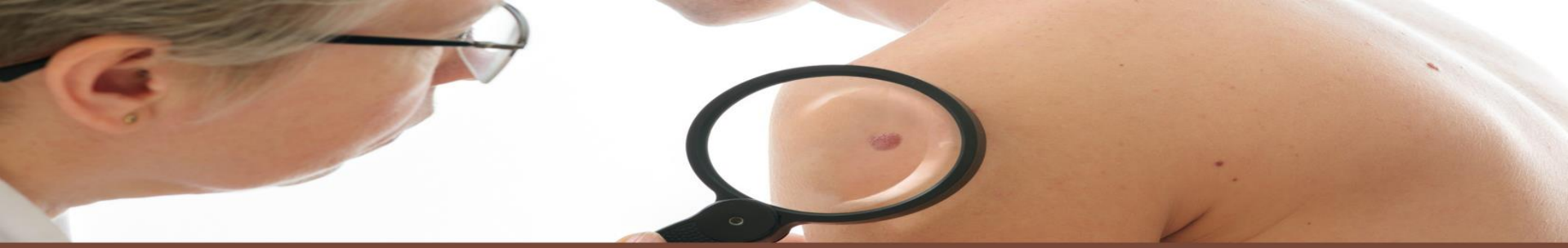


433 Teams

DERMATOLOGY

L8- cutaneous manifestations of systemic diseases

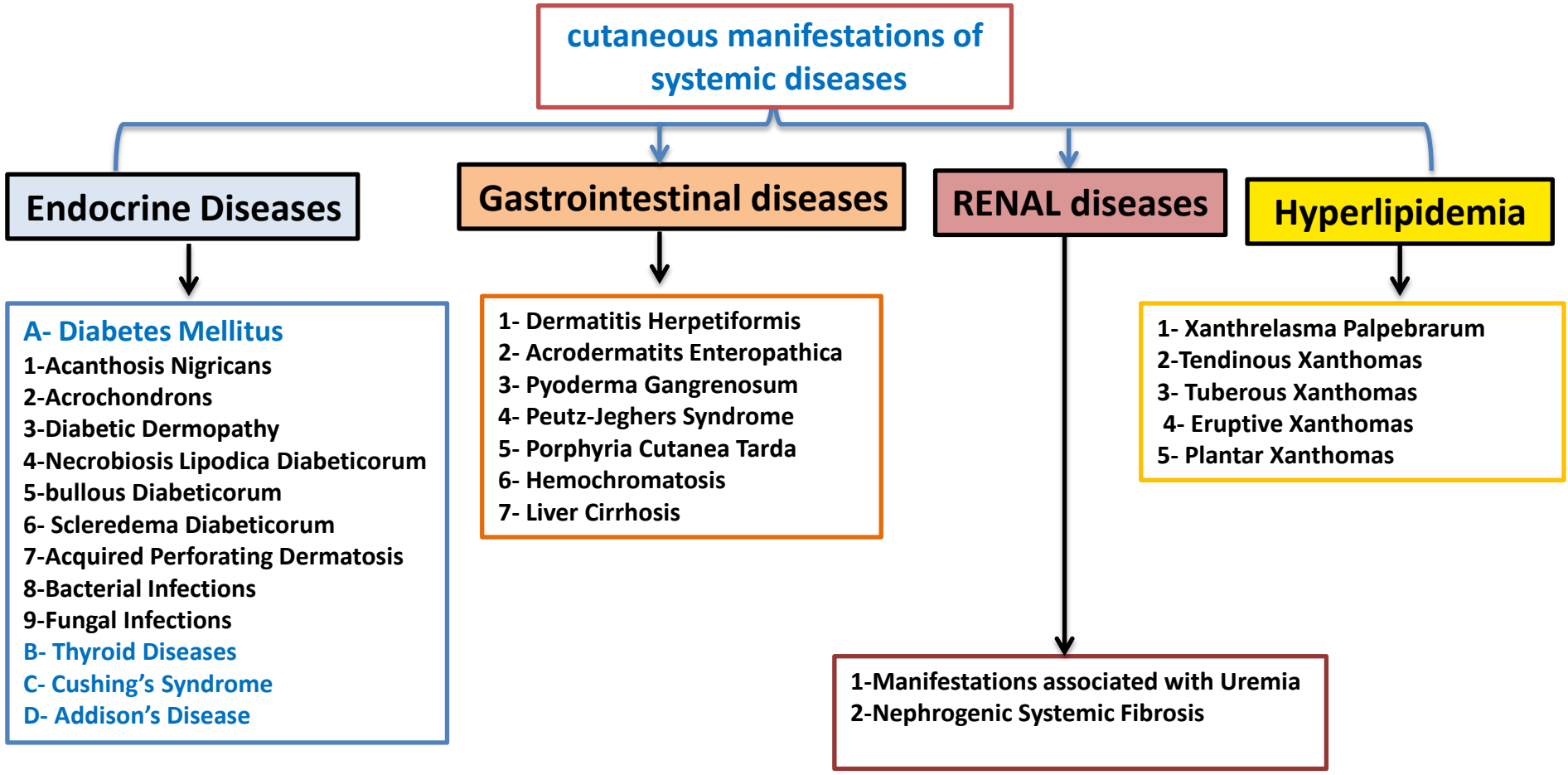
derm433team@gmail.com

جامعة الملك سعود
King Saud University

Objectives

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

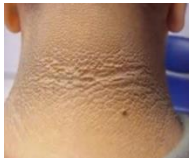
Mind Map



Cutaneous Manifestations Of Endocrine Diseases

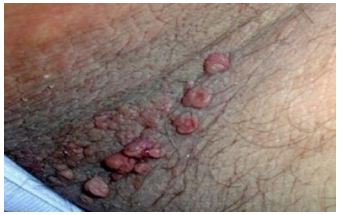
1- Diabetes Mellitus

1-Acanthosis Nigricans



- Hyperpigmented velvety plaques in body folds and creases.
- An indicator of insulin resistance.
  [High concentrations of insulin stimulate keratinocytes and dermal fibroblasts through high affinity binding to insulin-like growth factor 1 (IGF-1)]
- Treatment: Weight reduction and reducing insulin resistance.

2-Acrochondrons [Skin Tags]



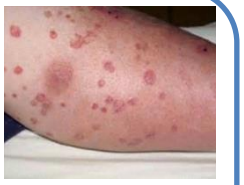
- Small, skin colored, pedunculated papules.
- Common sites: Neck, axilla, and the eyelids.

- Mostly associated with obesity and insulin resistance.
- Might be related to IGF activity
- If numerous, usually on top of acanthosis nigricans.

Cutaneous manifestations of Endocrine diseases

1- Diabetes Mellitus (CONT)

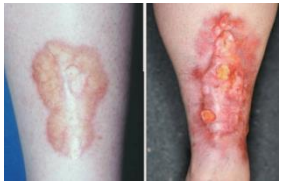
3-Diabetic Dermopathy



- Most Common cutaneous sign of diabetes
- Starts as red papules → atrophic, hyperpigmented papules and plaques on the shins.
- Possibly related to diabetic neuropathic and vascular complications.
- Usually don't require treatment, tend to go after few years with blood glucose control.

