433 Teams

DERMATOLOGY

Lecture (17)

CUTANEOUS MANIFESTATIONS OF SYSTEMIC DISEASES

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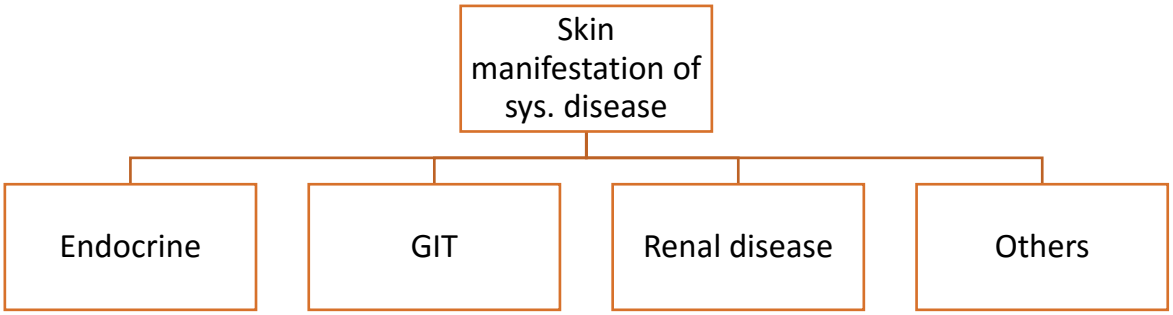
Main Title

Content of lecture:

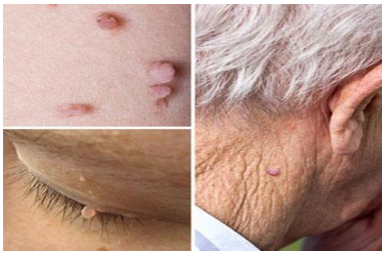
- To highlight the relation between skin manifestations and common systemic disorders.
- To understands various skin clues and their importance in investigating and managing different systemic diseases



#Skin and endocrine system:

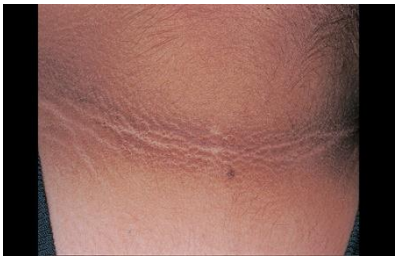
A. Diabetes mellitus.
B. Thyroid diseases.
C. Cushing's syndrome.
D. Addison's disease.

A- Diabetes mellitus:



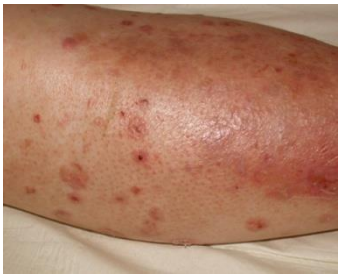
-Skin tags

small, pedunculated, soft papules on the eyelids, the neck, and the axillae. Mostly associated with obesity and insulin Resistance. If numerous usually on top of acantohsis nigricans.



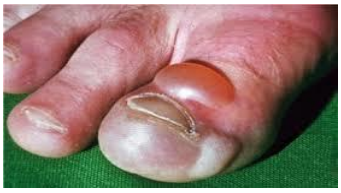
-Acanthosis nigricans

hyperpigmented, velvety **plaques** in body folds. Increased insulin, which binds to insulin-like growth factor receptors to stimulate the growth of Keratinocytes and dermal Fibroblasts. Treatment is by weight reduction and decrease insulin resistance.



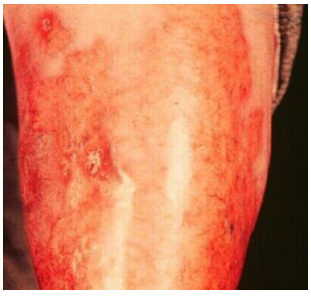
-Diabetic dermopathy

Very common, atrophic, hyperpigmented **papules** and **plaques** on Shins. Men are affected more often than women. Possibly related to diabetic neuropathy and vasculopathy. No effective treatment, but it does improve with diabetic control.



-Bullous diabeticorum

Rare. Spontaneous blistering of the hands and feet. Heals without scaring.

