# Cutaneous Manifestations of Gastrointestinal Diseases

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***no disclosures***

# Cutaneous Manifestations of Cirrhosis

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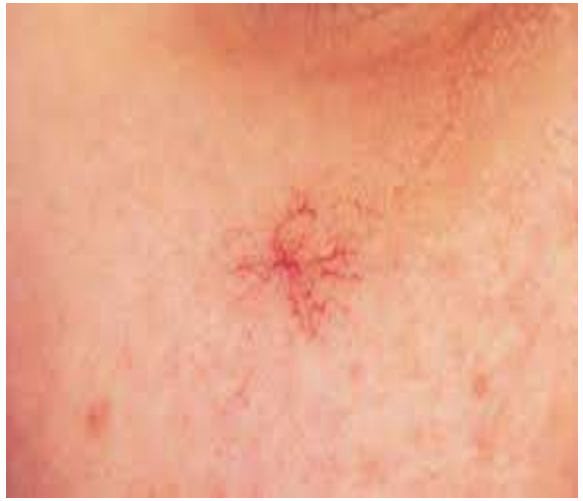
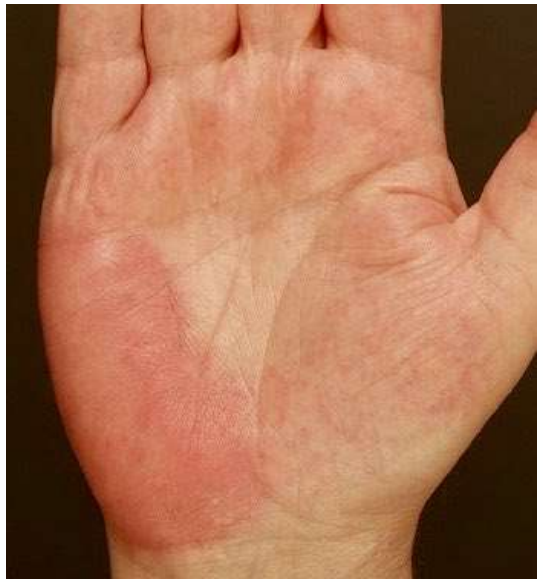
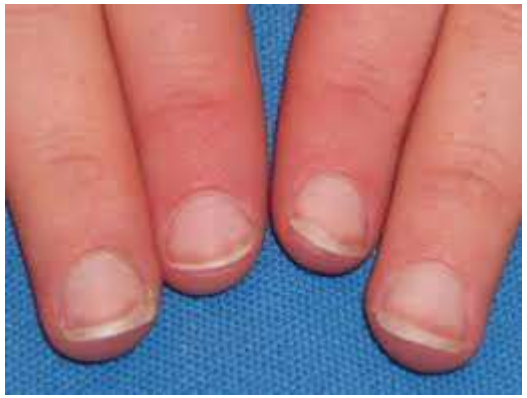
- Spider telangiectasia



- Palmar Erythema



- Terry nails
  - white proximal (leukonychia) and pink distal nail plate



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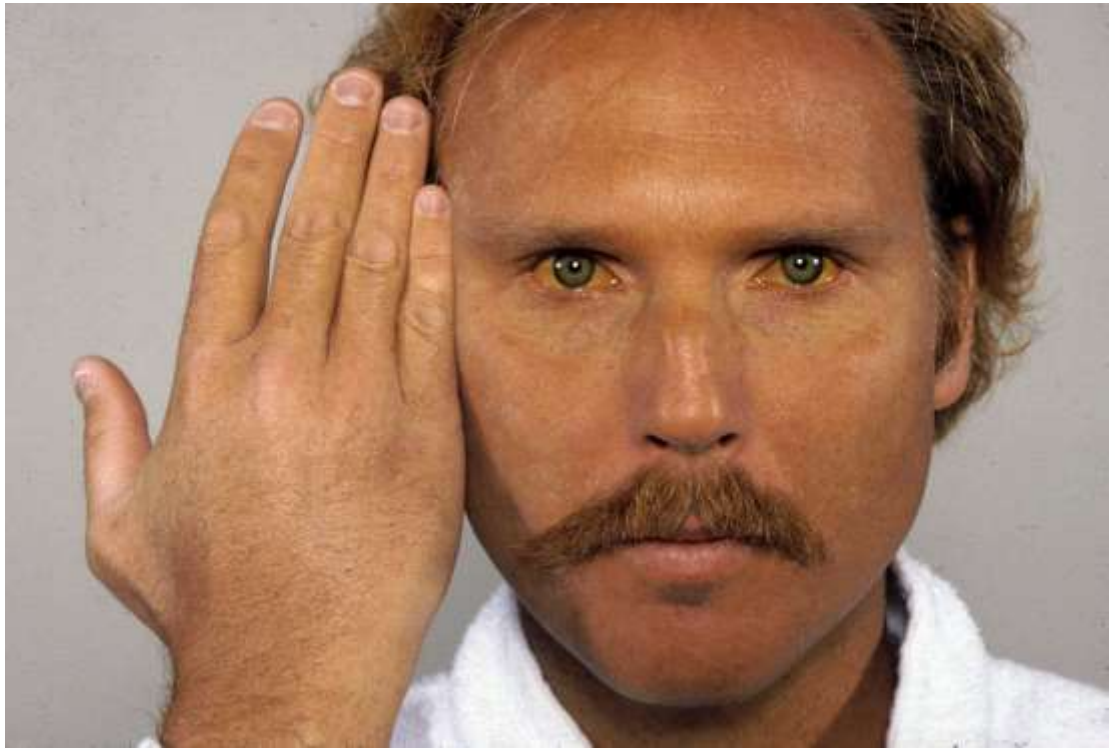
# Cutaneous Manifestations of Cirrhosis

- Hemochromatosis
  - An autosomal dominant iron-accumulation disorder that may cause generalized bronze hyperpigmentation



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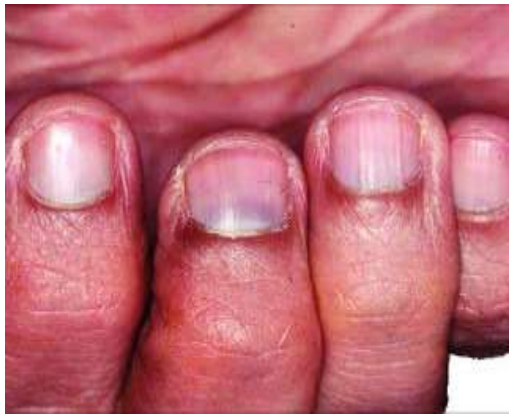
# Cutaneous Manifestations of Cirrhosis

- Wilson's Disease:
  - An autosomal recessive disorder of copper metabolism
    - Kayser-Fleischer ring around iris
    - Pigmentation over the shins
    - Blue lunulae (half-moons) on nails



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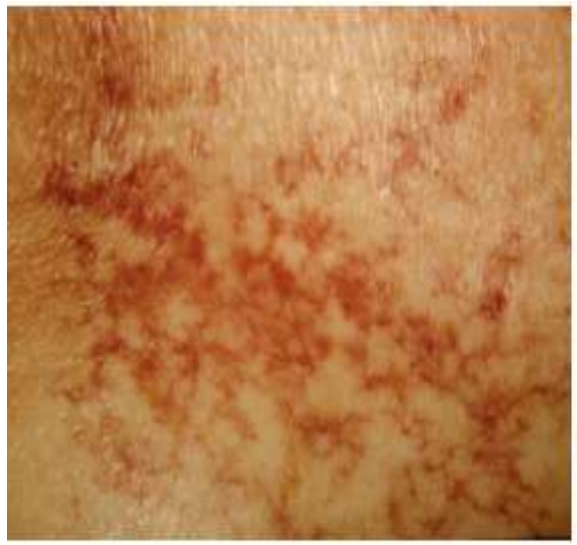
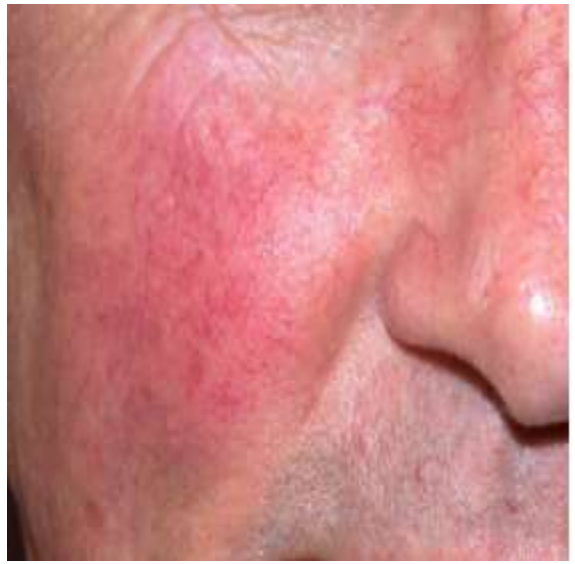
# Cutaneous Manifestations of Cirrhosis: Telangiectasia, Terry Nail, Facial Redness





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# Cutaneous Manifestations of Malnutrition

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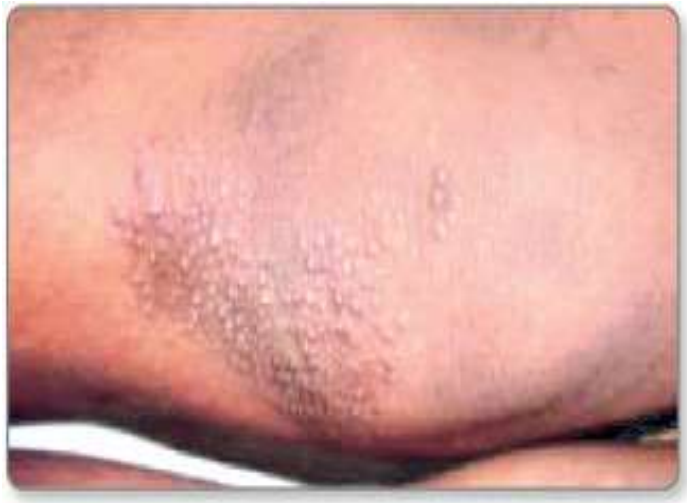
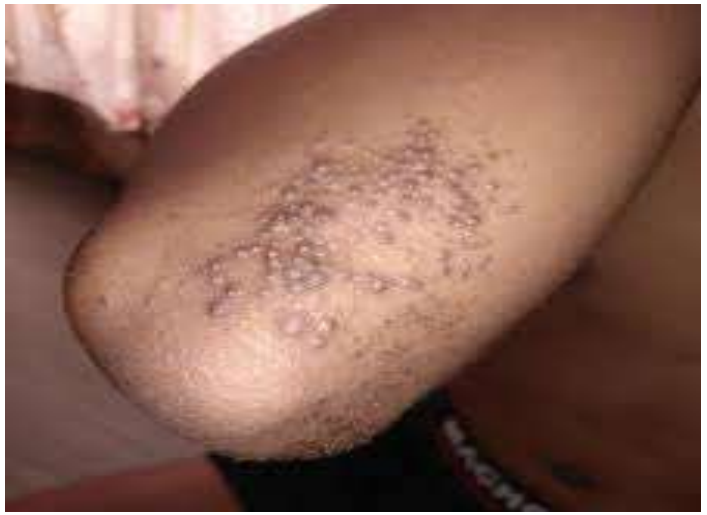
- Vitamin A (Retinol) Deficiency:
  - Phrynoderma → keratotic follicular papules on anterolateral thighs and posterolateral upper arms.





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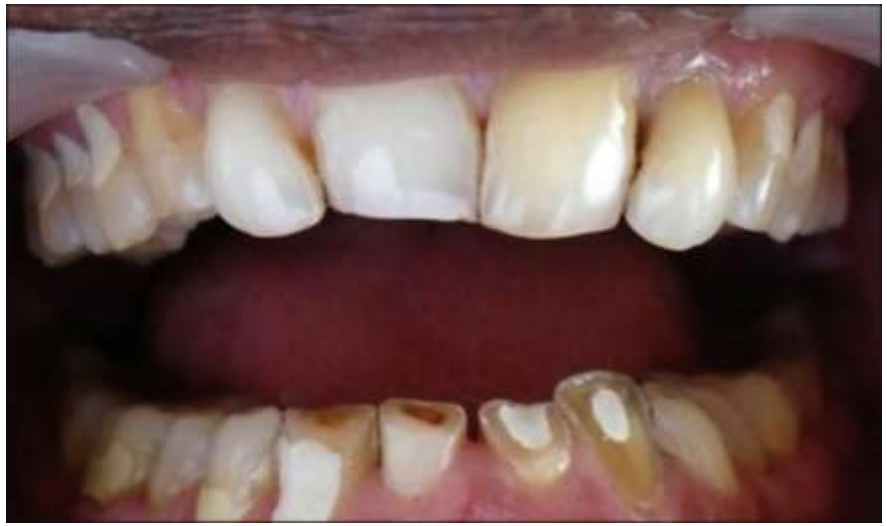
- Vitamin D (Cholecalciferol D3) deficiency:
  - Children → delayed tooth eruption with poor enamel
  - Adults → predisposes to caries



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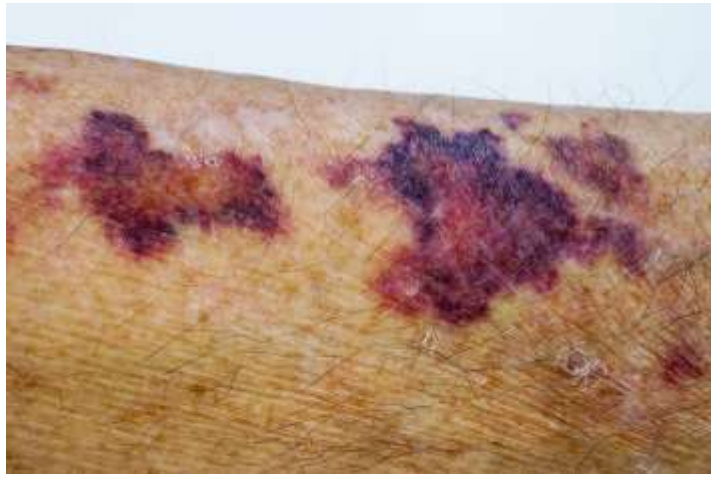
# Cutaneous Manifestations of Malnutrition