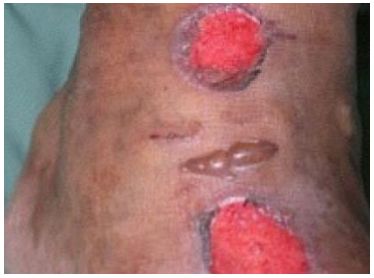
4-Necrobiosis Lipodica Diabeticorum

- Start as erythematous papules over the pretibial area, and evolve into yellowish brown plaques, with dilated blood vessels and central epidermal atrophy.
- Sometimes they ulcerate.
- Histopathology: granulomatous reaction.



- Treatment: Topical and intralesional steroids, tacrolimus, phototherapy, cyclosporine.

5- Diabetic Bullae or Bullae Diabeticorum (bullous Diabeticorum):



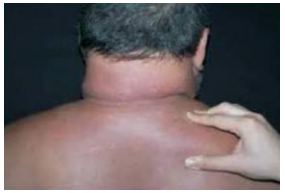
- Rare
- distinct marker of diabetes
- spontaneous blister- like lesions on the hands and feet
- heals Spontaneously without scarring

Cutaneous manifestations of Endocrine diseases

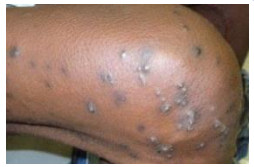
1- Diabetes Mellitus (CONT)

6- Scleredema Diabeticorum

- More common in males [obese with longstanding uncontrolled diabetes with complications (neuropathy, nephropathy retinopathy, atherosclerosis]
- Woody induration and thickening of the skin of the mid upper back, neck, and shoulders.
- Controlling diabetes DOES NOT affect the course of scleredema.



7-Acquired Perforating Dermatosis



- Occurs in patients with chronic renal failure, diabetes mellitus, or both.
- Characterized by the trans-epidermal elimination of collagen and elastic fibers. (Unknown pathogenesis)
- 2 to 10mm, firm, hyperkeratotic, often umblicated papules on the trunk and extremities.
- Treatment: topical keratolytics (e.g Urea), phototherapy, topical/systemic retinoids, topical/intralesional steroids, oral antihistamines, cryotherapy.

