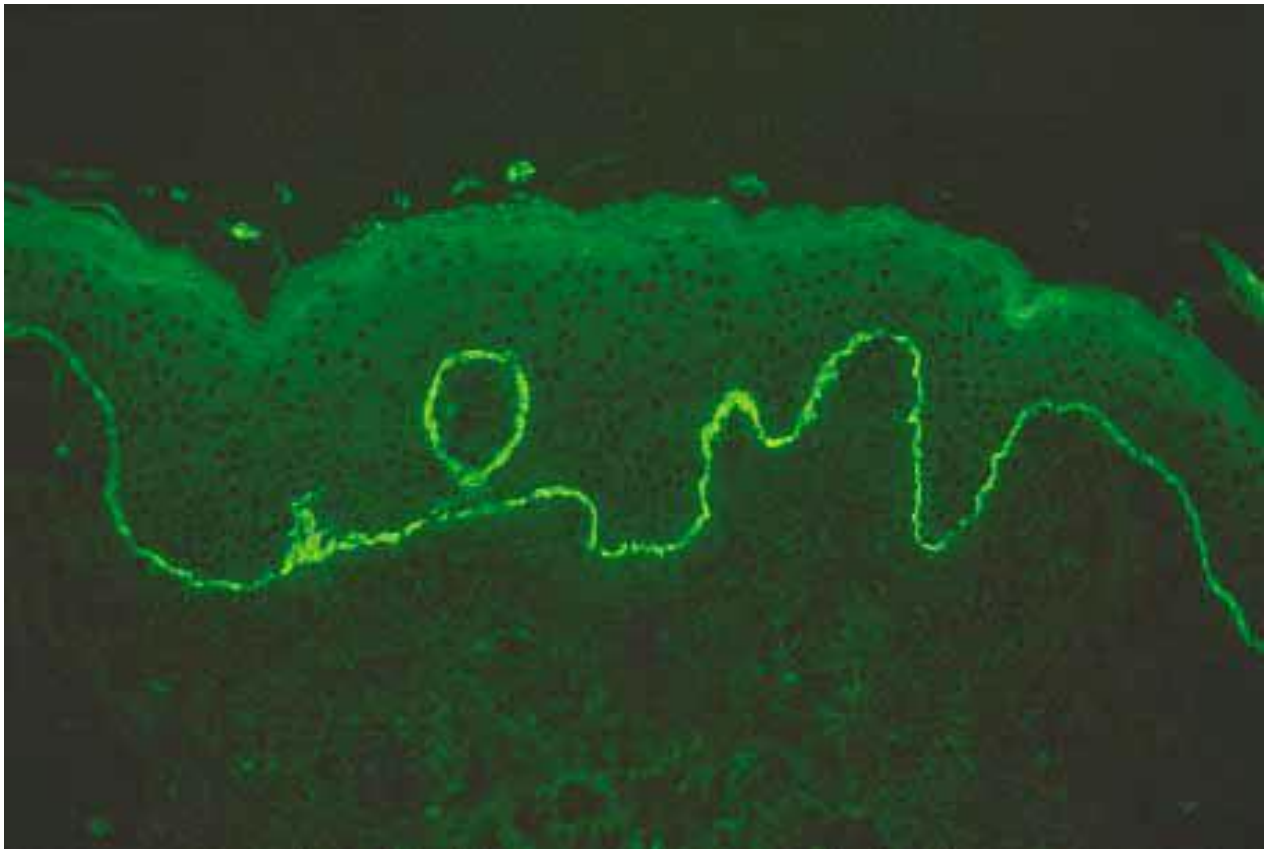
Investigations Immunoflourescence

Disorder	Direct Immunoflourescence	Indirect Immunoflourescence
Pemphigus	•IgG •Intercellular deposition in epidermis. •Fishnet pattern.	•+ve in 100 % of patients. •IgG to cell surface of epidermal cells of substrate tissue. •Titres correlate with disease activity.
Bullous pemphigoid	•C3 and IgG. •Dermoepidermal junction. •Linear.	•+ve in 70 % of patients. •IgG at the basement membrane of substrate tissue •Titres do not correlate with disease activity.