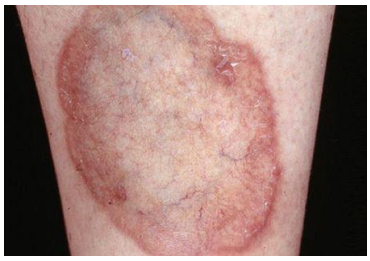


-Thickening of skin

Thickening of the hands with tiny papules on fingers and stiff joints.

Pebbled knuckles (or Huntley papules) are multiple minute papules, grouped on the extensor side of the fingers, on the knuckles, or on the periungual surface Generalized asymptomatic thickening of the skin (diabetic stiff skin) Scleredema on upper back and neck.

-Necrobiosis lipoidica diabeticorum



Yellow atrophic plaques on the shins. Sometimes they ulcerate.

Histopathology: shows tiered granulomatous Reaction.

Treatment: topical, intralesional steroids, tacrolimus, phototherapy, cyclosporine, and rarely surgery.

-Bacterial and fungal infections

Pyodermic infections such as impetigo, folliculitis, carbuncles, furunculosis, ecthyma, and erysipelas can be more severe and widespread in diabetic patients.

Erythrasma, caused by *Corynebacterium minutissimum mostly on axillae and groin.*

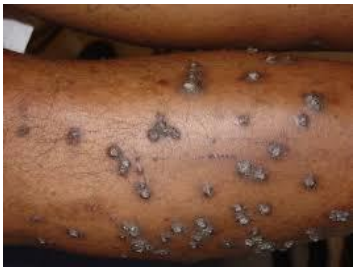
malignant otitis externa, often caused by *Pseudomonas aeruginosa*.

Tinea pedis (can lead to cellulitis) and onychomycosis.

Candidal infections like perleche on corners of mouth, and on vulva.

Rare infections like mucormycosis by Phycomycetes and anaerobic cellulitis by *Clostridium* species.

-Perforating dermatosis



Pruritic hyperkeratotic papules on the legs and trunk.

Histopathology shows transepidermal elimination of collagen and/or elastin.

Common in patients with diabetes and renal failure.

treatments include topical keratolytics, phototherapy, topical and systemic retinoids, topical and intralesional steroids, oral antihistamines, and cryotherapy.

B- Thyroid disorders

Hyperthyroidism

- Pretibial myxedema: is the most characteristic features of thyrotoxicosis appearing as shiny waxy papules and plaques having orange-skin appearance on the chin of the tibia.
- Warm skin and increased sweating and pruritus.
- Premature hair graying, alopecia with fine soft thinned scalp hair.
- Hyperpigmentation or vitiligo.
- Brittle nails.

Hypothyroidism

- Cold, pale and dry skin and pruritus.
- A yellowish hue to the skin due to carotenaemia.
- Slow growing ridged and brittle nails.
- Delayed wound healing.

C- Cushing's syndrome

caused by prolonged exposure to high levels of plasma glucocorticoid, adrenocortical hyperplasia, benign or malignant adrenal tumours, ectopic ACTH syndrome – secretion of ACTH by malignant or benign tumours arising in structures other than the pituitary or adrenal glands, exogenous steroid administration.

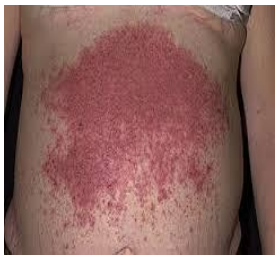
- Acne and hirsutism.
- Clitromegaly and male pattern alopecia (Hamilton pattern).
- Striae.
- Easy bruising and purpura.
- Moon face and buffalo hump with fat redistribution.
- Telangectasia on face.
- Poor wound healing.

D- Addison's disease

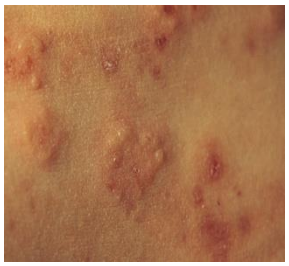
Adrenocortical hypofunction

- Hyperpigmentation at Sun exposed skin, sites of trauma, axillae, palmar creases, old scars ,nevi and mucous membranes.
- Diffuse pigmentation on skin and mucous membranes.
- Melanocytes stimulation by ACTH