Cutaneous manifestations of Endocrine diseases

1- Diabetes Mellitus (CONT)

8-Bacterial Infections

- Usually caused by:
staphylococcus aureus and beta-hemolytic strept:
[Styes, folliculitis, fruncles, carbuncles, infections around the nails, impetigo, erysipelas, cellulitis, and necrotizing fasciitis.]

- Corynebacteria minustissimum:
cause→erythrasma

- Peusdomonas aeruginosa:
cause→ malignant otitis externa

9-Fungal Infections

- Candida: in warm, moist folds of the skin. → (Genitocrural folds, nail folds, web spaces of fingers and toes, mouth corners, armpits.)

- Tinea pedis, tinea cruris, tinea corporis, onychomycosis.

- Rhinocerebral mucormycosis:

Extensive, life threatening infection.
Begins in the nasal passages and spreads into the orbit and cerebrum.

Cutaneous manifestations of Endocrine diseases

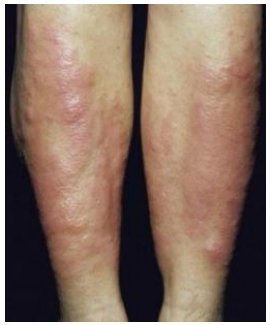
2- Thyroid Diseases

1-Hyperthyroidism

- Palmoplanta hyperhidrosis (Warm, moist skin.)
- Pruritis .
- Diffuse, non-scarring alopecia.
- Facial flushing
- Hyperpigmentation of the skin OR vitiligo
- Nail changes:
  [Plummer nails -> CONCAVE contour due to DISTAL onycholysis]

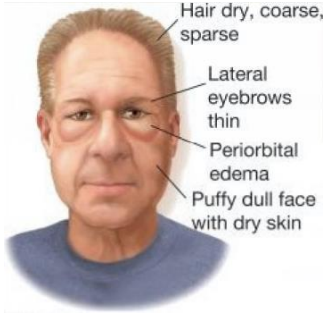
- Pretibial myxedema: the MOST CHARACTERISTIC feature of THYROTOXICOSIS.

Pretibial myxedema→presents as shiny waxy papules and plaques over the pretibial area



2-Hypothyroidism

- Coarse, rough, dry skin.
- Pallor.
- Pruritis.
- Carotenemia.*
- Diffuse hair loss.
- Loss of later 1/3 of the eyebrow.
- Myxedematous facies.
- Autoimmune diseases [Vitiligo, alopecia.]



***Carotenemia→** Carotenemia is a clinical condition characterized by yellow pigmentation of the skin
Due to increased beta-carotene levels in the blood

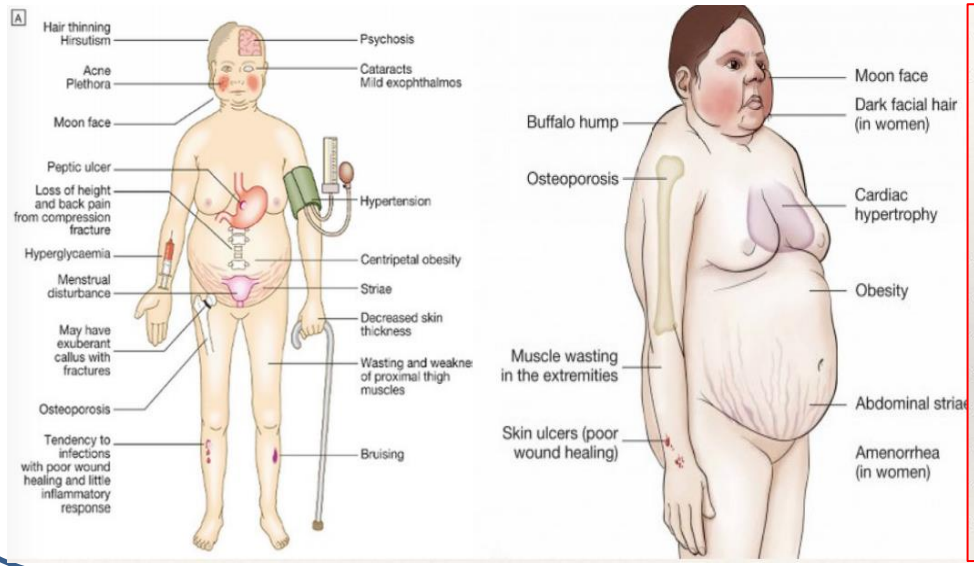
Cutaneous manifestations of Endocrine diseases

