DERMATOLOGY

Cutaneous manifestations of systemic diseases

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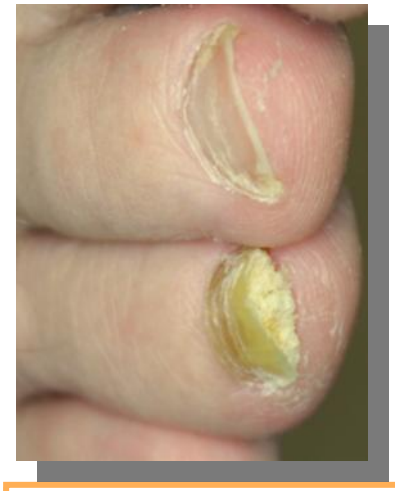
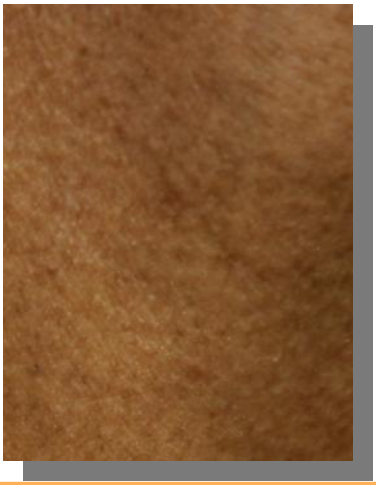
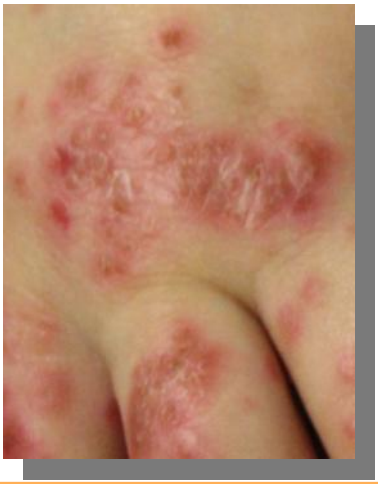
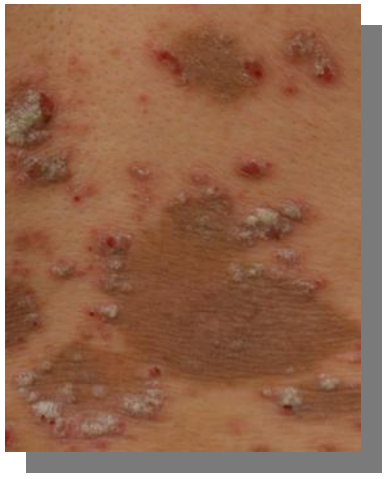





FROM SLIDES DOCTOR'S NOTES TEAM'S NOTES FROM BOOK IMPORTANT

1. Contents

2. Lecture Objectives

- **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.**

3.1- Diabetes Mellitus:

A-Skin tags:

Definition:

- They aresmall, pedunculated, soft papules on the eyelids, the neck, and the axillae.Most common found in the neck.

Etiology :

- They are mostly associated with obesity and insulin resistance.Increased insulin, which binds to insulin-like growth factor receptors to stimulate the growth of keratinocytes and dermal fibroblasts.

Clinical feature:

- If they are numerous , They will usually be on top of acantohsis nigricans.

B-Acanthosis nigricans:

Defenition :

- They are hyperpigmented, velvety plaques in body folds.

Etiology:

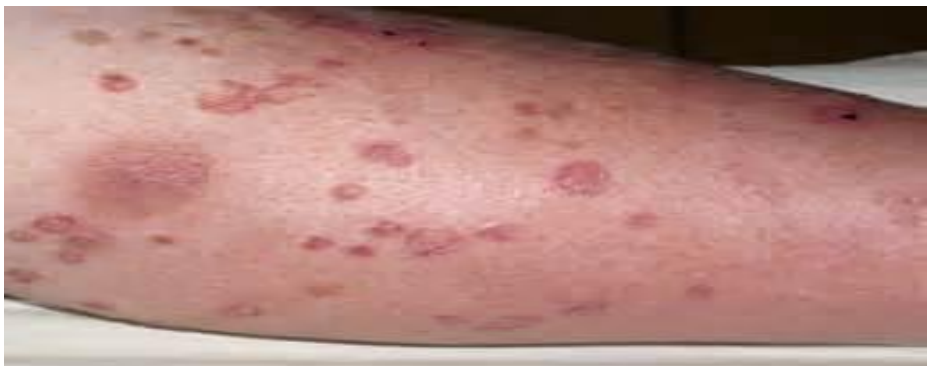
- Increased insulin, which binds to insulin-like growth factor receptors to stimulate the growth of keratinocytes and dermal fibroblasts, which leads to thickening of the epidermis.

Treatment:

- It is done by weight reduction and decrease insulin resistance+ metformin .(The more fatty cells the more needed insulin which bind to insulin like growth factor), It is very hard to treat by external procedures like lase skin exfoliation by chemicals.

3.2 Cutaneous manifestations of endocrine diseases:

C-Diabetic dermopathy:



Definition:

Atrophic, hyperpigmented papules and plaques on shins.

Clinical feature:

- It is very common
- It is harmless .it mostly causes cosmetic issues.
- Men are affected more often than women

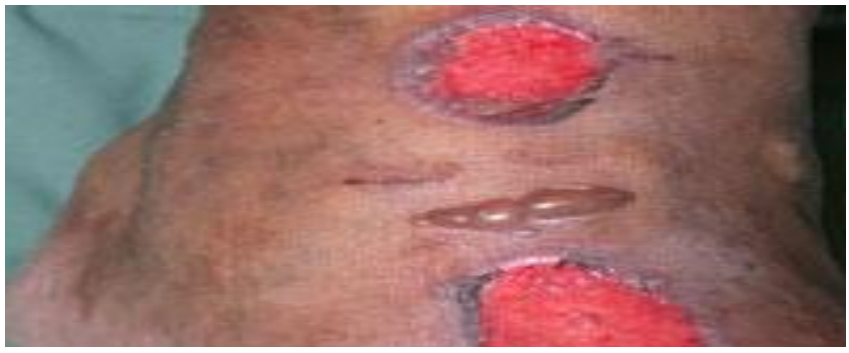
Etiology:

- Possibly related to diabetic neuropathy and vasculopathy.

Management:

- No effective treatment, but it does improve with diabetic control.

D-Bullous diabeticorum:



Definition:

They are Rare Spontaneous blistering of the hands and feet.

Prognosis:

- They Heal without scaring.

Management:

• No need for treatment Expect if there is erosions >>Occlusive ointment +Dressing

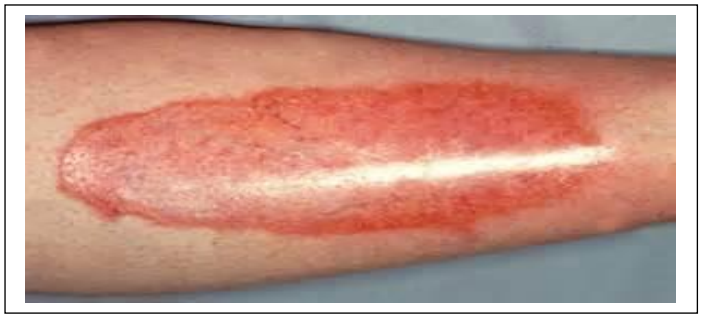
E-Thickening of skin:

Definition:

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

  Clinical presentation:
  - 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)
  - Scler**e**d**ema**(thinking of skin or fibrosis, you should differentiate from sclero**d**erma which is systemic sclerosis disease) on upper back and neck.
  - It like Acanthosis nigricans but without any skin discolouration And it is very Tiny

F-Necrobiosis lipoidica diabeticorum:



Definition:

- They are Yellow atrophic plaques on the shins.Sometimes they mayulcerate. May be unilateral or bilateral.

Histopathology:

- It shows tiered (multi-layers) **granulomatous reaction**. Not like TB or scrodiosis which are round shaped and associated with central necrosis.