Vitamin K (Phytonadione) deficiency:

- Interferes with coagulation → hemorrhage, purpura and ecchymoses



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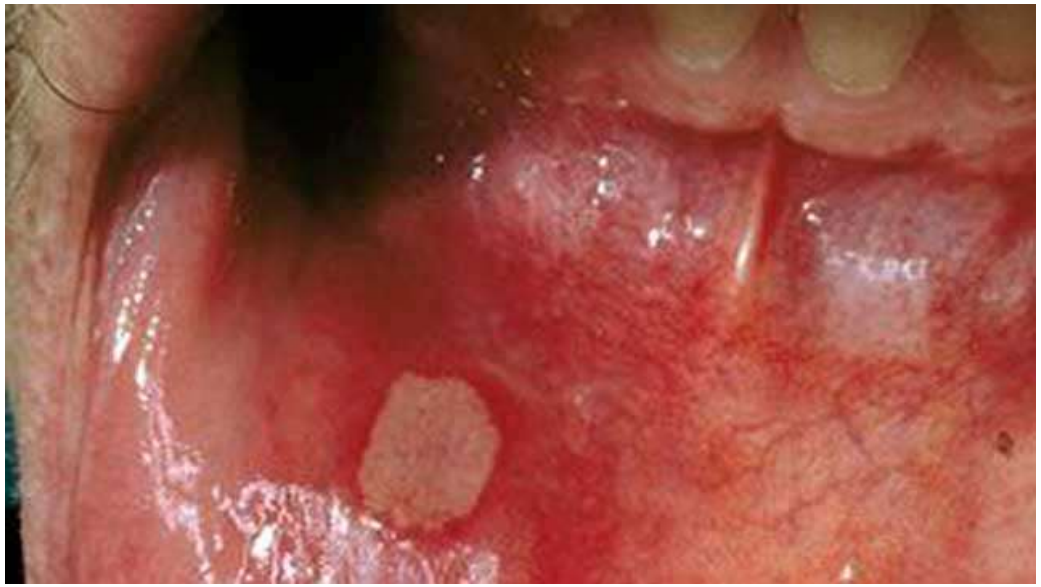
# Cutaneous Manifestations of Malnutrition

Acute vitamin B2 (Riboflavin) deficiency results in:

- Deep red erythema spots
- Epidermal necrolysis (peeling skin)
- Stomatitis



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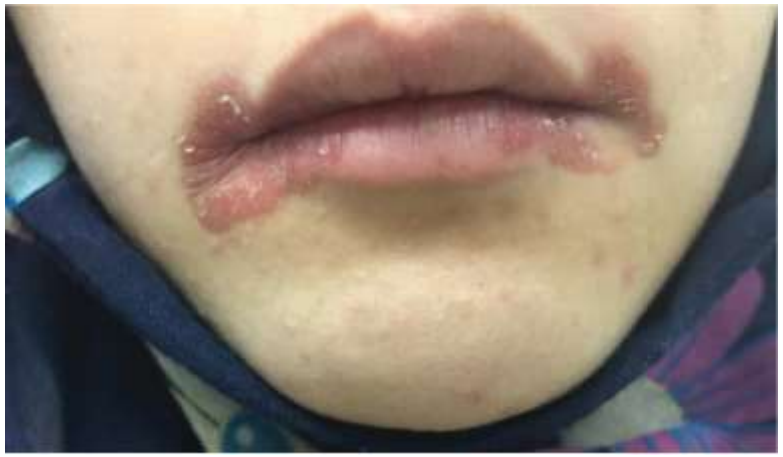
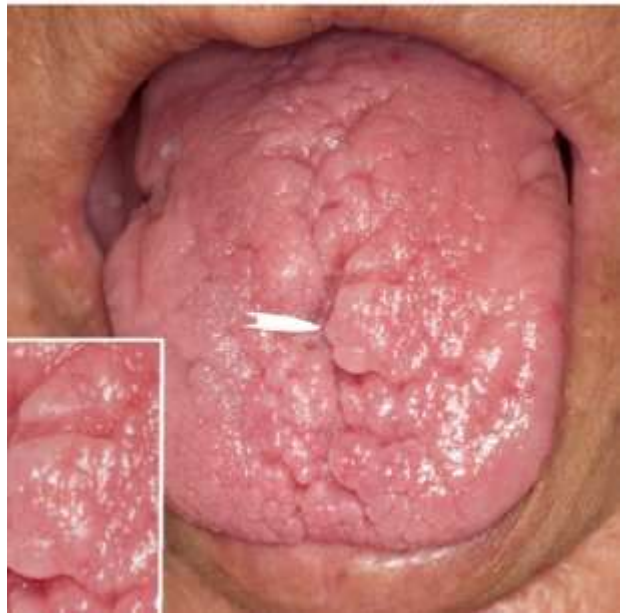
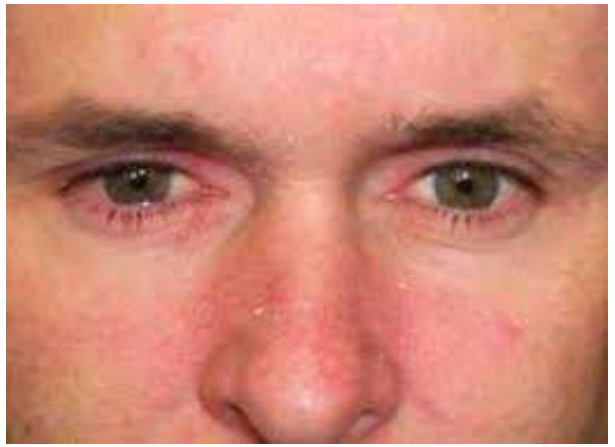
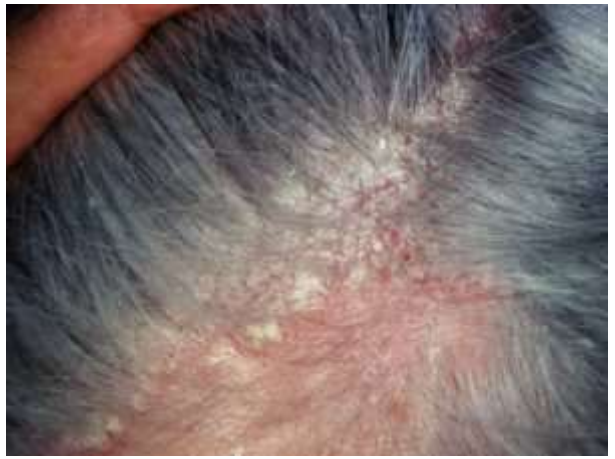
# Cutaneous Manifestations of Malnutrition

- Chronic vitamin B2 (Riboflavin) deficiency causes:
  - Prominent lingual papillae
  - Seborrheic dermatitis
  - Angular cheilitis









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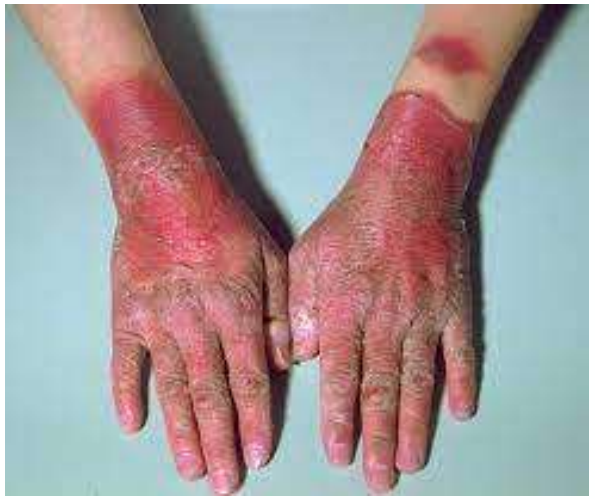
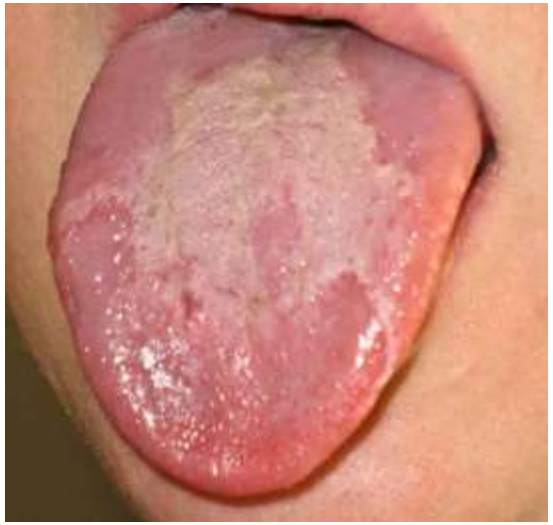
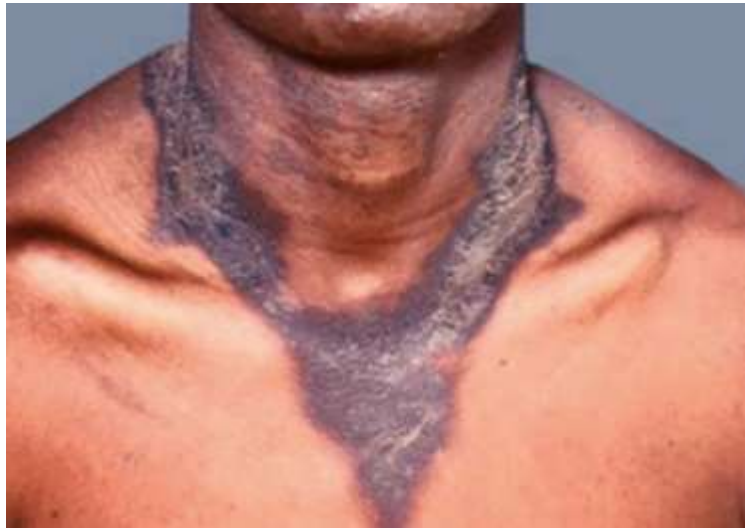
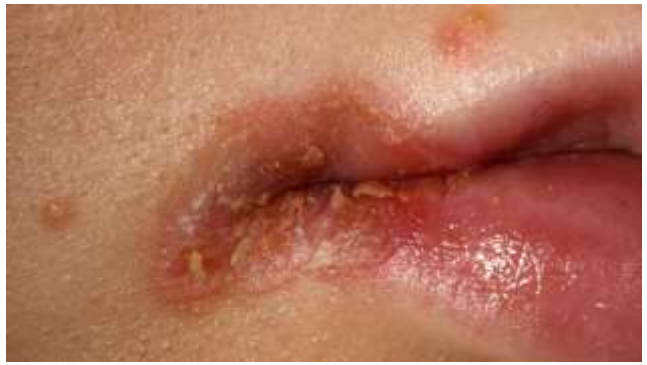
# Cutaneous Manifestations of Malnutrition

- Vitamin B3 (Niacin) deficiency:
  - Pellagra (diarrhea, dermatitis, dementia and death):
    - Photosensitivity
    - Casal necklace
    - Glossitis
    - Angular cheilitis
    - Oral/perioral ulcers









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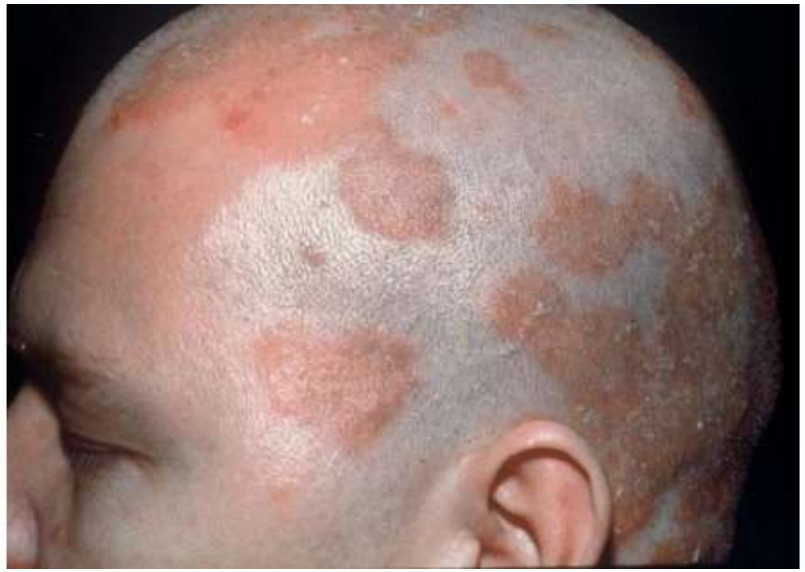
# Cutaneous Manifestations of Malnutrition

- Vitamin B6 (Pyridoxine)deficiency:
  - Seborrheic dermatitis
  - Glossitis
  - Angular cheilitis