3- Cushing's Syndrome

- Occurs when there is an excess of cortisol in the body.
- Causes:
  - The pituitary gland is releasing too much ACTH.
  - The adrenals are releasing too much cortisol.
  - Patient is taking large doses of glucocorticoids. [Asthmatics, RA]

The difference between cushing's disease & cushing's syndrome :

- HIGH ACTH [seen in pituitary adenomas] → CUSHING'S DISEASE
- NORMAL or LOW blood ACTH [due to medications that contain steroids or ectopic ACTH syndrome, seen in lung cancer] → CUSHING'S SYNDROME

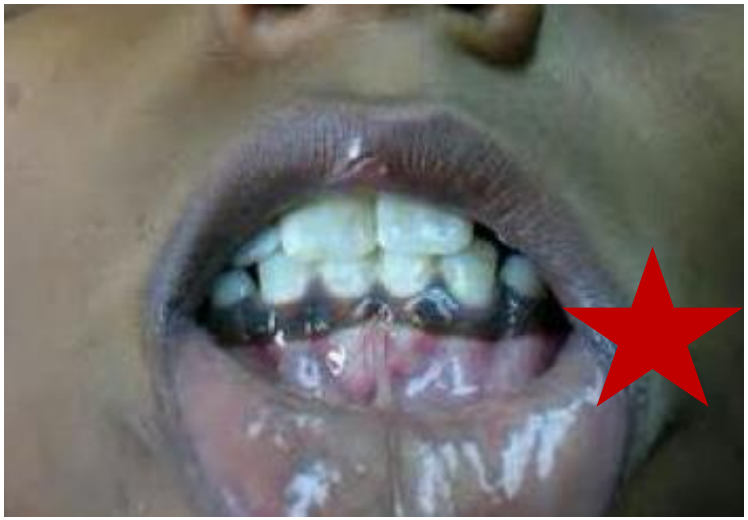


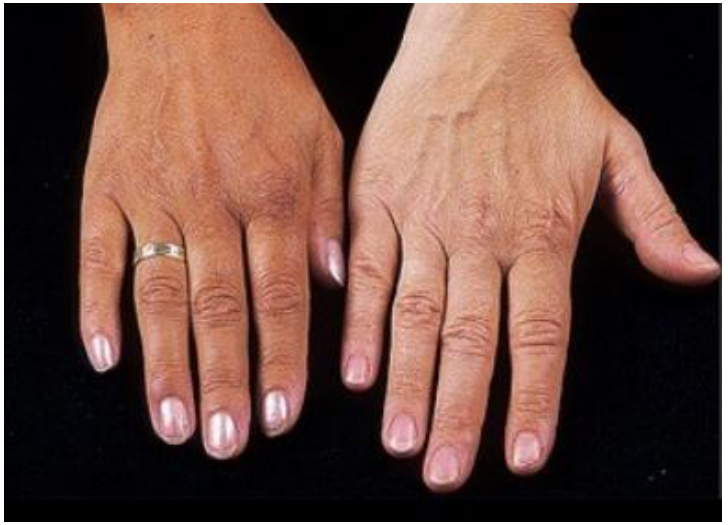
- Weight gain
- Skin changes -thin, fragile skin, increased susceptibility to infections, poor wound healing, striae, redistribution of fat (moon faces, buffalo hump, central obesity), hirsutism, acne
- Menstrual irregularities
- Muscle loss and weakness
- Osteoporosis
- Glucose intolerance
- High blood pressure
- Cardiovascular disease
- Psychological symptoms
- Hypercoagulability

Cutaneous manifestations of Endocrine diseases

4- Addison's Disease

- Destruction of ADRENAL CORTEX → Adrenocortical insufficiency.
- HALL MARK OF ADDISON'S: Hyperpigmentation of the skin and mucous membranes.
- Oral mucous membrane hyperpigmentation is PATHOGNOMONIC.
- Hyperpigmentation is caused by high levels or ACTH that bind to the MELANOCORTIN 1 receptors on the surface of dermal melanocytes.