Treatment:

- You can manage it with topical, intralesional steroids (If it is ulcerated), tacrolimus, phototherapy, cyclosporine, and rarely surgery.

G- Bacterial skin infections:

g-1 Pyogenic skin infection(Also called Pyodermal infectionsmostly caused by staph and streptococcus):

- Impetigo
- Folliculitis
- carbuncles
- Furunculosis

g-2Erythrasma:

- It caused by Corynebacterium minutissimum .It occurs mostly on axillae and groin.

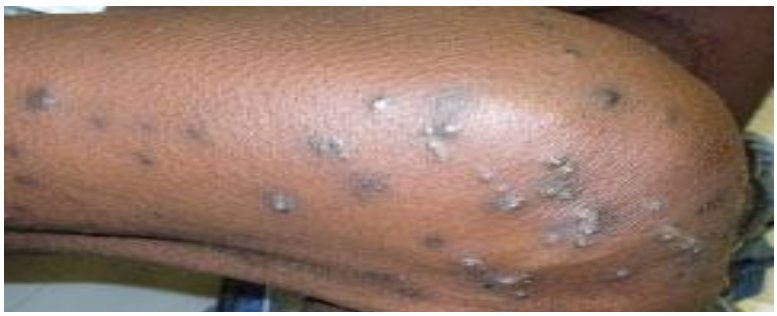
g-3 Malignant otitis externa

- It often caused by Pseudomonas aeruginosa.

H- Fungal skin infections:

- Tinea pedis 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
- Deep fungal infection usually they invade blood vessels especially in the face witch is very difficult to treat.

I- Perforating dermatosis:



Definition:

- Pruritic hyperkeratotic papules on the legs and trunk.

Histopathology:

- It shows transepidermal elimination of collagen and/or elastin.

Clinical pictures:

- Common in patients with diabetes and renal failure.

Treatment:

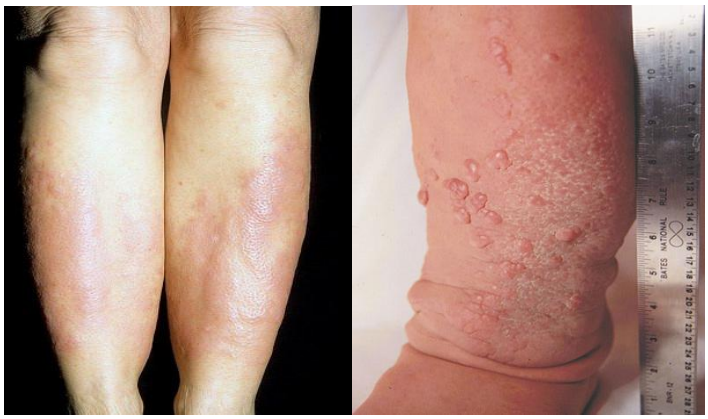
- include topical keratolytics, phototherapy, topical and systemic retinoids, topical and intralesional steroids, oral antihistamines, and cryotherapy.

3.2- Thyroid Diseases:

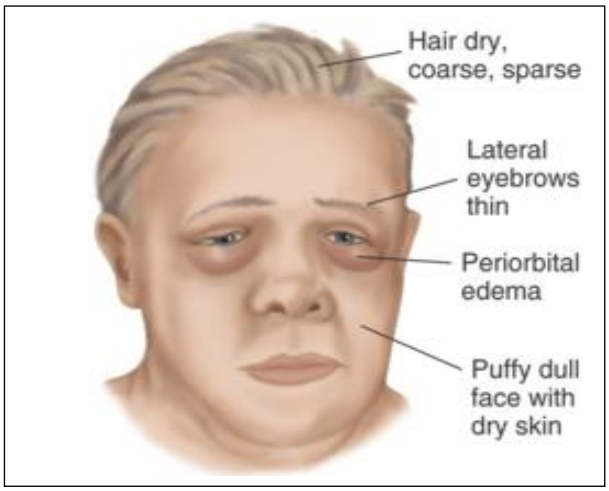
Thyroid disease is associated with changes in the skin, whichmay sometimes be the first clinical signs. There may beevidence of the effect of altered concentrations of thyroxine onthe skin, with changes in texture and hair growth.