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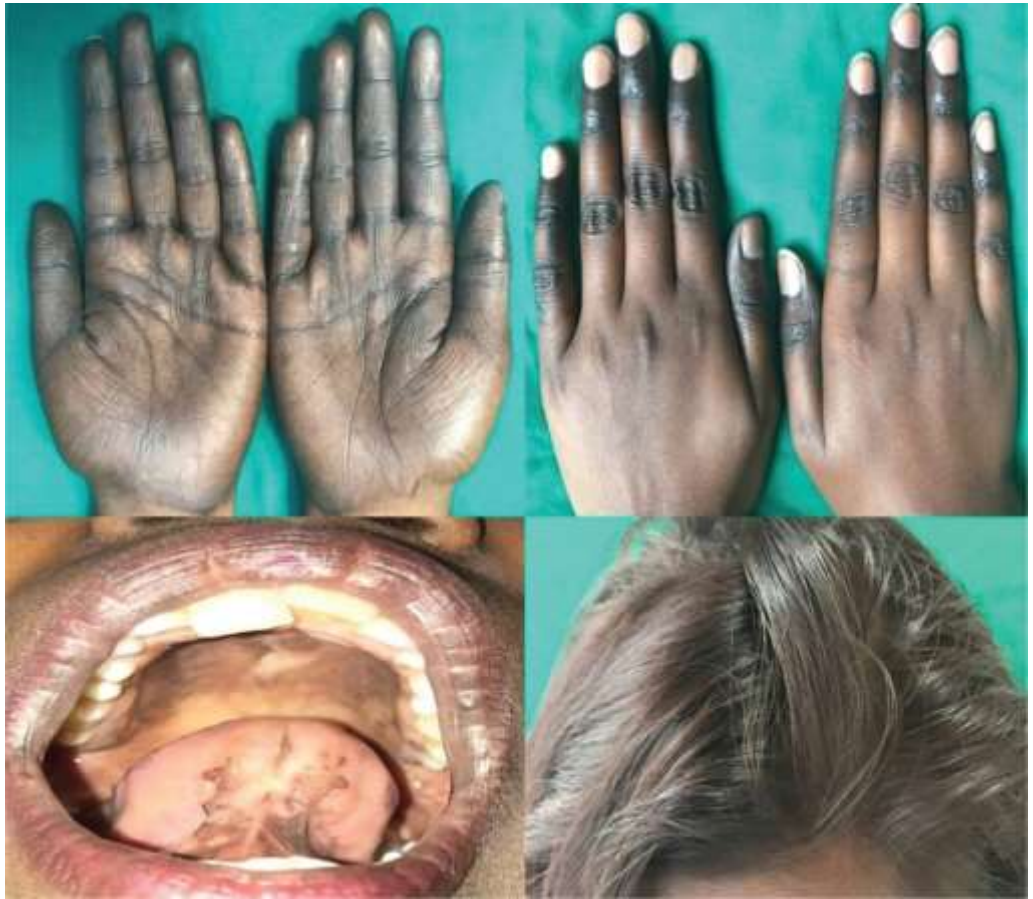
# Cutaneous Manifestations of Malnutrition

- Vitamin B9 (Folate)/B12 (Cobalamine) deficiency:
  - Angular cheilitis
  - Hunter glossitis
  - Hair depigmentation
  - Hyperpigmentation



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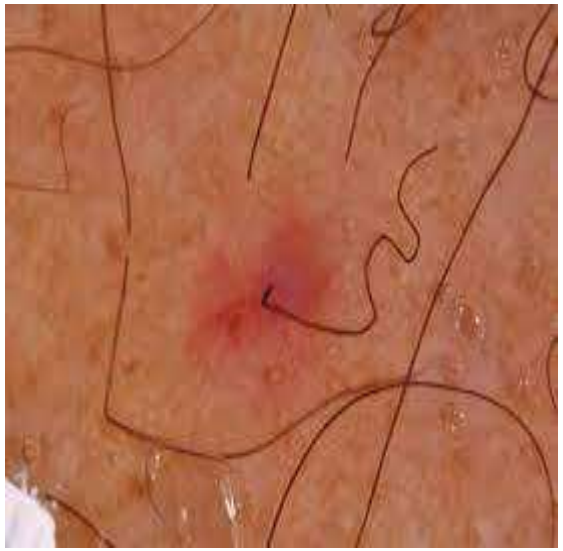
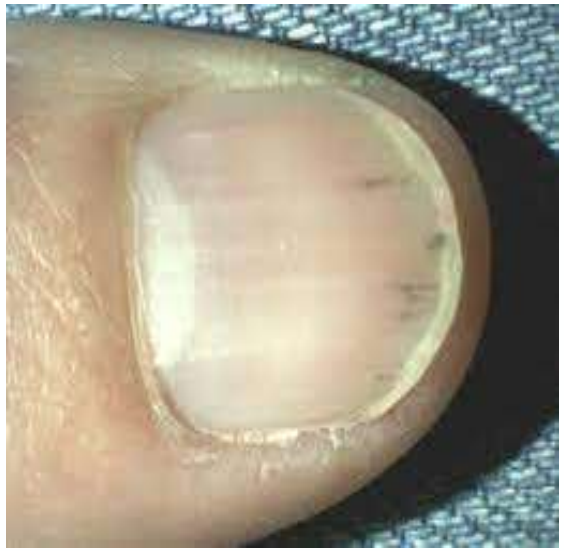
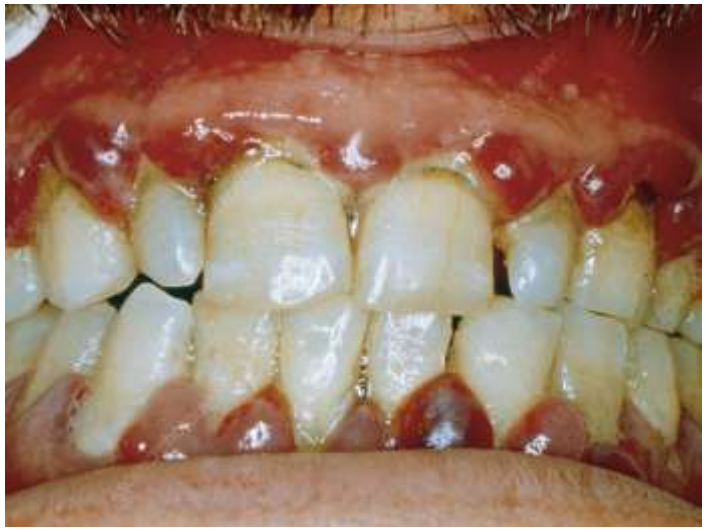
# Cutaneous Manifestations of Malnutrition

- Vitamin C (Ascorbic acid) deficiency → Scurvy
  - Enlarged, hyperkeratotic hair follicles on posterolateral aspects of arms
  - Abnormal hair development
  - Splinter hemorrhage of nails
  - Swelling of gums, erythema







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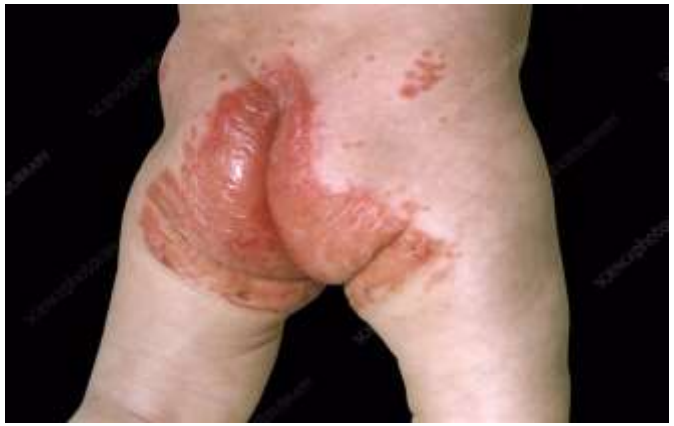
# Cutaneous Manifestations of Malnutrition

**Acrodermatitis enteropathica** → autosomal recessive inherited form of zinc deficiency with a clinical triad of dermatitis, alopecia and diarrhea

- Scaly patches and plaques on extremities, periorificial and anogenital sites
- Psoriasiform plaques → vesicular, bullous, pustular or erosive lesions
- Possible hair loss



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# Cutaneous Manifestations of Inflammatory Bowel Diseases

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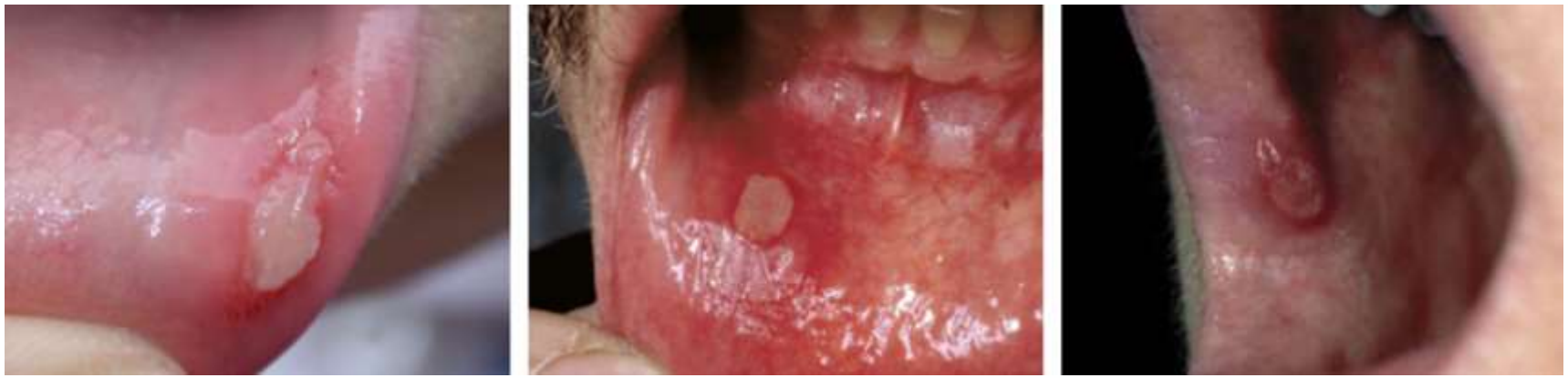
IBD – Crohns and Ulcerative Colitis

Oral aphthous ulcers:

- Common in both forms of IBD
- Round mucosal ulcers



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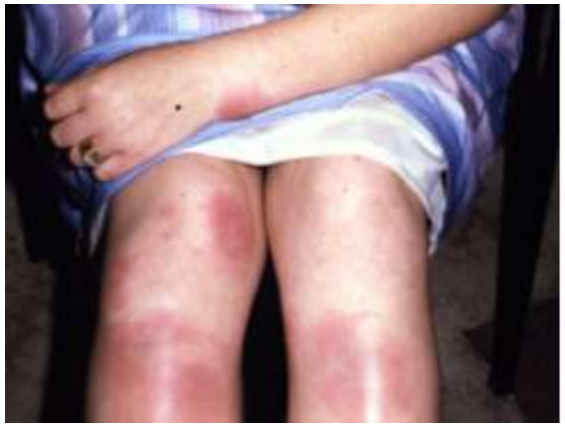
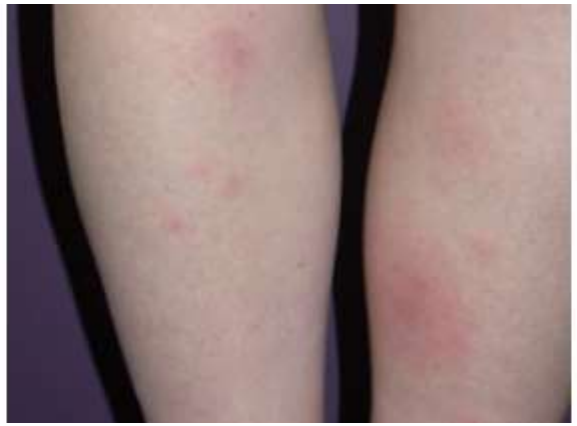
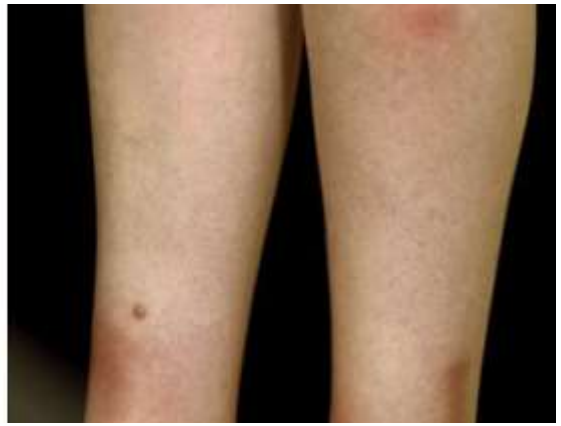
# Cutaneous Manifestations of Inflammatory Bowel Diseases

Erythema nodosum → (EN) is the most common skin sign of Crohn disease (4-6%) or ulcerative colitis (3%)

- A form of panniculitis
- Tender red subcutaneous nodules
  - Symmetrical extensor surfaces of lower legs, extensor surfaces of arms, trunk and face

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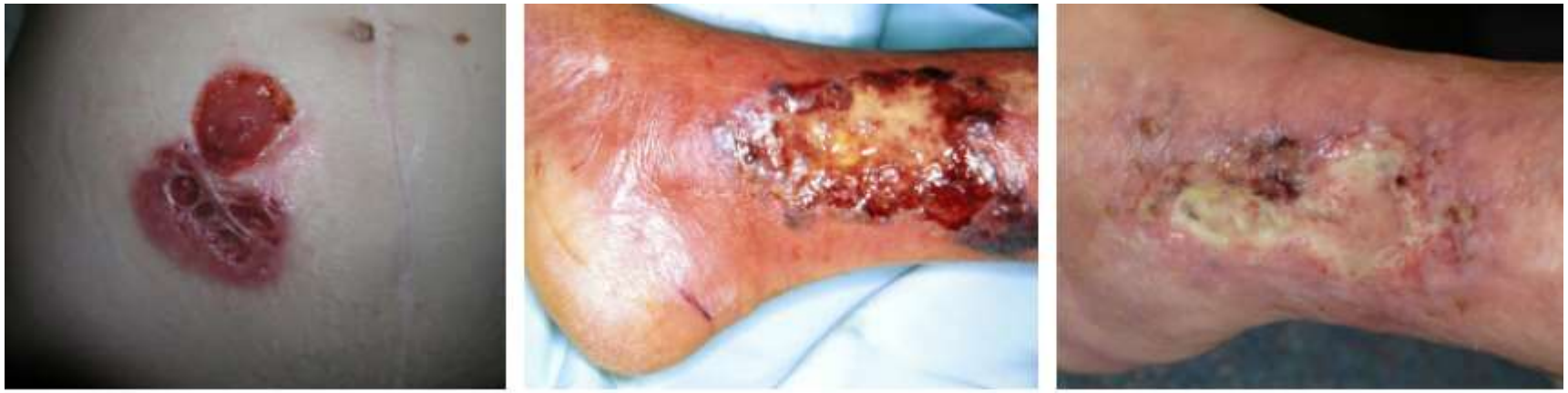
# Cutaneous Manifestations of Inflammatory Bowel Diseases