#Gastrointestinal diseases

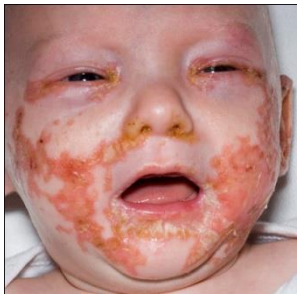


1- Dermatitis herpetiformis



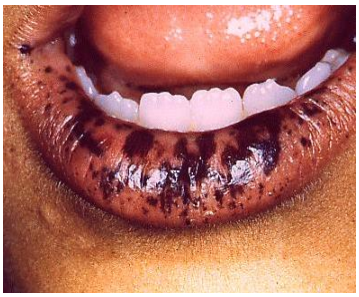
- Small severely pruritic vesicular lesions found in a symmetric distribution of both upper and lower extensor surfaces, buttocks and the scalp.
- direct immunofluorescence finding is granular deposition of **IgA** within the dermal papillae.
- celiac disease (also known as gluten-sensitive enteropathy and celiac sprue) are caused by the inability to absorb gluten from the diet.
- Treatment: gluten-free diet and dapsone.

2- Acrodermatits enteropathica



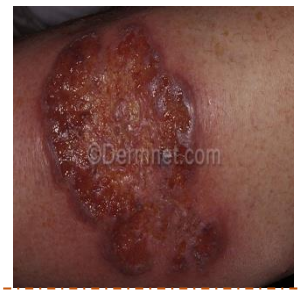
- ✓ a rare autosomal recessive disorder that impairs dietary zinc absorption in the jejunum and ileum.
- ✓ presents in infants several weeks after breastfeeding is discontinued.
- ✓ characterized by diarrhea, inflammatory rash, and hair loss.
- ✓ scaly, erythematous patches and plaques similar to atopic dermatitis, but progress to vesicles, crusts, erosions, and pustules on acral areas, perioral and perianal areas.
- ✓ Treatment by zinc supplementation for life.

3- Peutz Jeghers syndrome



- autosomal dominant disorder.
- mucocutaneous hyperpigmentation together with GI polyposis.
- The skin findings first appear in infancy or early childhood and involve brown macules on the lips and buccal mucosa.
- multiple hamartomatous polyps occurring most commonly in the jejunum.
- 2-3% of patients develop GI carcinoma during their lifetimes.

4- Pyoderma gangrenosum



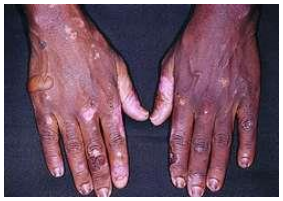
- a painful, ulcerative lesion with a well-defined, undermined violaceous border.
- start as small pustules, which subsequently burst and expand to form the larger noninfectious ulcer.
- Positive pathergy test.

Surgery is contraindicated

- Mostly associated with ulcerative colitis. Also with Crohn's disease, rheumatoid arthritis, and leukemia.

5- Porphyria cutanea tarda

- ✓ most common porphyria occurring in adults.
- ✓ skin photosensitivity with increased skin fragility, facial hypertrichosis, blisters, scarring with milia formation, and skin hyperpigmentation on the hands and other sun-exposed areas.
- ✓ results from the decreased activity of the enzyme uroporphyrinogen decarboxylase.
- ✓ Associated with Hep C virus.
- ✓ Treatment by removal of possible triggers, including iron supplementation, alcohol, and estrogens. Also by phlebotomy and hydroxycholorquine



6-Hemochromatosis

- a disorder of iron overload leading to excess deposition in multiple body organs.
- metallic gray or bronze-brown color that is generally diffuse.
- skin atrophy, ichthyosis, partial hair loss (most often in the pubic region), and koilonychia.
- cirrhosis may develop, and might lead to hepatocellular carcinoma.
- treatment involves phlebotomy and chelating agents.

7- Liver Cirrhosis

Clinical features: Some of the associated abnormalities are the following:

• Pruritus: this is related to obstructive jaundice and may precede it.

• Pigmentation with bile pigments and sometimes melanin.

• Spider naevi: these are often multiple inchronic liver disease.

• Palmar erythema.

• White nails: these associate with hypo-albuminaemia.

• Porphyria cutanea tarda.

• Xanthomas: in primary biliary cirrhosis.

• Hair loss

• Generalized asteatotic eczema: it may occur in alcoholics with cirrhosis who have become zinc deficient.

#Renal diseases

Xerosis: occurs in 50-92% of the dialysis population.