3.2.1- 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.
- Pruritus
- Premature hair graying.
- Alopecia with fine soft thinned scalp hair.
- Hyperpigmentation or vitiligo.
- Brittle nails.



3.2.2- Hyporthyroidism:



A-Clinical Presentation:

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

Clinical signs of thyroid disease	
Hypothyroidism	**Hyperthyroidism**
Dry skin	Soft, thickened skin
Pale "ivory" skin	Diffuse pigmentation
Oedema of eyelids and hands	Pretibial myxoedema
Absence of sweating	Increased sweating (palms and soles)
Coarse, thin hair—loss of pubicaxillary, and eyebrow hair	Thinning of scalp hair
Brittle poorly growing nails	Rapidly growing nails
Purpura, bruising, andtelangiectasia	Palmar erythema and Facial flushing

Source: ABC of Dermatology, PAUL K BUXTON, Fourth Edition

3.3- Adrenal Diseases:

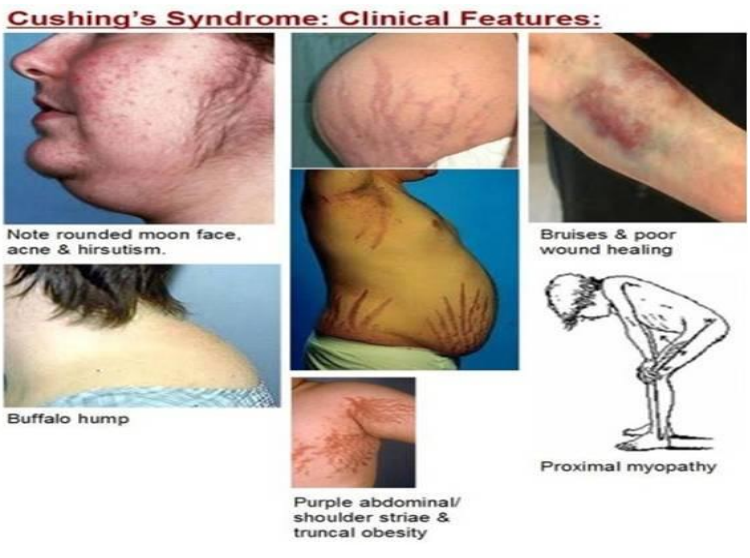
3.3.1-Cutaneous manifestation of Cushing's syndrome:

A-Etiology:

- It is caused by prolonged exposure to high levels of plasma glucocorticoid, which may due to one of the following:
  - 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.

B-Clinical cutaneous menifestations:

- Acne and hirsutism.
- Clitromegaly and male pattern alopecia.
- Striae.
- Easy bruising and purpura.
- Moon face and buffalo hump
- Telangectasia on face.
- Poor wound healing.



3.3.2- Cutaneous manifestation of Addison Disease :

A-Etiology:

- It caused by Adrenocortical hypofunction.

B-Clinical cutaneous menifestations:

- Diffuse pigmentation on skin and mucous membranes (Due to Melanocytes stimulation by ACTH).

4. Cutaneous manifestation of gastrointestinal diseases:

4.1- Dermatitis herpetiformis:

A-Definition:

- **Grossly** :Small severely pruritic vesicular lesions found in a symmetric distribution of both upper and lower extensor surfaces, buttocks and the scalp.
- **Microscopically**:direct immunofluorescence finding is granular deposition of IgA within the dermal papillae.

B-Etiology:

- celiac disease (also known as gluten-sensitive enteropathy and celiac sprue) are caused by the inability to absorb gluten from the diet.

C-Management:

- Gluten-free diet
- Dapsone.

4.2- Acrodermatits enteropathica:

A-Definition:

- It is a rare autosomal recessive disorder that impairs dietary zinc absorption in the jejunum and ileum.

B-Clinical menifestation:

- presents in infants several weeks after breastfeeding is discontinued.