Pyoderma gangrenosum (PG) → affects 0.7% of patients with Crohns disease and 2% with ulcerative colitis

- Ulcers with irregular undermined necrotic borders
- Usually affects lower extremities but can arise anywhere
- Peristomal PG  develops two months up to 25 years after stoma creation



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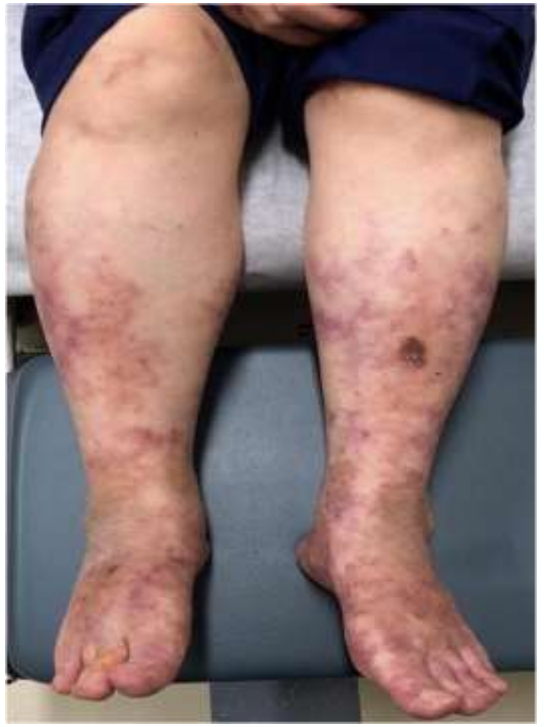
# Cutaneous Manifestations of Inflammatory Bowel Diseases

Cutaneous polyarteritis nodosa → rare form of necrotizing vasculitis

10% of cases are associated with IBD

- Causes tender arterial nodules, palpable purpura and deep ulceration
- Lower extremities



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# Cutaneous Manifestations of Gastrointestinal Diseases

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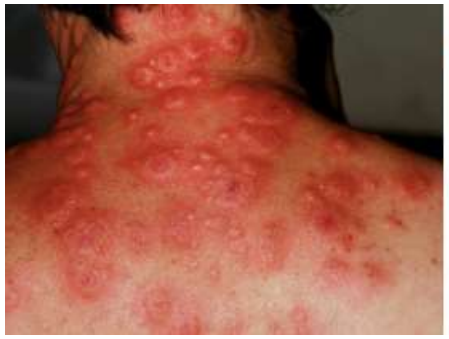
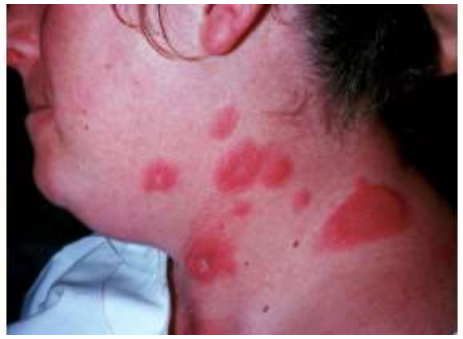
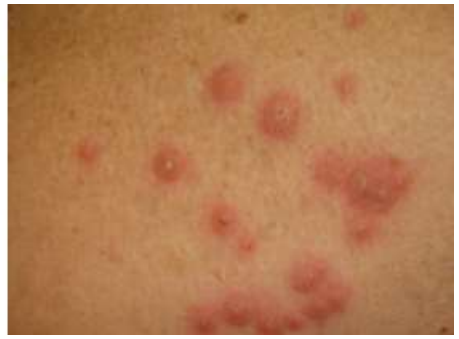
# Cutaneous Manifestations of Gastrointestinal Diseases

Acute neutrophilic dermatosis (Sweet Syndrome) → autoinflammatory disorder sometimes associated with IBD:

- Tender erythematous papules and plaques
- Vesicles, bullae or pustules within the plaques
- Typically affects the head, neck and extremities but can be widespread
- Usually resolves spontaneously within 5–12 weeks

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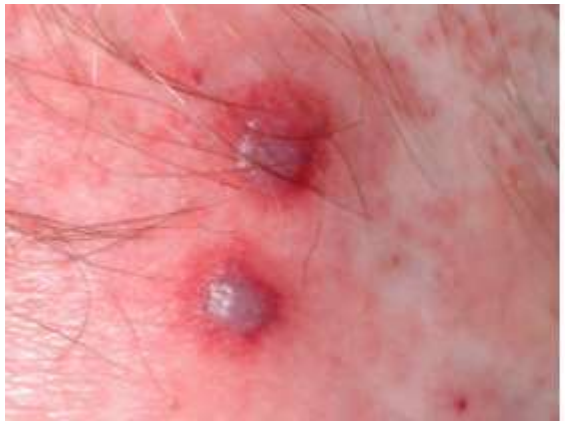
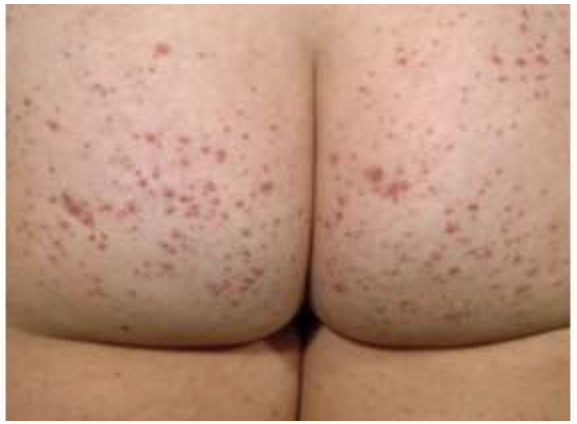
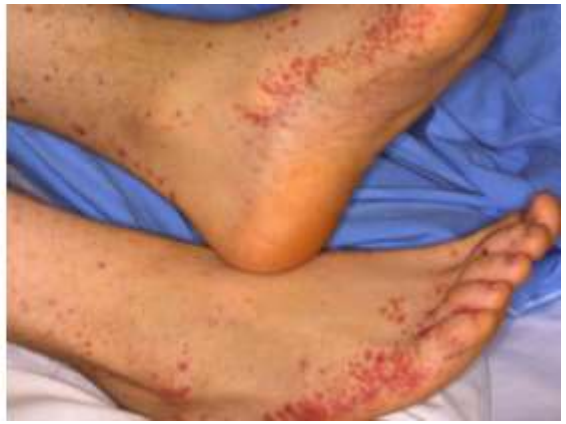
# Cutaneous Manifestations of Gastrointestinal Diseases

Henoch-Schönlein purpura → immunoglobulin A-mediated, leukocytoclastic small vessel vasculitis

- Palpable purpuric lesions usually affect buttocks and legs
- Gastrointestinal vasculitis causing mucosal edema, hemorrhage and rarely, bowel infarction, perforation and/or intussusception

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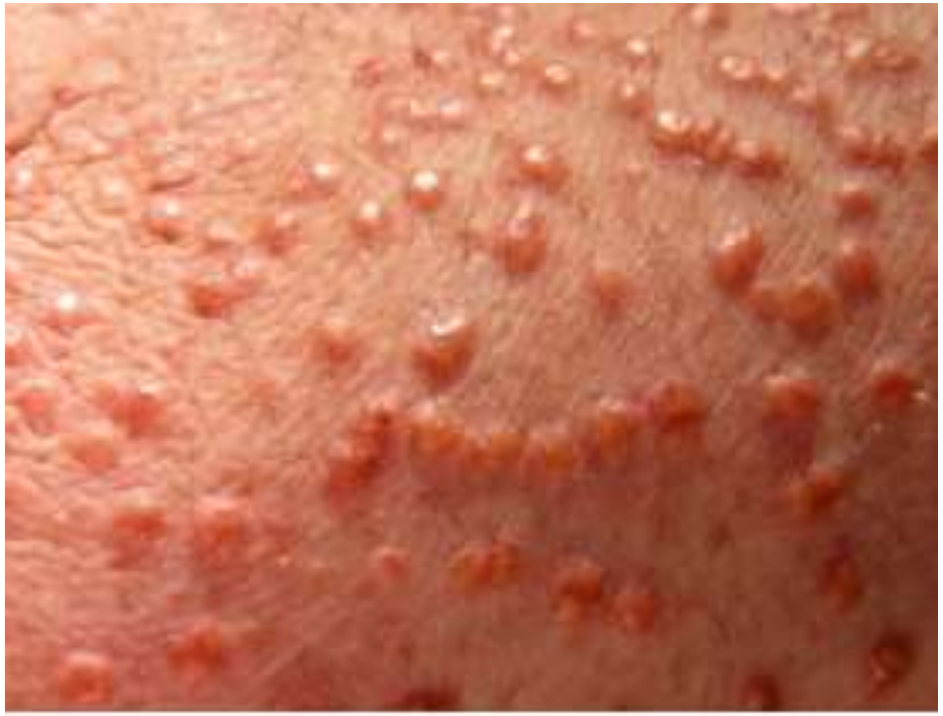
# Cutaneous Manifestations of Gastrointestinal Diseases

## Eruptive xanthomas

- Caused by the accumulation of fat in macrophage cells in the skin
- Associated with hypertriglyceridemia and diabetes



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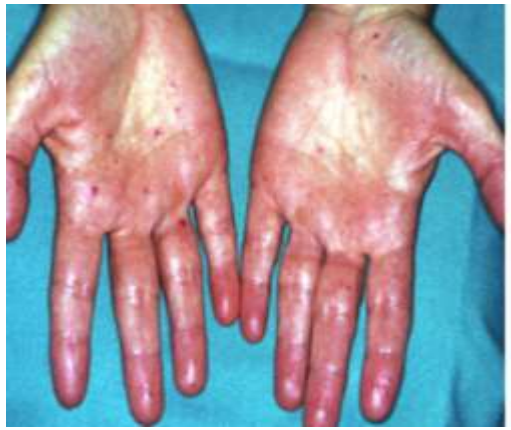
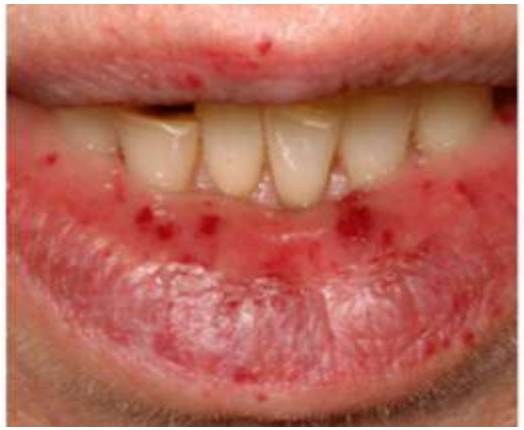
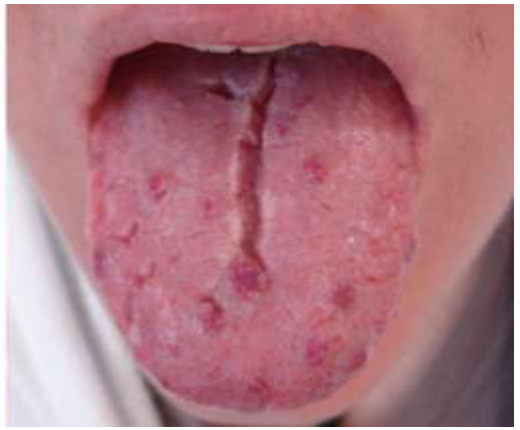
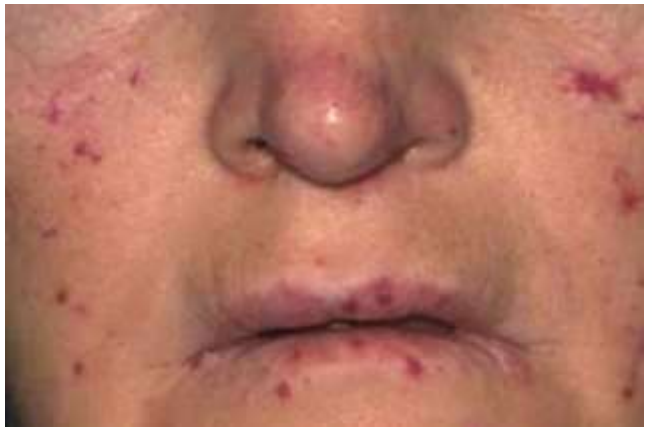
# Cutaneous Manifestations of Gastrointestinal Diseases

Hereditary Hemorrhagic Telangiectasia (HHT) → hereditary, autosomal dominant, bleeding disorder:

Multiple, small punctate telangiectasis affects 50% of patients by 30 years old

- Telangiectasias are predominantly found on face and mouth
- Recurrent nosebleeds affect 90% of patients with HHT
- Bleeding may also occur from gastrointestinal arteriovenous malformations