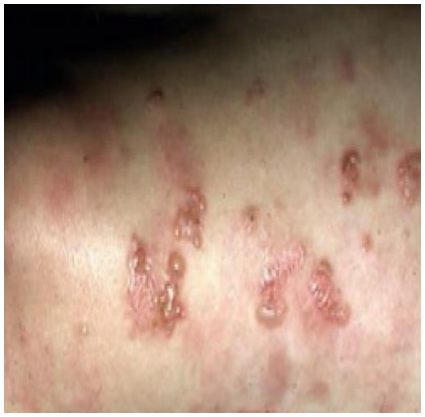
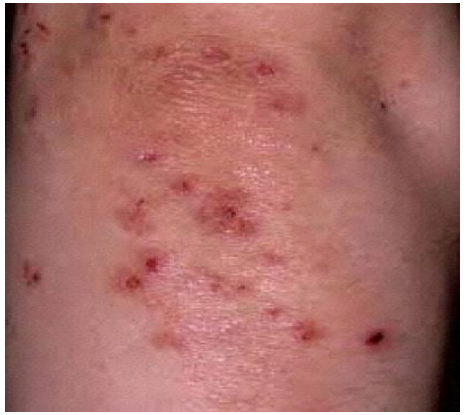
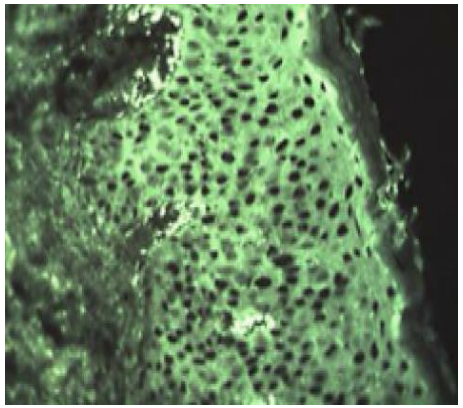
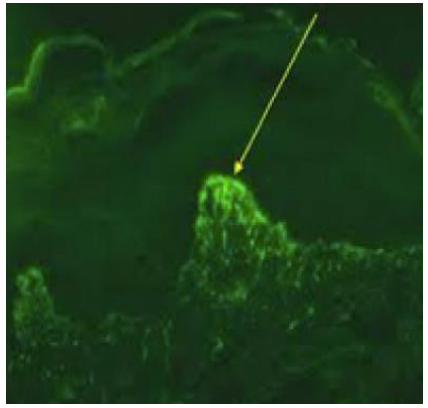


Cutaneous manifestations of Gastrointestinal diseases

1- Dermatitis Herpetiformis

- Autoimmune blistering disorder that is often associated with a GLUTEN SENSITIVE ENTEROPATHY. [Lesions are considered as the cutaneous manifestation of Celiac disease 'known as Celiac Disease of the Skin]
- Dietary intake of glutens makes it worse.
- Small, severely pruritic, clustered vesicular papules or plaques that are symmetrically distributed over the extensor surfaces [elbows, knees, buttocks and shoulders.]
- Direct immunofluorescence of precessional skin shows granular IgA deposits in dermal papillae.

The name herpetiformis is used because the skin lesions that develop in this disorder often resemble those seen in herpes-related infections, and not because dermatitis herpetiformis is caused by the herpes virus.

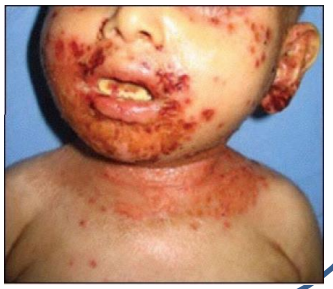





Cutaneous manifestations of Gastrointestinal diseases

2- Acrodermatits Enteropathica

Acrodermatits Enteropathica

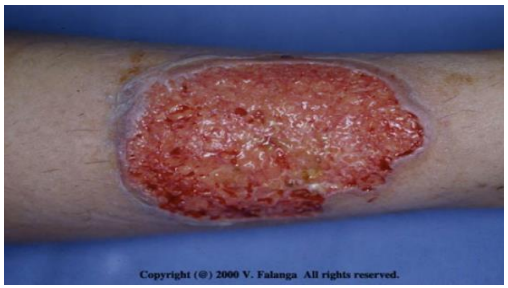
- Rare, autosomal recessive disorder that impairs zinc absorption in the jejunum and ileum.
- Features manifest when the infant is weaned from breast milk.
- It is a triad of.→ Periorificial and acral dermatitis, alopecia, and diarrhea.
- Scaly erthematous patches and plaques similar to atopic dermatitis, but progress to vesicles, crusts, erosions and pustules in the acral, perioral, and perianal areas.
- Treatment: life long dietary zinc supplementation.



3- Pyoderma Gangrenosum

Pyoderma Gangrenosum

- Painful ulcerative lesion with well-defined undermined violaceous border.
- Starts as a small red papule or pustule that subsequently burst and expand to form a large non-infectious ulcer.
- +ve pathergy test (Behcet's disease)
- Mostly associated with ulcerative colitis (mostly), also crohn's disease, rheumatoid arthritis, and leukemia.
- Surgery is contraindicated.