- **characterized by diarrhea, inflammatory rash, and hair loss**.(Triad of acrodermatitis enteropathica)
- scaly, erythematous patches and plaques similar to atopic dermatitis, but progress to vesicles, crusts, erosions, and pustules on acral areas, perioral and perianal areas.

C-Management:

- Zinc supplementation for life.

4.3- Pyoderma gangrenosum:

A-Definition:

- It is a painful, ulcerative lesion with a well-defined, undermined violaceous border.

B-Clinical menifestation:

- start as small pustules, which subsequently burst and expand to form the larger noninfectious ulcer.
- Positive pathergy test.You prick the skin and after 3-6 H the patient will develop pastule and this pastule will converted into ulcer.We usually avoid this test
- Mostly associated with ulcerative colitis. Also with Crohn's disease, rheumatoid arthritis, and leukemia.

C-Management:

- **Surgery is contraindicated.**

4.4- Peutz Jeghers syndrome:

A-Definition:

- It is an autosomal dominant disorder characterized by mucocutaneous hyperpigmentation together with GI polyposis.

B-Clinical features:

- 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.

C-Prognosis:

- 2-3% of patients develop GI carcinoma during their lifetimes.

4.5- Porphyria cutanea tarda:

A-Definition:

- It is an inherited as autosomal dominant disorder resulted from accumulation and increased excretion of porphyrins (a part of the hemogl.obin molecule.).

B-Etiology:

- It results from the decreased activity of the enzyme Uroporphyrinogen decarboxylase (enzyme involved in hemoglobin synthesis pathways).

C-Clinical features:

- Skin photosensitivity with increased skin fragility.
- Blistering of the skin (in areas exposed sunlight)
- Facial hypertrichosis.(is an abnormal amount of hair growth ovet the face)
- Skin hyperpigmentation on the hands and other sun-exposed areas.
- It is often associated with Hepatitiss C .

D-Management:

- Removal of possible triggers, including iron supplementation, alcohol, and estrogens.
- Phlebotomy (a procedure that removes blood from the body).
- Hydroxycholorquine.

4.6- Hemochromatosis:

A-Definition:

- It is a disorder of iron overload leading to excess deposition in multiple body organs.

B-Clinical features:

- Metallic gray or bronze-brown skin colorthat is generally diffuse.
- Skin atrophy.
- Ichthyosis.(thickened, rough, fish scale skin).
- Partial hair loss (most often in the pubic region).
- Koilonychia.(spoon-shaped fingernails).

C-Prognosis:

- Cirrhosis may develop, and might lead to hepatocellular carcinoma.

D-Management:

- treatment involves phlebotomy and chelating agents.

4.7- Liver Cirrhosis:

A-Clinical features:

Some of the associated abnormalities are the following.

- Pruritus. This is related to obstructive jaundice andmay precede it
- Pigmentatio With bile pigments and sometimesmelanin.
- Spider naevi :These are often multiple inchronic liver disease.
- Palmar erythema.
- White nails. These associate with hypoalbuminaemia.
- Porphyria cutanea tarda .
- Xanthomas: In primary biliary cirrhosis .
- Hair loss
- Generalized asteatotic eczema: It mayoccur in alcoholics with cirrhosis who have becomezinc deficient.

5. Cutaneous manifestation of renal diseases:

5.1 Xerosis:

A-Definition:

It is abnormal dryness of the skin, mucous membranes, or conjunctiva . Xerosis occurs in 50-92% of the dialysis population.

B- Etiology:

The exact cause of xerosis in ESRD remains unknown.

C- Management:

Many patients respond to routine use of emollients.

5.2 Pruritus:

A-Definition:

Itch (Latin: pruritus) is a sensation that causes the desire or reflex to scratch.

B- Etiology:

Uremia is the most common metabolic cause of pruritus.

C- Clinical presentation:

- Pruritus affects 15-49% of patients with chronic renal failure and 50-90% of the dialysis population.
- Excoriations
- Prurigo nodularis (It is a skin disease characterised by pruritic (itchy) nodules which usually appear on the arms or legs)
- Lichen simplex chronicus (See Eczema Lecture)

D- Management:

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

E- Prognosis:

Pruritus typically resolves after transplantation.