Pemphigus Vulgaris

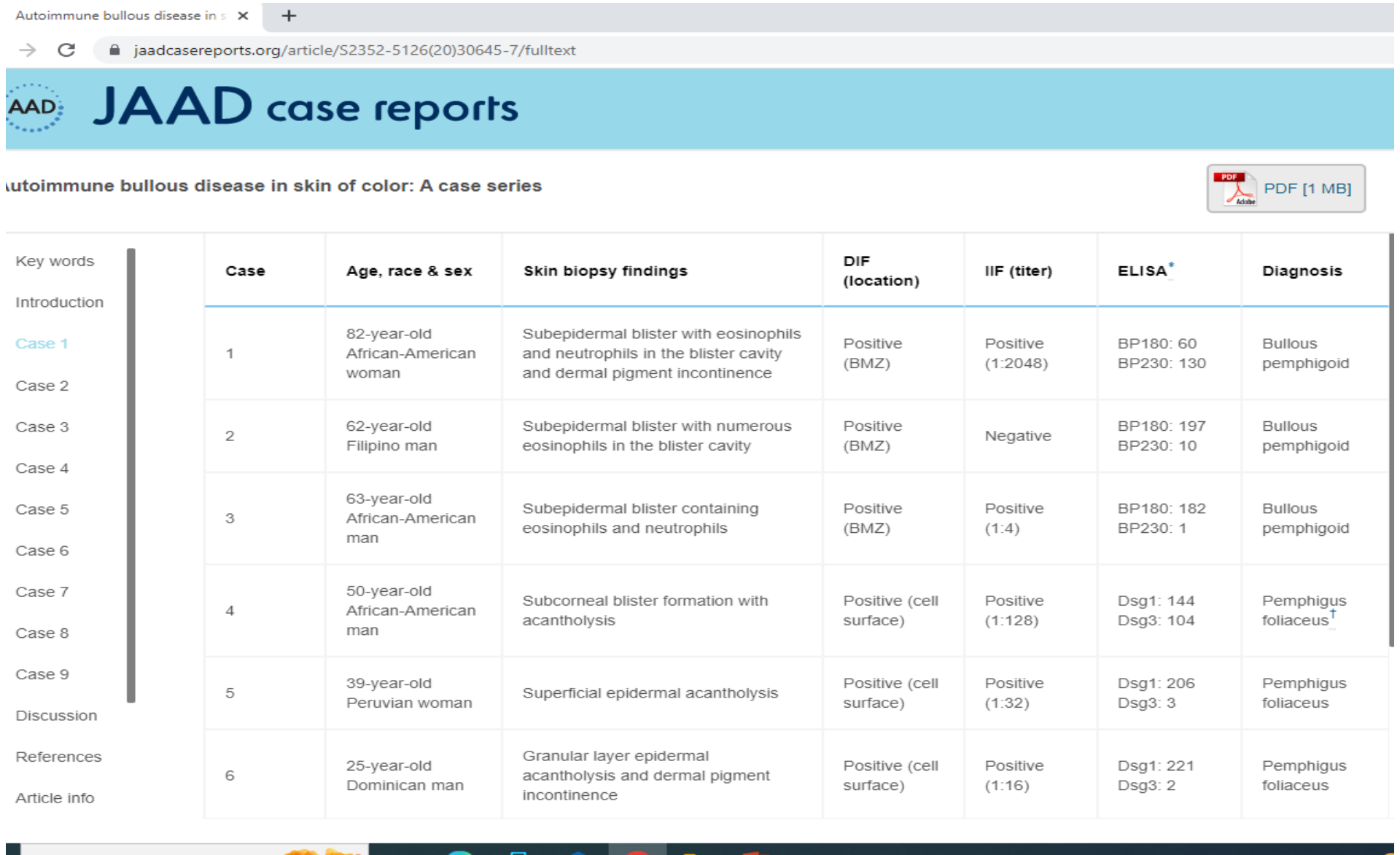


Bullous Pemphigoid



VERTICAL INTEGRATION

RESEARCH



Autoimmune bullous disease in s

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Key words
Introduction
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Discussion
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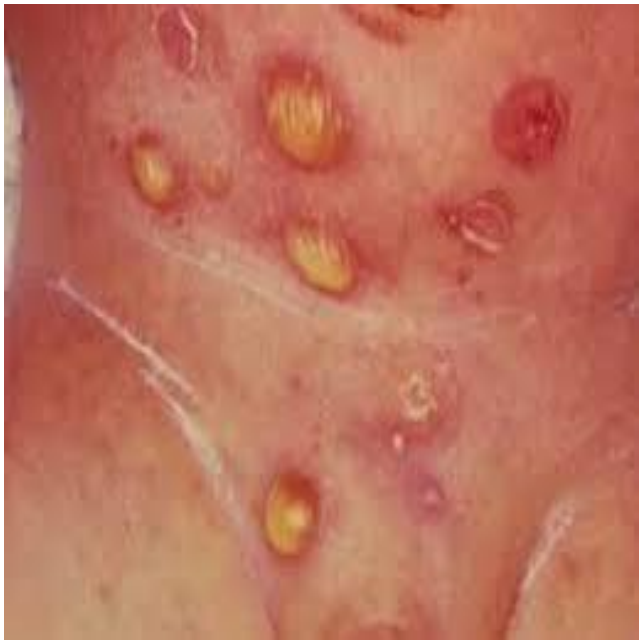
Article info