Cutaneous manifestations of Gastrointestinal diseases

4- Peutz-Jeghers Syndrome

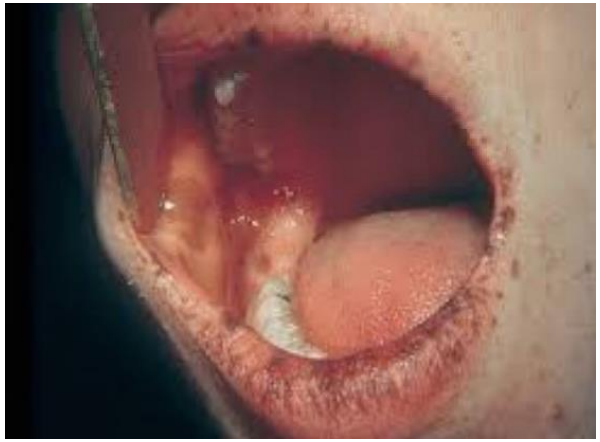
Peutz-Jehgers Syndrome

- Autosomal dominant disorder characterized by hyperpigmentation of the skin and mucous membranes along with intestinal hamartomatous polyps (mostly jejunum).

- The skin findings first appear in infancy.
- [Brown macules on the lips and buccal mucosa]

- Increased lifetime risk of intestinal and extra-intestinal malignancy.

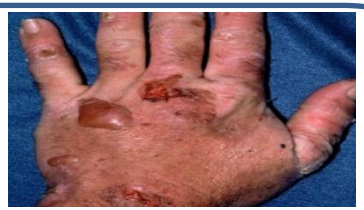
- Increased risk of malignancy in younger individuals.



Cutaneous manifestations of Gastrointestinal diseases

5- Porphyria Cutanea Tarda

[Dracula's or The Vampire Disease]



- **The most common prophyria in adults.**
- **Prophyria is a group of inherited metabolic disorder resulting from a deficiency of an enzyme in the heme production pathways. [overproducing of toxic heme precursors]**
- Porphyria cutanea tarda (PCT) is a term encompassing a group of acquired and familial disorders in which activity of the heme synthetic enzyme **uroporphyrinogen decarboxylase (UROD) is deficient**
- **Photosensitivity, skin fragility of sun exposed areas after mechanical trauma, leading to erosions and bullae typically on the hands and other sun exposed areas.**
- **Healing of crusted erosions and bullae leaves milia, hyperpigmentationm and atrophic scars.**
- **Facial hypertrichiosis.**
- **Frequently associated with hepatitis C infection.**
- **Treatment: the goal is to reduce serum ferritin levels to the lower limit of the reference range.**
  [avoid the followings: iron supplementations, phlebotomy, hydroxychloroquine, sun exposure.]

Cutaneous manifestations of Gastrointestinal diseases

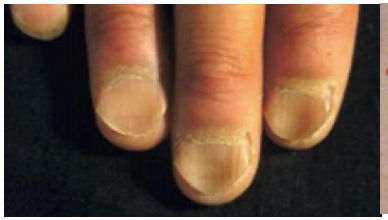
6- Hemochromatosis

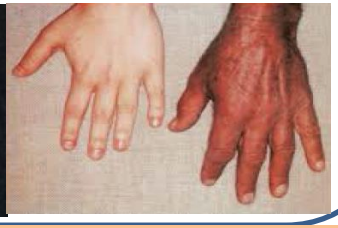
When the hepcidin level is abnormally low →iron overload

- Abnormal accumulation of iron in several organs leading to organ toxicity [Autosomal recessive disorder]
- Cutaneous pigmentation is one of the earliest signs of the disease, on sun exposed areas, particularly the face, brownish bronze or slate gray.

- Other skin changes: Icythosis-like changes, koilonychia, and hair loss mostly over the pubic area [due to the rise in the ratio of estrogen to androgen in males].

  Treatment: the goal is to remove the iron before irreversible damage.
  - Phelebotomy
  - Chelation therapy
  - Surgery indications:
    1. End-stage liver disease/Hepatocellular carcinoma.
    2. Severe arthropathy.