5.3 Half and half nails:

A-Definition:

Proximal portion of the nail with white color and the discoloration of the distal half (red, pink, or brown), with a sharp demarcation (line f between the two halves).

C- Clinical presentation:

- It occurs in around 40% of patients on dialysis.
- Usually involve fingernails

E- Prognosis:

It typically resolves after transplantation.

5.4 Nephrogenic systemic fibrosis:

A-Definition:

It is a disease of fibrosis of the skin and internal organs reminiscent but distinct from scleroderma or scleromyxedema.

B- Etiology:

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

C- Clinical features:

- Nephrogenic systemic fibrosis mostly seen in ESRD and dialysis patients.
- Presents as thick, indurated plaques on the extremities and the trunk similar to scleroderma.

D- Management:

Treatment includes:

- immunosuppressive agents.
- phototherapy.
- topical steroids.
- Retinoids.
- Photophoresis.

6. Cutaneous manifestation of hyperlipidemia syndromes:

6.1 Xanthelasma palpebrarum:

A-Definition:

It is a yellow plaque that occur near the inner canthus of the eyelid and are often associated with atherosclerosis, dyslipidemia, and coronary artery disease.

B- Clinical presentation:

- Xanthelasma palpebrarum is the most common of the xanthomas
- It is asymptomatic
- bilateral and symmetric
- Can be associated with any type of primary hyperlipoproteinemia .Also, could be without hyperlipidemia.

C- Management:

- topical trichloroacetic acid.
- Electrodesiccation.
- laser therapy.
- surgical excision.

6.2 Tendinous xanthomas:

A-Definition:

It is a Cholestrol deposition and clinically manifested by papules and nodules found in the tendons of the hands, feet, and Achilles.

B- Clinical presentation:

- It is commonly seen on the Achilles tendon followed by the hands, feet, elbows, and knees.
- It is the least responsive xanthoma to treatment.
- Mostly seen in patients with familial hypercholesterolemia.

6.3 Tuberous xanthomas:

A-Definition:

They are firm and non-tender cutaneous and subcutaneous yellowish nodules on extensor surfaces.

B- Clinical presentation:

- It is mostly associated with familial dysbetalipoproteinemia.
- It may resolve after months of treatment with lipid lowering agents.

6.4 Eruptive xanthomas:

A-Definition:

They are painless, yellowish papules on an erythematous base that present as grouped lesions on trunk, elbows and buttocks.

B- Etiology:

They are usually associated with Hypertriglyceridemia.

C- Clinical presentation:

- They could be seen in poorly controlled diabetes and acute pancreatitis.
- They usually resolve in few weeks after therapy**.**

6.5 Planar xanthomas:

A-Definition:

They are elevated cutaneous yellowish-orange deposits on palmar creases.

B- Clinical presentation:

It is usually associated with familial dysbetalipoproteinemia.

7. MCQs

1- A patient who is known case of ulcerative colitis referred to the dermatology clinic due to his complaining of a painful well defined ulcerative lesion. The most likely diagnosis is :

A-Pyoderma gangrenosum

B- Dermatitis herpetiformis

C- Necrobiosis lipoidica

D- Porphyria cutanea tarda.

2- 47 Year old female presents with brown patches in the axilla، Which One of the following is the most recognized clinical feature of acanthosis nigricans:

A - They are small, pedunculated, soft papules on the skin found coomnly on the neck.

B -Thickening of the hands with tiny papules on fingers and stiff joints , but without discoloration

C- It is usually found in obese patints.

D- Diffuse purple skin pigmentation fond on the back, abdomen and shoulder.

3- Patients has abnormal bowel movements, Perianal ulcer,Mouth apthous ulcer(Patient diagnosed with Crohn Disease). What is the most likely of the following skin manifestations can go with the previous clinical picture?

A- Non-tender Ulcers on the calf.

B- Un ulcerated tender nodule on the shin.

C-Small, pedunculated, soft papules on the neck

D-Small severely pruritic vesicular lesions found in a symmetric distribution of both upper and lower extensor surfaces.

Answers:
1-A , 2-C, 3-B