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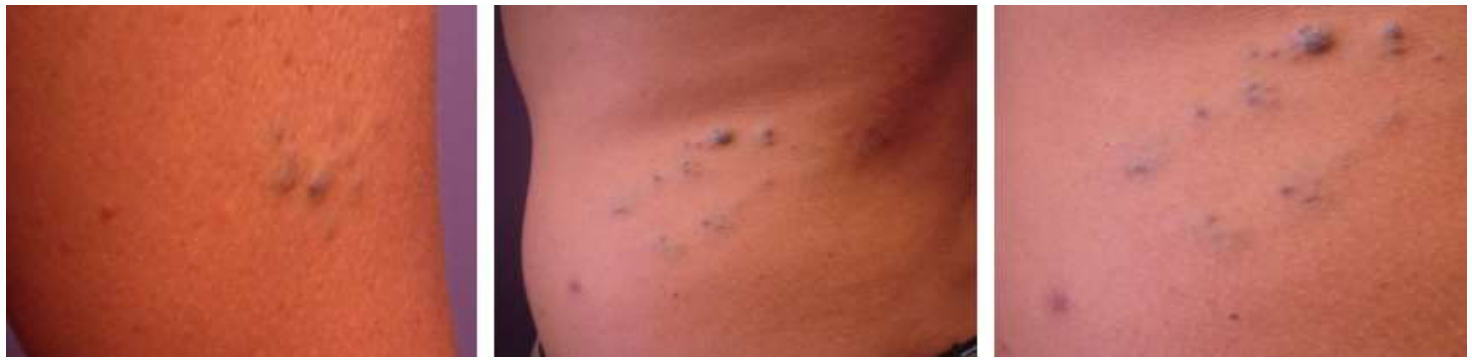
Blue rubber bleb naevus syndrome → vascular malformations of skin and gastrointestinal tract anywhere from the mouth to the anus, most commonly in the small bowel

- The “blue rubber nipple” is a blue-purple, soft, rubbery subcutaneous nodule that fills after compression
- Associated hemangiomas are tender macular lesions found on upper limbs, trunk, perineum



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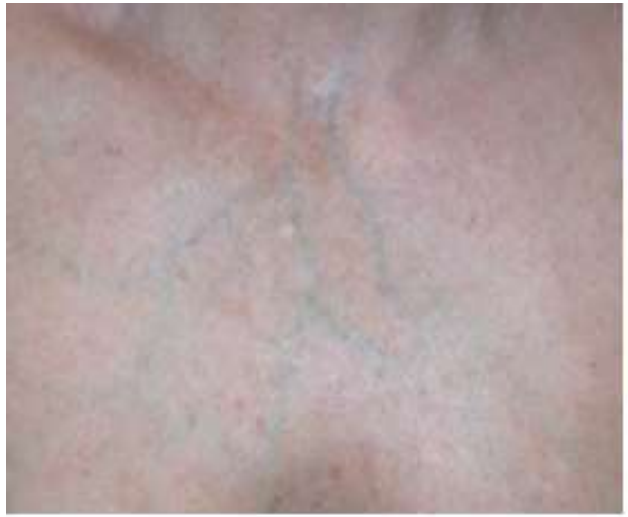
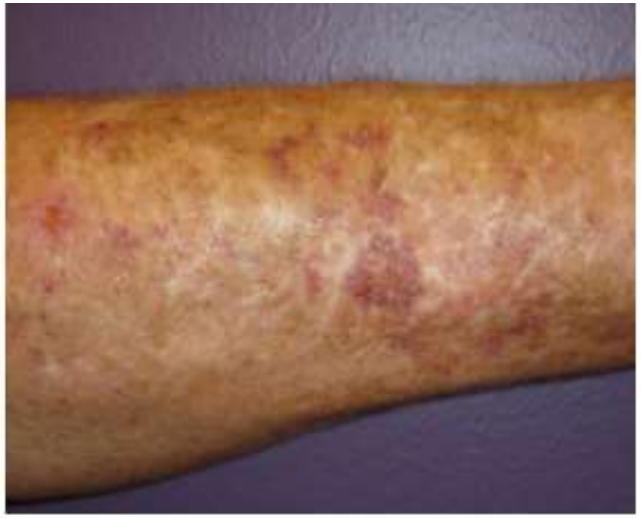
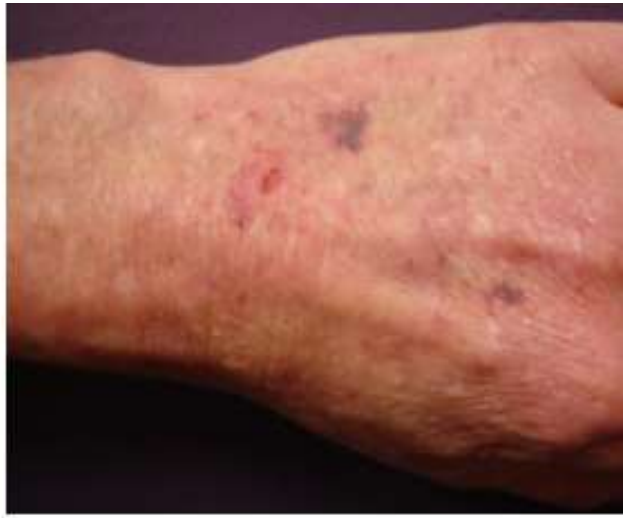
# Cutaneous Manifestations of Gastrointestinal Diseases

Ehlers-Danlos syndrome (EDS) type IV is an inheritable connective tissue disease due to a defect in collagen synthesis:

- Translucent skin
- Varicosities
- Delayed wound healing and keloids







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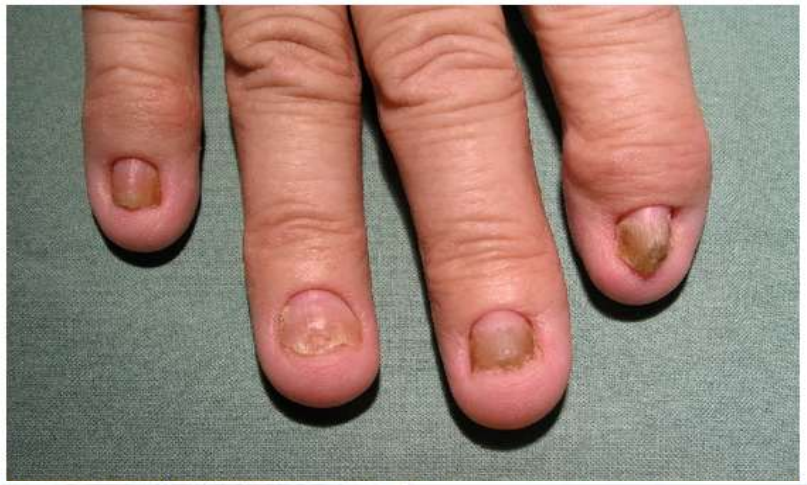
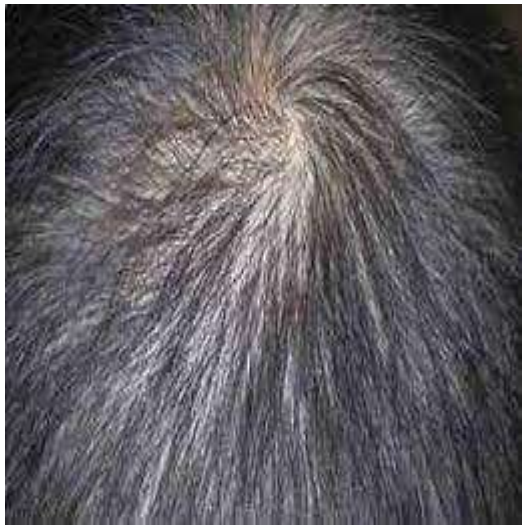
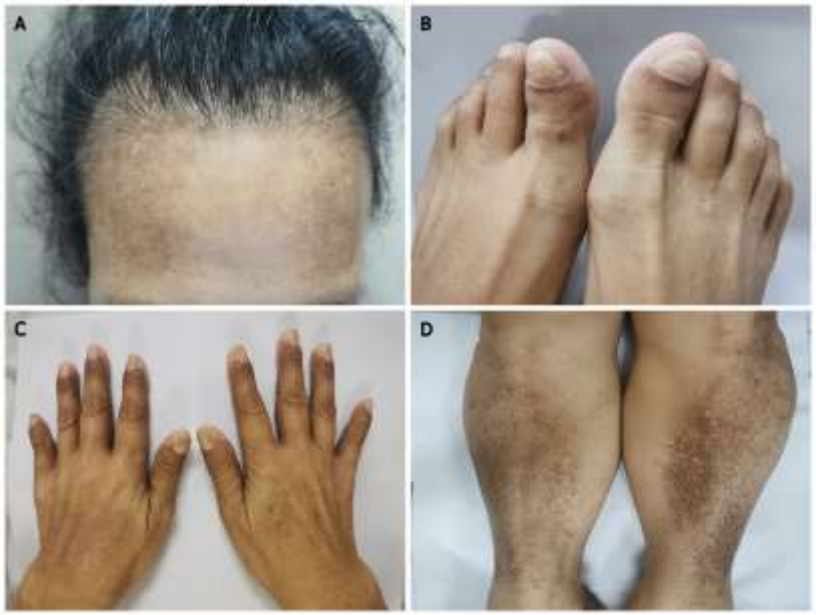
# Cutaneous Manifestations of Gastrointestinal Diseases

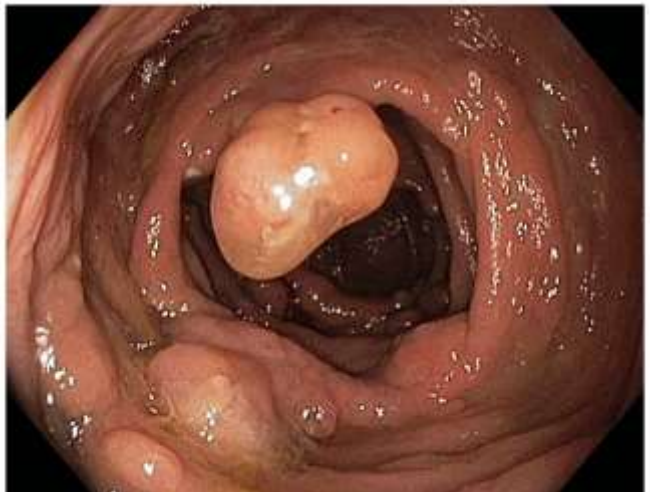
Cronkhite-Canada Syndrome → sporadic syndrome that affects older adults

- Nail dystrophy
- Hair loss
- Diffuse hyperpigmentation
- Intestinal polyps







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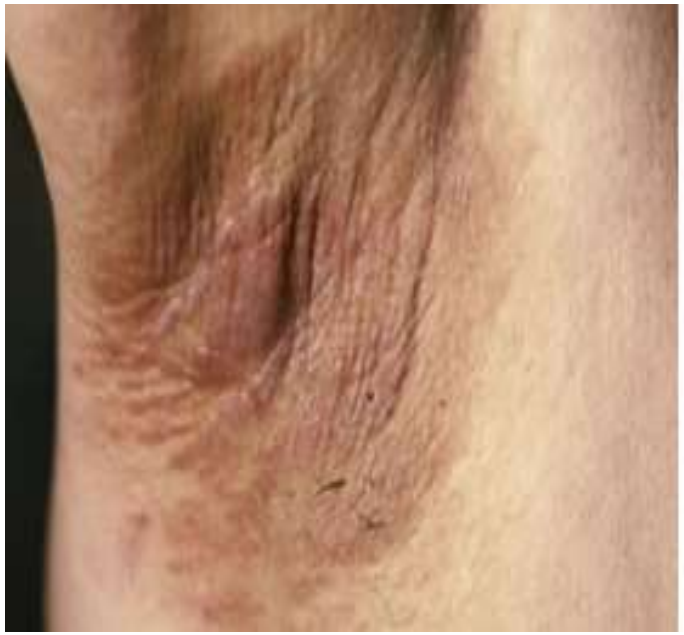
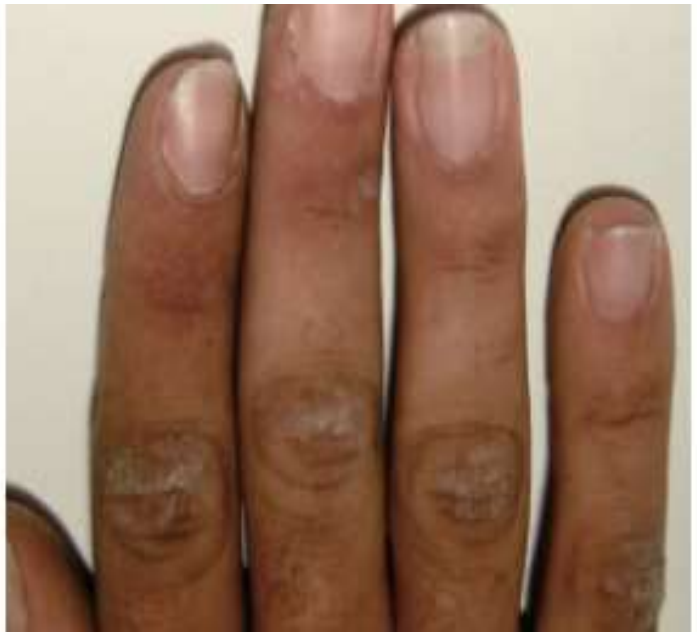
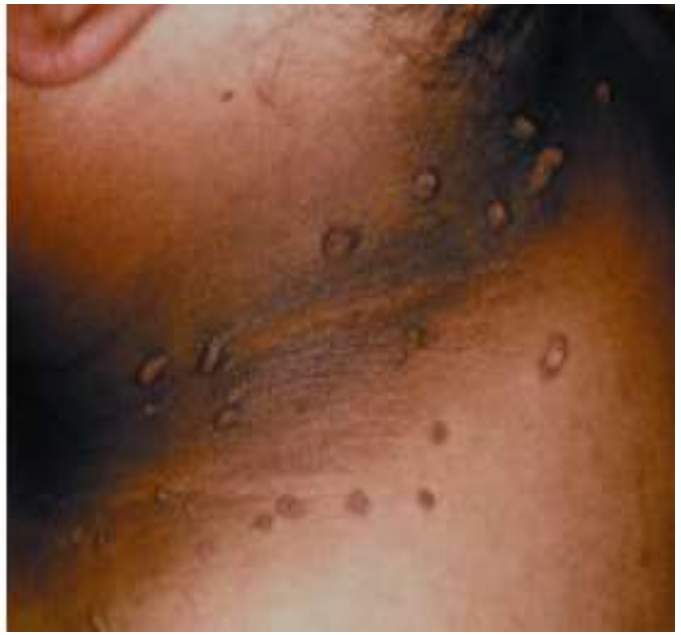
# Cutaneous Manifestations of Gastrointestinal Diseases