Case	Age, race & sex	Skin biopsy findings	DIF (location)	IIF (titer)	ELISA*	Diagnosis
1	82-year-old African-American woman	Subepidermal blister with eosinophils and neutrophils in the blister cavity and dermal pigment incontinence	Positive (BMZ)	Positive (1:2048)	BP180: 60 BP230: 130	Bullous pemphigoid
2	62-year-old Filipino man	Subepidermal blister with numerous eosinophils in the blister cavity	Positive (BMZ)	Negative	BP180: 197 BP230: 10	Bullous pemphigoid
3	63-year-old African-American man	Subepidermal blister containing eosinophils and neutrophils	Positive (BMZ)	Positive (1:4)	BP180: 182 BP230: 1	Bullous pemphigoid
4	50-year-old African-American man	Subcorneal blister formation with acantholysis	Positive (cell surface)	Positive (1:128)	Dsg1: 144 Dsg3: 104	Pemphigus foliaceus†
5	39-year-old Peruvian woman	Superficial epidermal acantholysis	Positive (cell surface)	Positive (1:32)	Dsg1: 206 Dsg3: 3	Pemphigus foliaceus
6	25-year-old Dominican man	Granular layer epidermal acantholysis and dermal pigment incontinence	Positive (cell surface)	Positive (1:16)	Dsg1: 221 Dsg3: 2	Pemphigus foliaceus

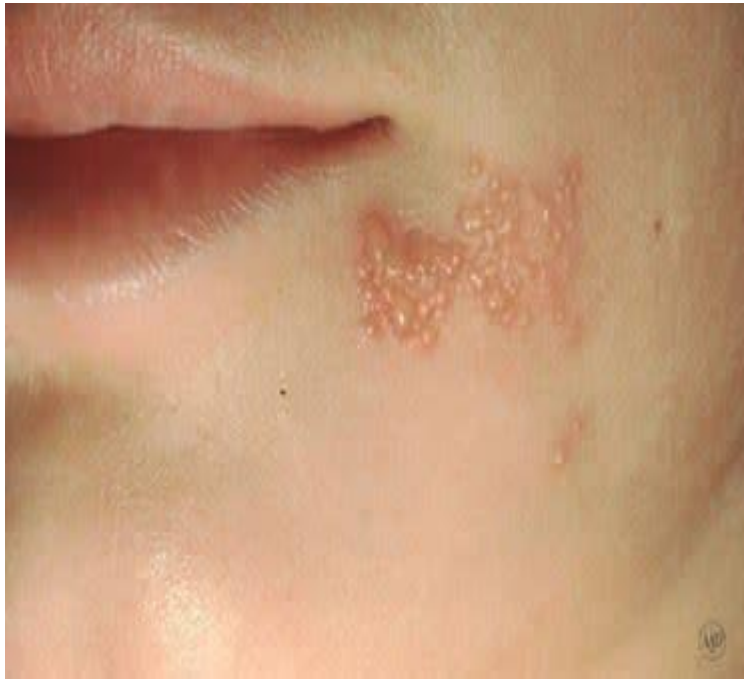
VERTICAL INTEGRATION

FAMILY MEDICINE

BULLOUS IMPETIGO



HERPES SIMPLEX



VERTICAL INTEGRATION

BIOETHICS

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Ethical Use of Topical Corticosteroids

Abir Saraswat

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Abstract

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Dermatologists rely very heavily on corticosteroids for treating many common dermatoses. Concerns about their incorrect use are widely expressed both in lay public and specialist discourse. From the point of view of medical ethics, issues of autonomy, beneficence and non-maleficence are all raised frequently when we prescribe topical corticosteroids to our patients. We need to be aware of situations when conflicts between these issues arise and have a clear thought process about resolving them. This can only be achieved if we have a thorough understanding of the skin disease being treated coupled with expertise in the use of the varied potencies and available dosage forms of topical corticosteroids. A good understanding of human psychology and effective communication is also needed to use these agents optimally.

Keywords: *Adverse effects, medical ethics, topical corticosteroids*

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Thank You