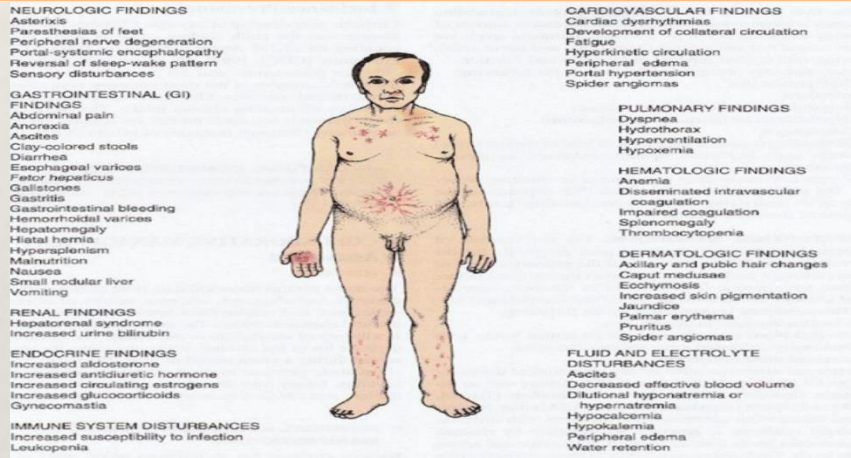




7- Liver Cirrhosis

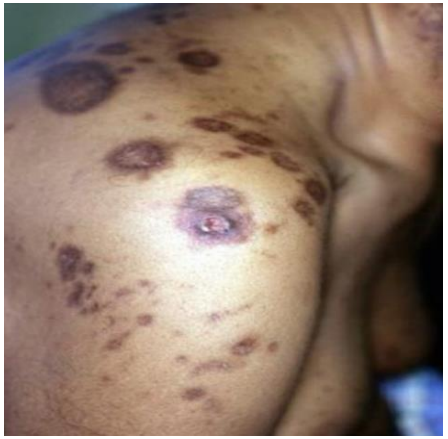
- Jaundice
- Pruritis
- Spider angioma
- Palmar erythema
- Purpura

- Petechiae
- Caput medusa
- Loss of body hair



Cutaneous manifestations of RENAL diseases

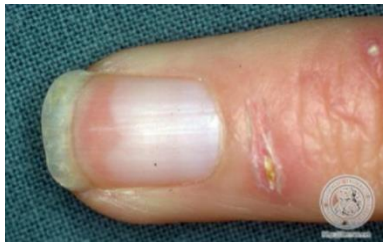
1-Manifestations associated with Uremia



- **Xerosis**
- Occurs in 50-90% of dialysis patients.
- Some develop acquired ichthyosis.
- The exact cause of xerosis in ESRD is unknown.
- Many patients respond to routine use of emollients.

- **Pruritus**
- Uremia is the most common metabolic cause of pruritis.
- Affects 15-49% of patients with CRF, and 50-90% of the dialysis population.
- (Excoriations, prurigo nodularis, lichen simplex chronicus)→ caused by scratching.
- Resolves after transplantation
- Treatment:
  Sedating anti-histamines, emollients, phototherapy, thalidomide, gabapentin.

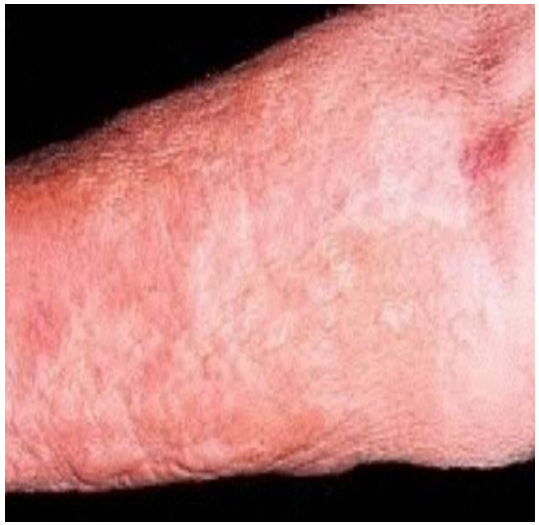
- **Half-and-half nails**
- NOT pathognomic for renal failure, but it occurs in around 40% of patients on dialysis.
- Disappears several months after renal transplantation.
- Dark reddish brown distal band and a white proximal band.
- More commonly affects fingernails but could be seen on toenails.

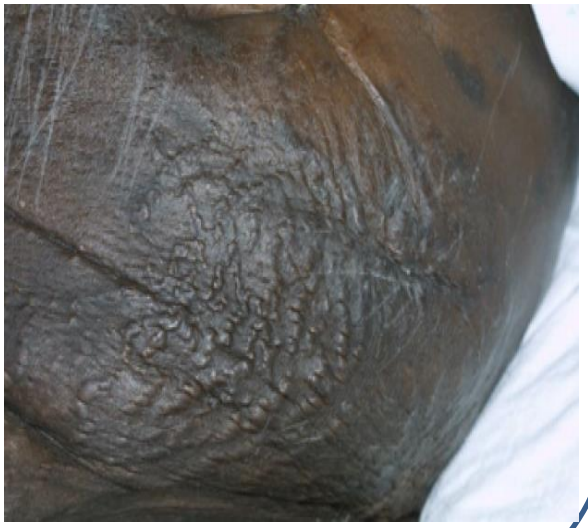


Cutaneous manifestations of RENAL diseases

2-Nephrogenic Systemic Fibrosis

- It's a disease of fibrosis of the skin and internal organs similar to scleroderma.
- Caused by Gadolinium exposure, used in imaging patients who have renal insufficiency.
- Large areas of thick, indurated skin with fibrotic nodules and plaques on the extremities and trunk.
- Treatment:
  - Extracorporeal photopheresis
  - Immunosuppressive therapy
  - Phototherapy
  - IVIG
  - Topical steroids.