Acanthosis nigricans → associated with insulin resistance

- Hyperpigmented, velvety, hyperkeratotic plaques
- Axillary, inguinal, neck folds, mammary, umbilical and anogenital regions
- Acanthosis nigricans associated with malignancy:
  - Affect areola, digital web spaces, extensor sites and mucous membranes
  - Arises spontaneously
  - Rapidly progressive
- Gastrointestinal adenocarcinoma can be responsible for acanthosis nigricans

# Acanthosis Nigricans

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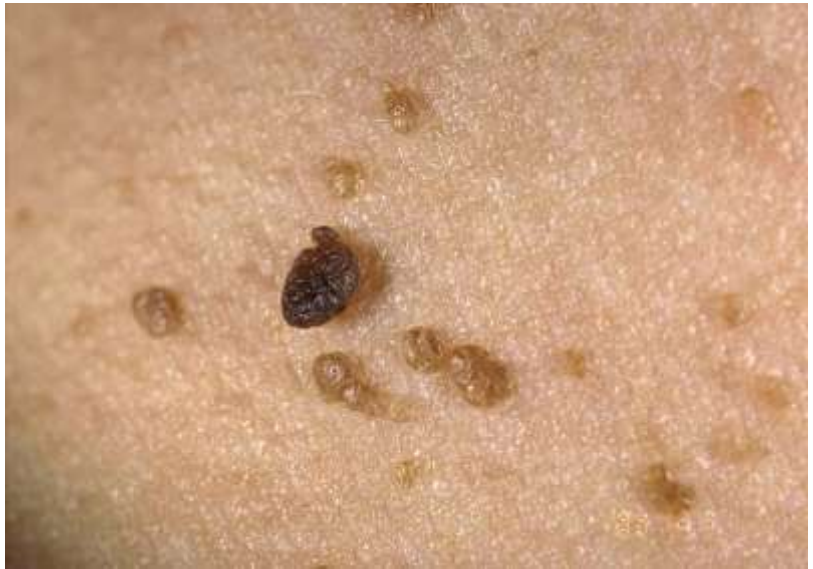
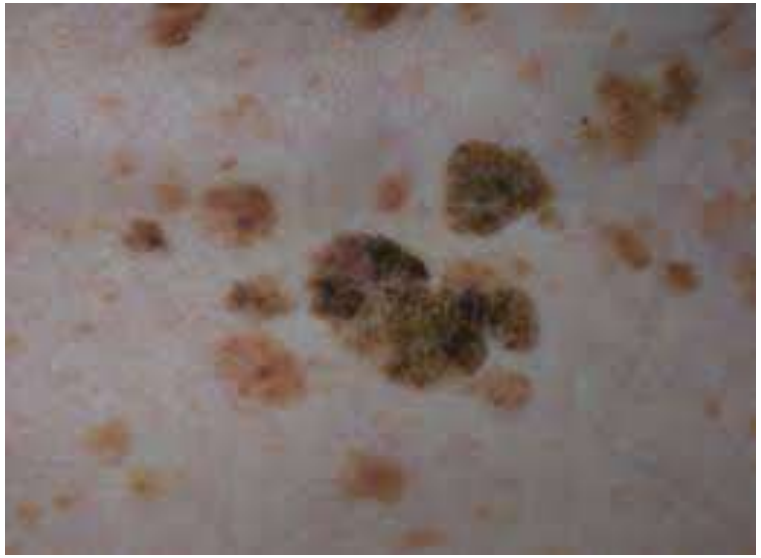
# Cutaneous Manifestations of Gastrointestinal Diseases

Multiple seborrheic keratoses → acute onset of multiple, eruptive keratosis is known as the sign of Leser- Trelat

- Seborrheic keratoses initially erupt on the trunk, then on the extremities or face
  - When associated with gastrointestinal adenocarcinomas, the sign of Léser-Trelat carries a poor prognosis





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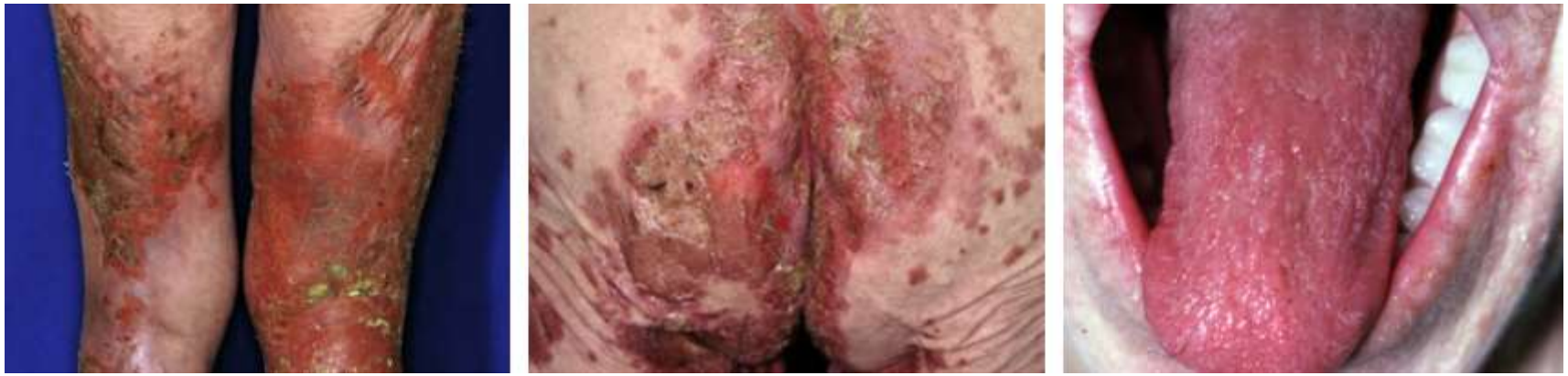
# Cutaneous Manifestations of Gastrointestinal Diseases

Glucagonoma → a rare, glucagon-secreting, pancreatic, alpha cell tumor

- Glucagonoma is associated with necrolytic migratory erythema
  - Painful, pruritic, annular, erythematous eruption with central blisters, erosions, crusting and post-inflammatory pigmentation



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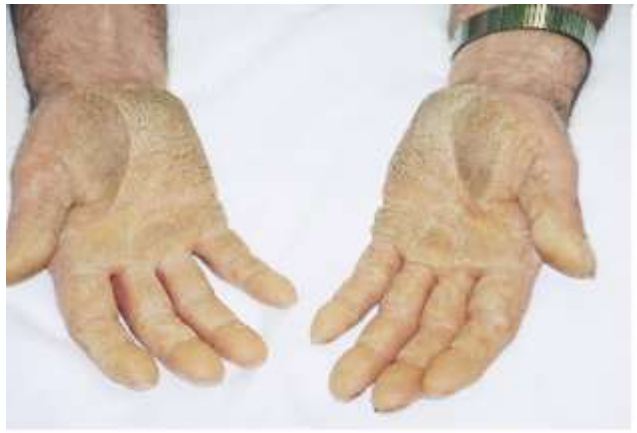
# Cutaneous Manifestations of Gastrointestinal Diseases

Tylosis → autosomal dominant form of focal, non-frictional and nonepidermolytic, palmoplantar keratoderma

- Involves more than 50% of the surface of palms and soles
  - Type A tylosis usually arises between 5 and 15 years of age; it is associated with esophageal carcinoma at a later age
  - Type B arises in the first year of life and is generally benign



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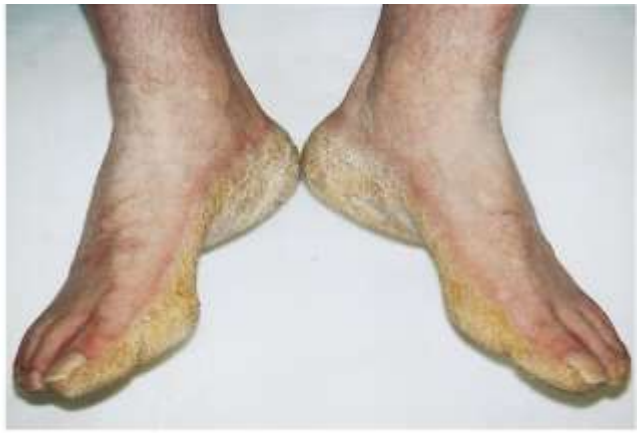

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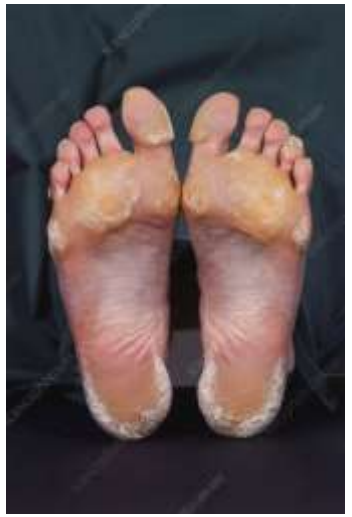

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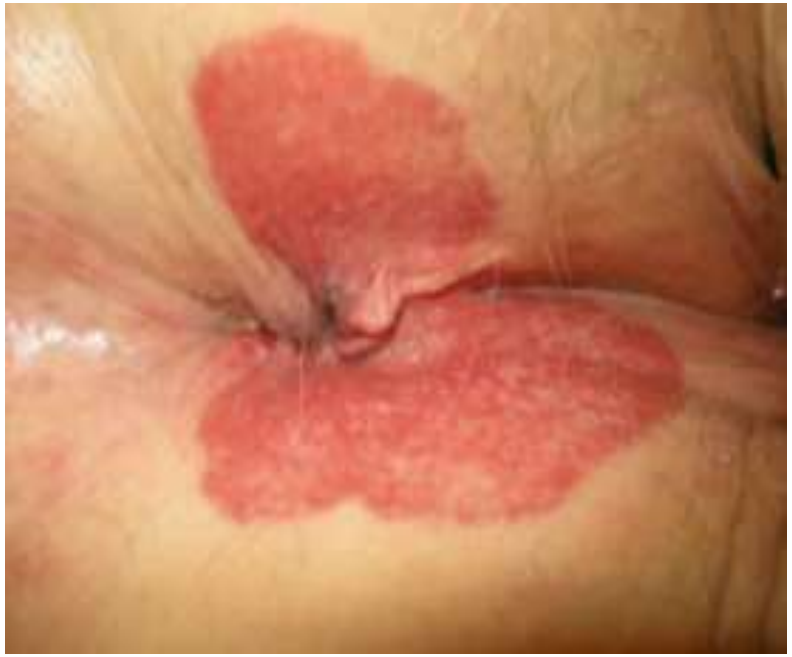
# Cutaneous Manifestations of Gastrointestinal Diseases

Extramammary Paget disease → intraepithelial adenocarcinoma located in and around the anal verge, vulva, or male genitalia

- Anal Paget disease is associated with anorectal adenocarcinoma
- Slowly-enlarging, unilateral, dry, scaly plaque



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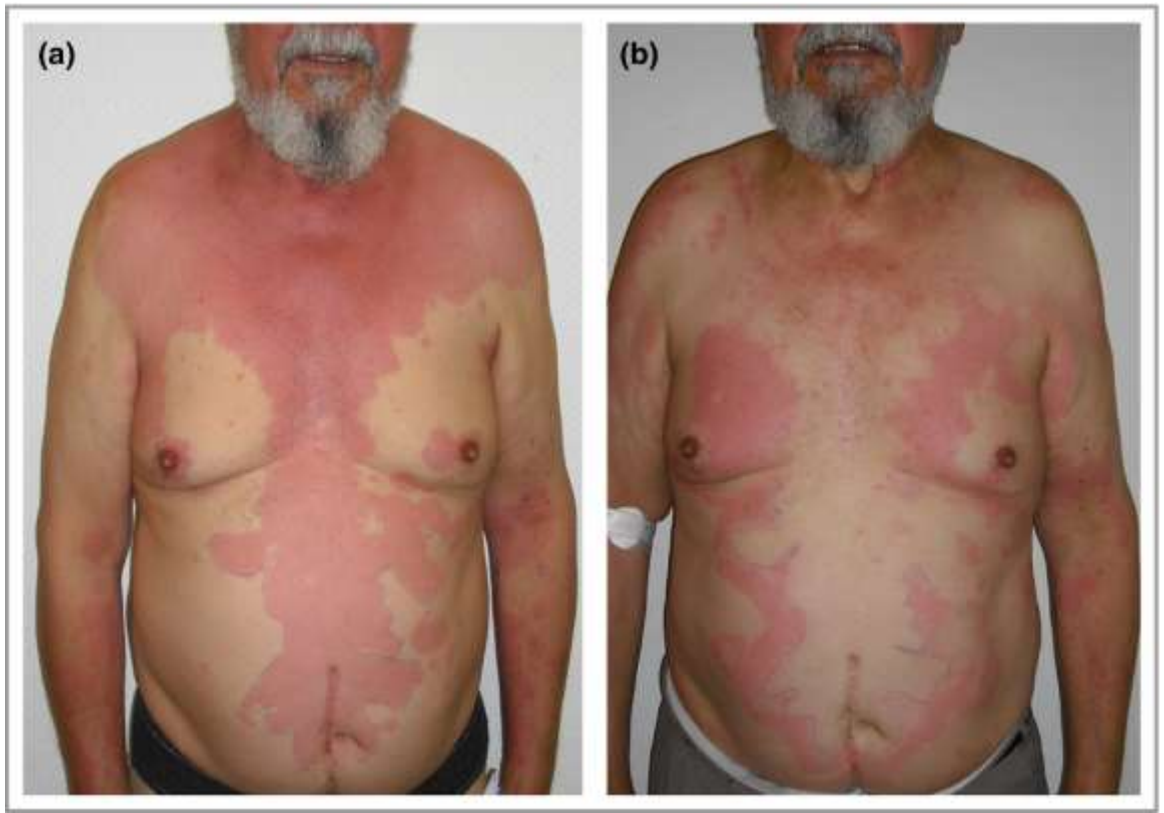
# Cutaneous Manifestations of Gastrointestinal Diseases