Some patients may develop acquired ichthyosis.

the exact cause of xerosis in ESRD remains unknown.

Many patients respond to routine use of emollients

Pruritus: affects 15-49% of patients with chronic renal failure and 50-90% of the dialysis population.

Uremia is the most common metabolic cause of pruritus.

Cutaneous manifestations of pruritus include excoriations, prurigo nodularis, and lichen simplex chronicus.

Pruritus typically resolves after transplantation.

Treatment include sedating antihistamines, emollients, phototherapy, thalidomide, and gabapentin .

Half and half nails: occur in around 40% of patients on dialysis.

Kidney transplant usually resolve this sign.

Usually involve fingernails.

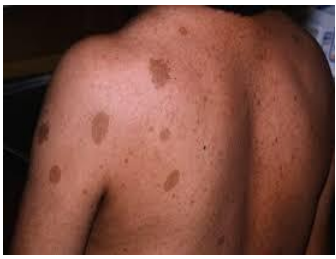
Nephrogenic systemic fibrosis: mostly seen in ESRD and dialysis patients.

Presents as thick, indurated plaques on the extremities and the trunk similar to scleroderma.

gadolinium might have a role in the pathogenesis of this condition.

Treatment includes immunosuppressive agents, phototherapy, topical steroids, retinoids, and photophoresis.

#Neurocutaneous Disorders



- ✓ Autosomal dominant
- ✓ Café-au-lait macules(light brown) (PIC)
- ✓ Neurofibromas (soft pink or skin- colored papules and nodules)
- ✓ Axillary freckling(Crowe sign), the presence of the macules on the axillae.
- ✓ Optic glioma
- ✓ Lisch nodules (iris hamartoma, seen by slit-lamp examination)
- ✓ Associated with Neurological complications e.g. tumors, seizures and mental retardation.

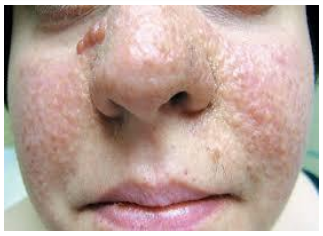
-Tuberous Sclerosis (Epiloia) :

Epi = Epilepsy

Loi = Low intellegence

A= adenoma sebaceoum

Skin Features :



- ❖ Adenoma sebaceoum (anigofibroma): red papules around the nose and on chin
- ❖ Ash-leaf hypopigmention: oval area of hypopigmention ***This is the earliest sign of TS***
- ❖ Periungal fibroma: multiple papules & nodules around the nail
- ❖ Shagreen patch: skin colored plaque on the trunk with “orange-peel” surface

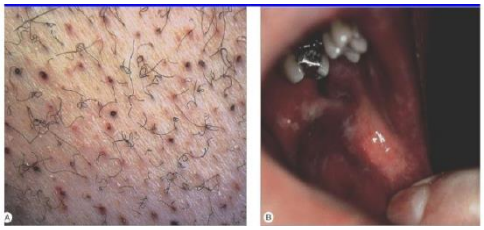
#Others

Behcet's Syndrome:

- Oral ulcer (the most common).
- Genital ulcers (mainly scrotal).
- Iritis and arthoropathy.
- May have CNS involvement.

Scurvy :

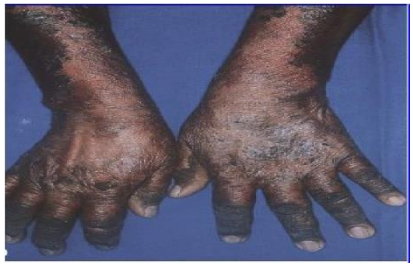
- Vitamin C deficiency, Bleeding gums
- Can cause teeth loss (permanent complication)
- Easy bruishing
- Diagnosis : Low ascorbic acid (Vit-C) level in Leukocyte



Pellagra: Nictonic acid deficiency

4 "D"s:

- Dermatitis (Photodermatitis)
- Death (if not treated)
- Diarrhea
- Dementia



Causes of generalized pruritus without skin lesions:

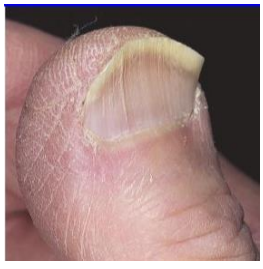
- Endocrine: DM, hypo& hyperthyroidsm
- Haematological: polycythemia rubra vera, iron def anemia Malignancy; e.g. Lymphoma ü Hepatic: primary biliary cirrhosis Renal: CRF, The commonest manifestation of CRF is pruritus Neurological : e.g. Tabes dorialis
- Others: Psychognic, Drugs, Idiopthaic.