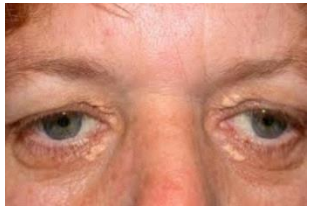
Cutaneous manifestations of Hyperlipidemia

1- Xanthrelasma Palpebrarum

Xanthelasma Palpebrarum

- **Most common cutaneous xanthoma**
- **Occurs most commonly near the inner canthus, more often on the upper eyelid.**
- **Asymptomatic, and usually bilateral and symmetrical.**
- **Can be associated with any type of primary hyperlipoproteinemia, but can also occur without hyperlipidemia.**
- **Treatment:**

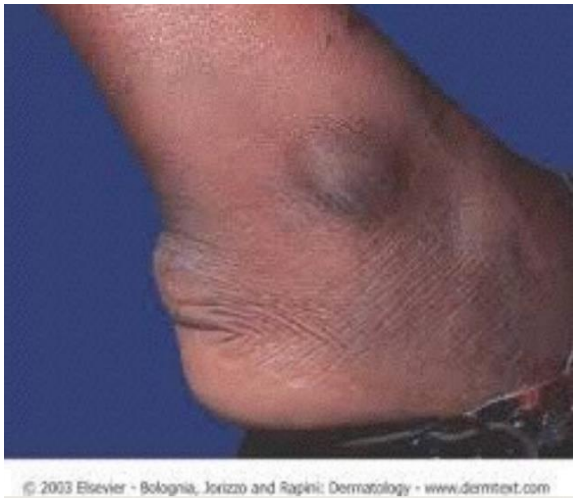
Surgical excision, CO2 laser ablation chemical cauterization (tricoloroacetic acid), electrodesiccation, cryotherapy.



2- Tendinous Xanthomas

Tendious Xanthomas

- **Commonly seen on the Achilles tendon, followed by the hands, feet, elbows, and knees.**
- **The least responsive to treatment.**
- **Mostly seen in patients with familial hypercholestrolemia.**

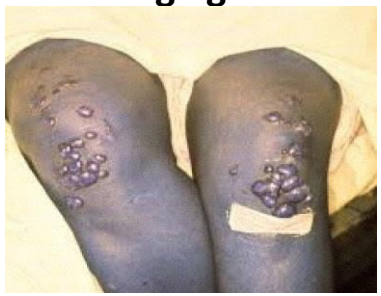


Cutaneous manifestations of Hyperlipidemia

3- Tuberous Xanthomas

Tuberous Xanthomas

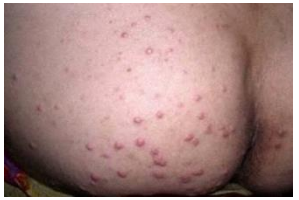
- Firm non-tender, cutaneous and subcutaneous yellowish nodules on the extensor surfaces.
- Mostly associated with familial dyslipoproteinemia.
- May resolve after months of treatment with lipid lowering agents.



4- Eruptive Xanthomas

Eruptive Xanthomas

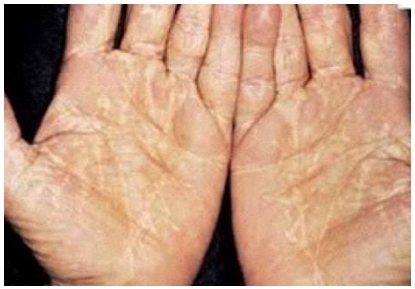
- Painless, yellowish papules on an erythematous base
- present as grouped lesions on: the trunk, elbows, and buttocks.
- Usually associated with hypertriglyceridemia.



- Could be seen in poorly controlled diabetes and acute pancreatitis.
- Usually resolves in few weeks after therapy.

5- Plantar Xanthomas

- Elevated cutaneous yellowish-orange deposits on the plantar creases.
- Usually associated with dysbetalipoproteinemia.



Done By:

Norah Alnaeim

Nawal Asiry

Revised by:

Nawal Asiry