Carcinoid syndrome → is the association of intestinal carcinoid with hepatic metastases

- Carcinoid syndrome causes flushing and diarrhea

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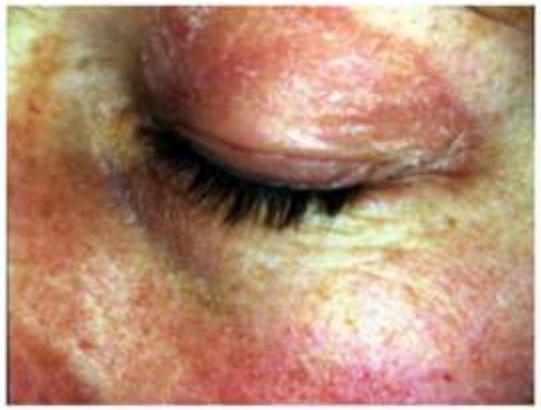
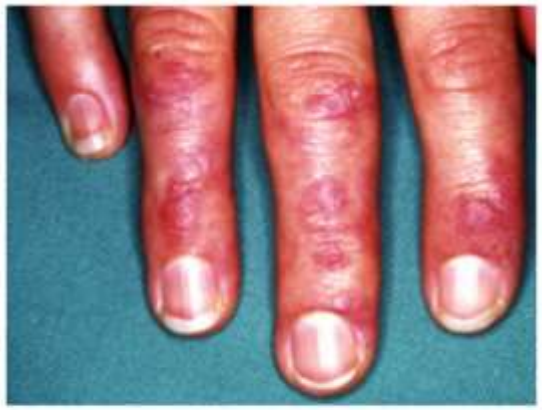
# Cutaneous Manifestations of Gastrointestinal Diseases

Dermatomyositis → autoimmune disease, 15–50% have an underlying malignancy of the gastrointestinal tract, pancreas, lung, breast, ovaries, or non-Hodgkin lymphoma

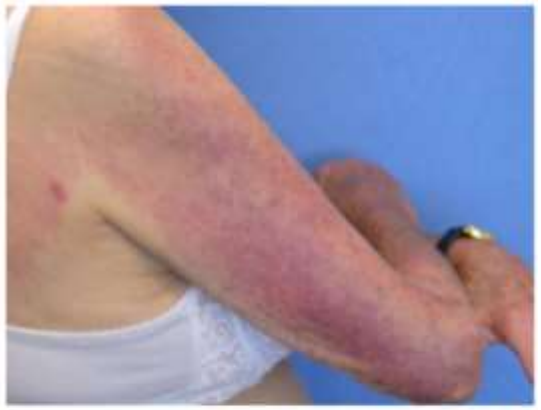
- Dermatomyositis associated with malignancy may precede the diagnosis of cancer by about six months
- Dermatological manifestations of paraneoplastic dermatomyositis include:
  - Heliotrope rash with periorbital edema
  - Gottron papules on finger joints
  - Violaceous poikiloderma over the chest, upper back, elbows and knees
  - Nail signs: “ragged cuticle” and nail fold telangiectasias



Heliotrope eyelids



Gottron's papules



Dermatomyositis

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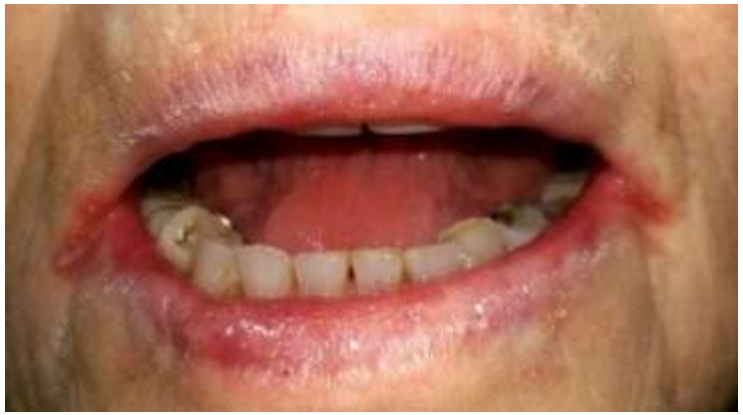
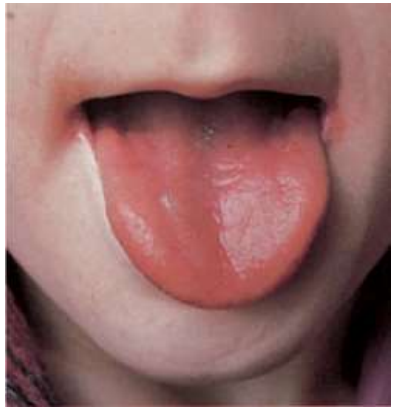
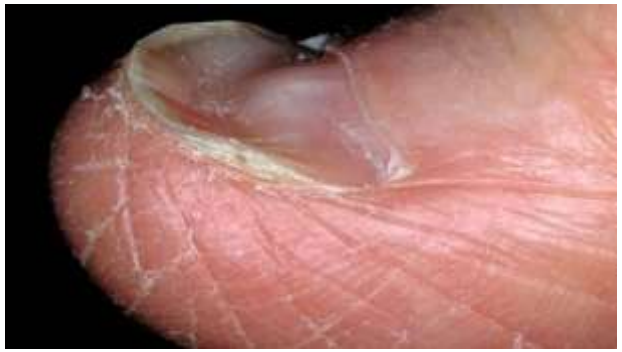
# Cutaneous Manifestations of Gastrointestinal Diseases

Plummer-Vinson syndrome → rare triad of dysphagia, iron deficiency anemia and esophageal webs



- Plummer-Vinson syndrome is associated with esophageal squamous cell cancer
- Skin signs include:
  - Brittle koilonychia (spoon-shaped, thin nail plates)
  - Oral leukoplakia (d/t intraepithelial carcinoma)
  - Angular cheilitis
  - Pale, atrophic buccal mucosa
  - Glossitis





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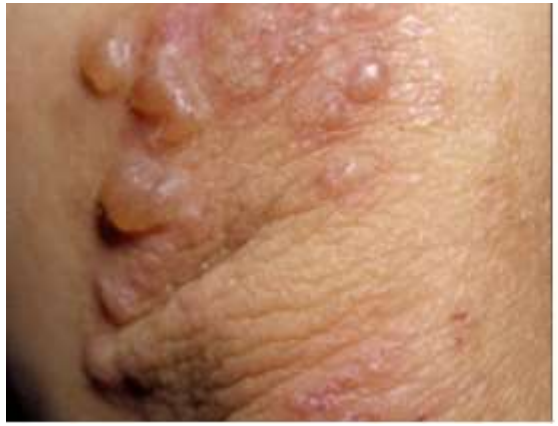
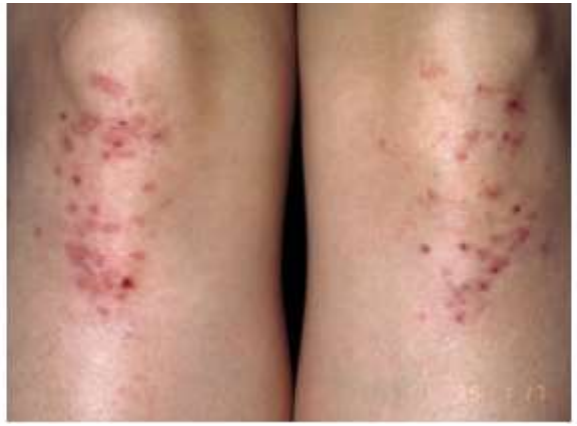
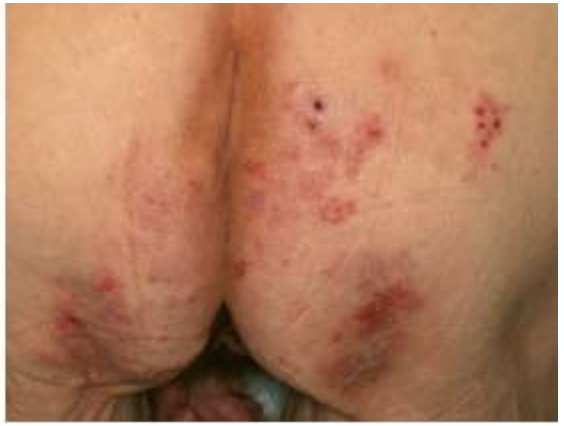
# Cutaneous Manifestations of Gastrointestinal Diseases

Dermatitis herpetiformis → associated with Human leukocyte antigens (HLA) DQ2 or DQ8 and with villous atrophy of duodenum (Celiac Disease)

Skin signs of dermatitis herpetiformis include:

- Pruritic herpetiform vesicles within erythematous/urticated plaques
- Excoriated erythematous, crusted, grouped papules
- Symmetrical distribution on extensor surfaces of elbows, knees, shoulders and buttocks

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# Cutaneous Manifestations of Hereditary Polyposis Syndromes

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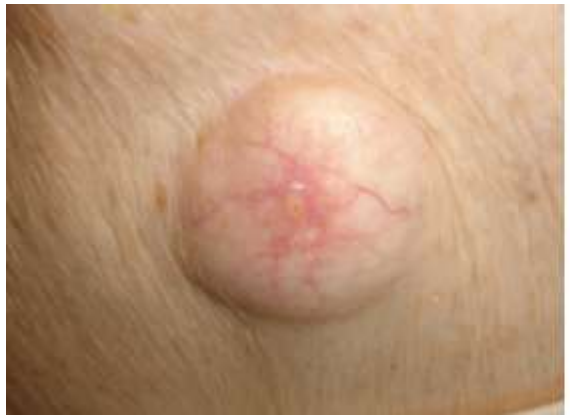
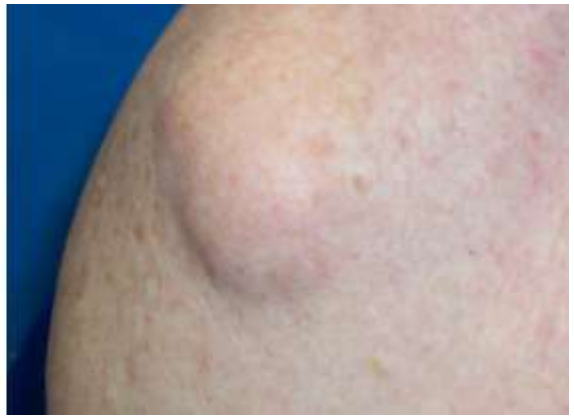
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

Gardner Syndrome → a variant of familial adenomatous polyposis with numerous adenomatous polyps and mucocutaneous findings:

- Epidermoid cysts on face or extremities
- Lipomas
- Desmoid tumors → non-tender, well-circumscribed, firm, flesh-colored tumors located on the abdomen wall, shoulder, chest wall and inguinal area





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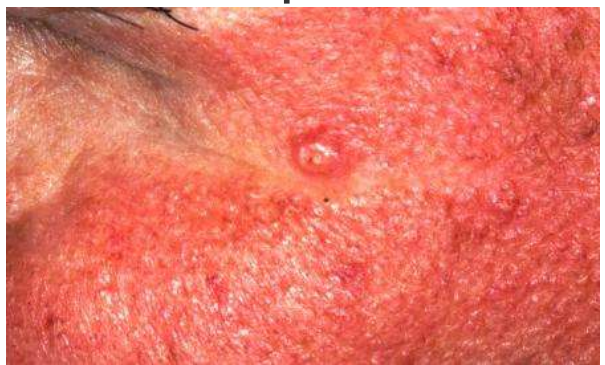
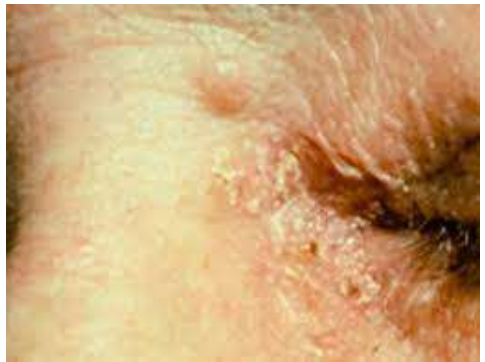
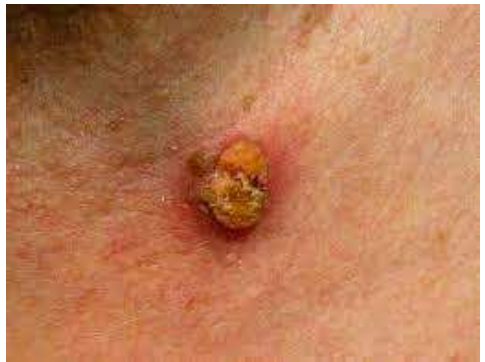
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

Hereditary nonpolyposis colorectal cancer (Lynch Syndrome) is the most common hereditary cancer, autosomal dominant disorder, accounting for 3% of all colon cancers

- Muir-Torre variant has most of the skin manifestations:
  - Sebaceous adenoma → yellow papules or nodules usually on the face
  - Sebaceous carcinoma → a periorbital or extraocular yellow nodule that may ulcerate and invade
  - Multiple keratoacanthomas and squamous cell cancers







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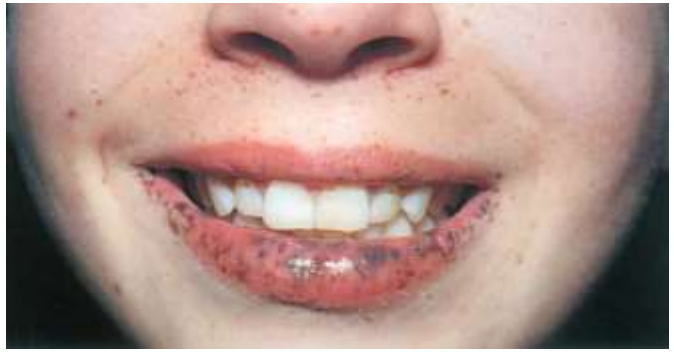
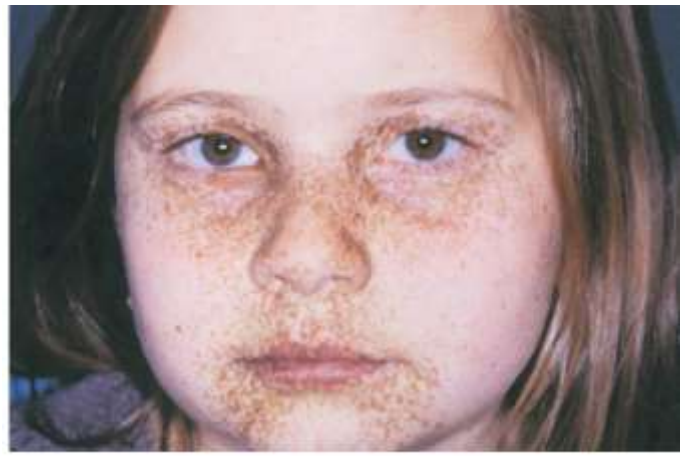
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

Peutz-Jeghers syndrome → hamartomatous polyposis syndrome with autosomal dominant inheritance

- Mucocutaneous pigmentation affects 95%, with small melanocytic macules that appear during childhood
- The lentigines cluster around the mouth, nostrils, eyes, digits, hands, feet, and perianal region (fade in adults)
- Oral-buccal pigmentation is usually permanent





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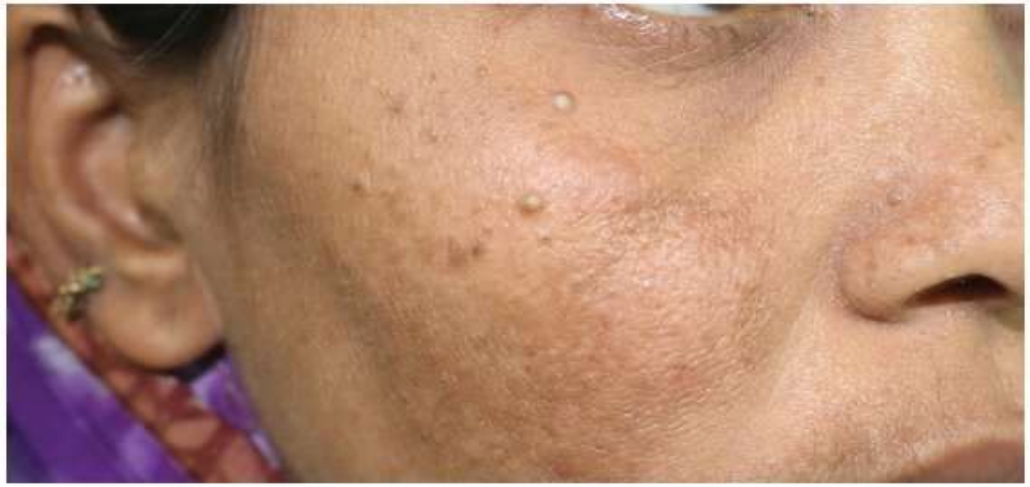
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

Cowden Syndrome (multiple hamartoma syndrome) → characterized by skin lesions and polyposis of the bowel

Trichilemmomas:

- Benign hamartomas of the outer sheath of hair follicles:
- Flesh-colored, smooth papules 1–5 mm in size on face, head, neck and hairline



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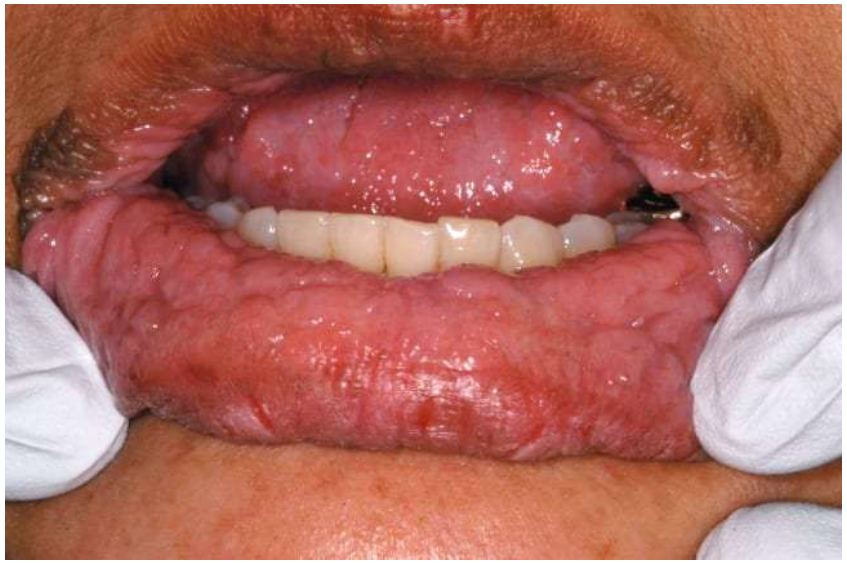
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

Other skin lesions described in Cowden syndrome include:

- Benign papillomas on the face, oral mucosa and acral surfaces
- Cobblestone appearance within the mouth
- Lipomas
- Café au lait macules
- Acral and plantar keratoses
- Fibromas
- Hemangiomas
- Furrowed tongue
- Neuromas
- Xanthomas
- Lentigines around the mouth, on hands, feet and genitals
- Vitiligo
- Acanthosis nigricans



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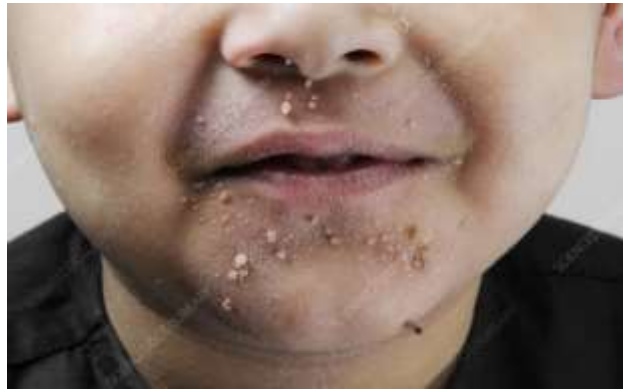
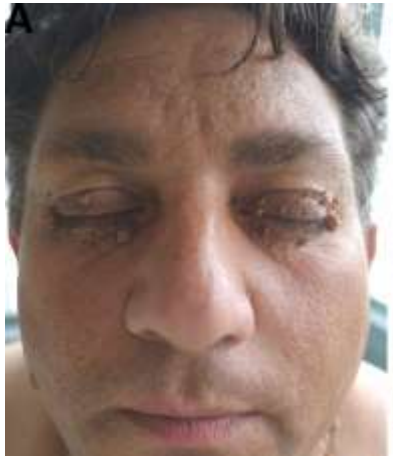
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

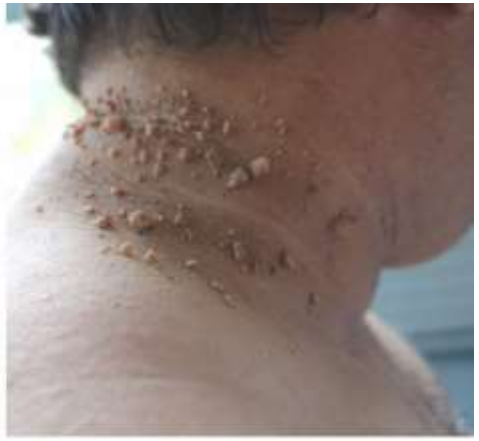
Bannayan-Riley-Ruvalcaba (BRR) syndrome→rare germline mutation with hamartomatous polyposis, macrocephaly and mental retardation

Dermatological manifestations:

- Genital lentigines
- Facial papillomas
- Lipomas
- Multiple acrochordons
- Acanthosis







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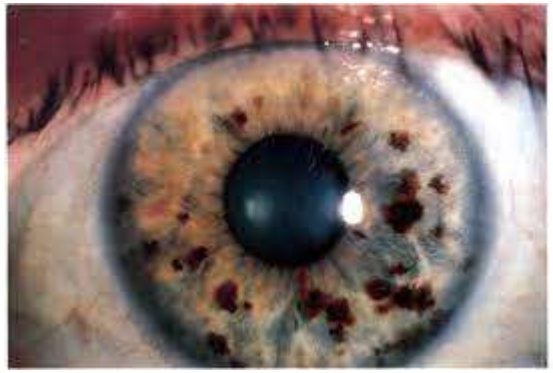
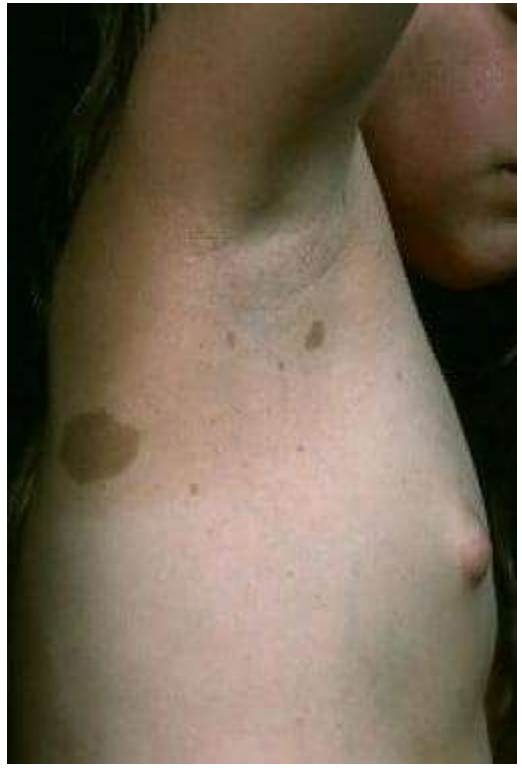
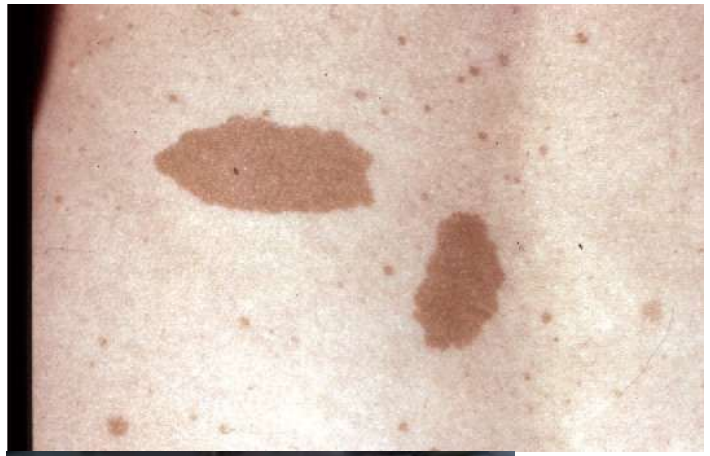
# Cutaneous Manifestations of Hereditary Polyposis Syndromes

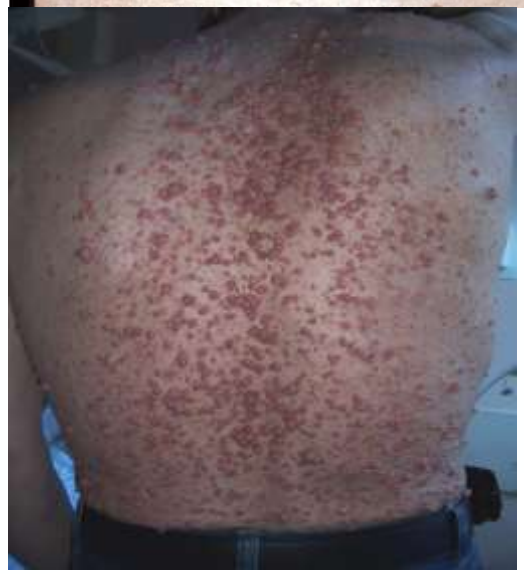
Neurofibromatosis → autosomal dominant neurodermatitis:

- Café au lait macules
- Axillary and inguinal freckling
- Dermal neurofibromas
- Lisch nodules of the iris









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# THANK YOU


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