Nails:

Koilonychia : Spoon- shaped appearance

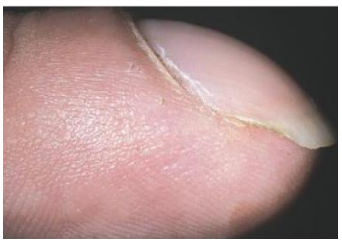
Causes:

- ✓ Iron deficiency anemia
- ✓ Thyroid disease
- ✓ Physological; early childhood
- ✓ Dermatosis: Lichen planus, Alopecia Areata and others



Clubbing :

Exaggeration of the normal nail curve associated with loss of the normal angle between nail and posterior nail fold.



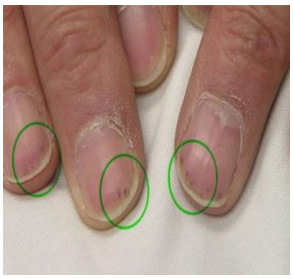
Causes:

- ✓ Thoracic: Lung abscess, Lung CA
- ✓ CVS: Congenital cyanotic heart disease
- ✓ GIT:GI carcinoma, Inflammatory bowel disease
- ✓ Endocrine: Thyroid disease
- ✓ Idiopathic.

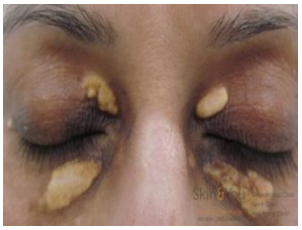
Splinter Haemorrhages :

Causes :

- Bacterial endocarditis
- Septic emboli
- CTD
- Trauma
- Idiopathic



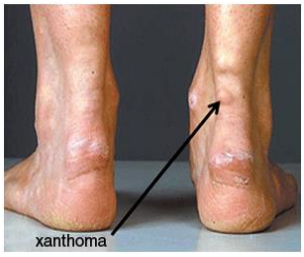
Hyperlipidemia:



-Xanthelasma palpebrarum: is the most common of the xanthomas asymptomatic and usually bilateral and symmetric.

Can be associated with any type of primary hyperlipoproteinemia and could be without hyperlipidemia.

often treated with topical trichloroacetic acid, electrodesiccation, laser therapy, and surgical excision.



-Tendinous xanthomas: commonly seen on the Achilles tendon followed by the hands, feet, elbows, and knees.

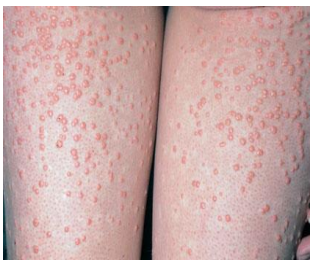
The least responsive xanthoma to treatment.

Mostly seen in patients with familial hypercholesterolemia.

-Tuberous xanthomas: are firm and nontender cutaneous and subcutaneous yellowish nodules on extensor surfaces.

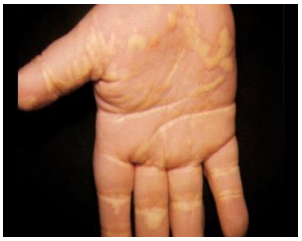
Mostly associated with familial dysbetalipoproteinemia.

May resolve after months of treatment with lipid lowering agents.



-Eruptive xanthomas: are painless, yellowish papules on an erythematous base that present as grouped lesions on trunk, elbows and buttocks.

- Usually associated with hypertriglyceridemia.
- Could be seen in poorly controlled diabetes and acute pancreatitis.
- Usually resolve in few weeks after therapy.



-Planar xanthomas are elevated cutaneous yellowish-orange deposits on palmar creases.

- Usually associated with familial dysbetalipoproteinemia.

Types of Xanthomas :

- **Eruptive**: small papules appear in crops over buttocks &extensors
- **Tendinous**: Nodules over tendons e.g. extensor tendons of hands & feet and Achilles tendon.
- **Palmar crease** xanthoma: on palms
- **Tuberous:** Papules & nodules over knees and elbows
  Xanthelasma: Bilateral symmetrical over both eyelids.

Done By: