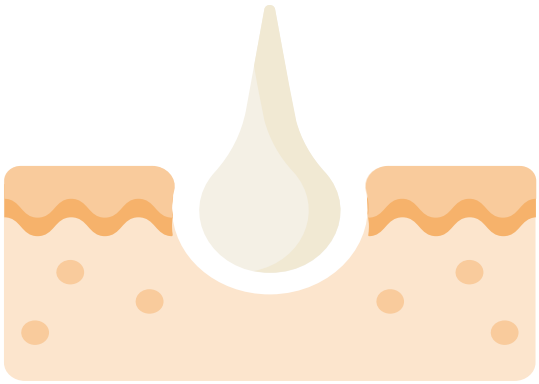


Cutaneous Manifestations Of Systemic Diseases

(Cutaneous Manifestations Of Systemic Diseases & Purpura And Vasculitis)

Objectives:

1. Not given
2.
3.

Male slides

Team leader:
Lama Alyahya

Done by:
Rema AlMutawa
Sarah Maghribi

Reviewd by
Noura & Alhanouf

Academic leader
Noura Alturki

Color index:

- Important
- Doctors Notes
- Extra

Contact us:
Dermatologyteam438@gmail.com

Editing file

Cutaneous Manifestations Of Systemic Diseases

Introduction:

Cutaneous manifestation of:

- Endocrine disorders
- Metabolic disorders (Hyperlipoproteinemias)
- GIT disorders
- Liver cirrhosis
- Renal failure

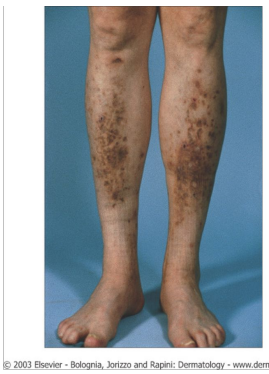
Cutaneous Manifestation of Endocrine Disorders

Cutaneous manifestation of:

- **Diabetes Mellitus**
- Hypothyroidism
- Hyperthyroidism
- Addison's disease
- Cushing syndrome

01 Diabetic Dermopathy or "Shin Spots":

- Most common cutaneous manifestation of diabetes; M >F, males over age 50 years with long standing diabetes
- Possibly related to diabetic neuropathy and vasculopathy
- There are bilateral asymptomatic red-brown atrophic macules on shins
- There is no effective treatment



02 Necrobiosis Lipoidica (NLD):

- Patients classically present with single or multiple red-brown papules, which progress to sharply demarcated yellow-brown atrophic, telangiectatic plaques with a violaceous,irregular border.
- Common sites include shins followed by ankles, calves, thighs and feet.
- Ulceration occurs in about 35% of cases. Cutaneous anesthesia, hypohidrosis and partial alopecia can be found

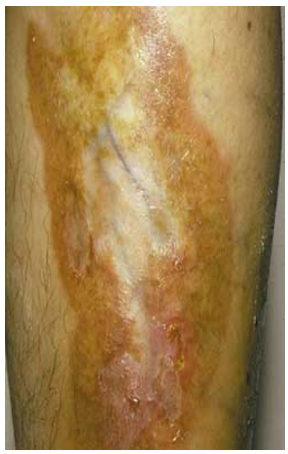
Pathology: Palisading granulomas containing degenerating collagen (necrobiosis).

What cause degeneration of collagen?

- Approximately 60% of NLD patients have diabetes & 20% have glucose intolerance. Conversely , up to 3% of diabetics have NLD.
- Women are more affected than men.
- Pathogenesis is thought to involve the nonenzymatic glycosylation of dermal collagen and elastin

Treatment: Ulcer prevention, No impact of tight glucose control on likelihood of developing NLD.

- Intralesional steroids - Aspirin -Antiplatelet -Pentoxyfylline.
- Preilesional heparin injection

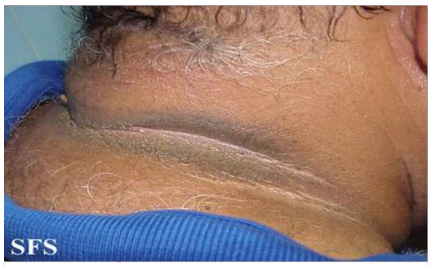


Cutaneous Manifestation of Endocrine Disorders

Cutaneous manifestation of:

- **Diabetes Mellitus**
- Hypothyroidism
- Hyperthyroidism
- Addison's disease
- Cushing syndrome

03 Acanthosis Nigricans:



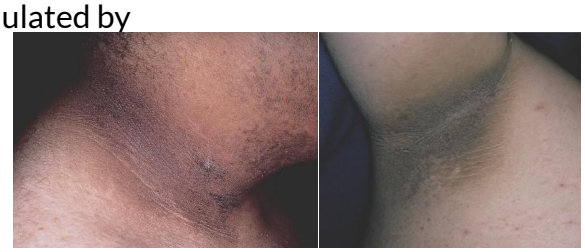
Hyperpigmented velvety plaques of the flexures any skin fold. The face, external genitalia, medial thighs, dorsal joints, lips and umbilicus can be involved in extensive cases (notice the skin tag)

Causes:

- obesity & insulin resistance & endocrinopathy (DM ,acromegaly ,cushing syndrome ,hypothyroidism & hyperandrogenic state as HAIRAN syndrome (hyperandrogen, insulin resistance, acanthosis nigricans)
- Malignancy (esp. GIT, Lung & Breast CA)
- Whenever you see Acanthosis nigrican extensively envolveing the palms think of malignancy we call it trip palm
- Medications (nicotinic acid ,niacinamide , testosterone, OCP & Glucocorticoid)

Pathogenesis:

- Genetic sensitivity of the skin to hyperinsulinemia
- Aberrant keratinocyte and fibroblast proliferation stimulated by excess growth factor(e.g., Insulin like growth factor)

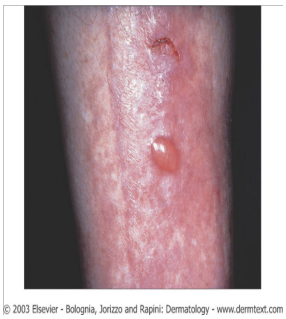


Treatment:

- Treat the underlying cause :

1. Tight blood glucose control,
2. treatment of underlying malignancy,
3. weight control
4. discontinuation of offending agent
5. Dose bleaching agent help ? No

04 Diabetic Bullae or Bullae Diabeticorum:



- Rarest cutaneous complications of diabetes; M > F, long standing diabetics.
- Trauma and microangiopathy may play a role

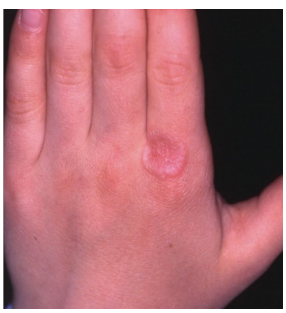
Clinical: Rapid onset of painless tense blisters on the hands and feet

Pathology:

- Intraepidermal and/or subepidermal split without acantholysis.
- DIF is negative

Treatment: Spontaneous healing without scarring

05 Granuloma Annulare:



- Association between granuloma annulare and diabetes is controversial.
- Asymmetric
- Generalized form of GA is the most closely associated with DM.
- It has a chronic and relapsing course

Treatment :

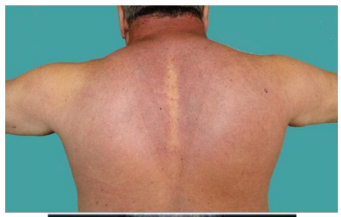
- IL steroid
- Systemic steroid
- PUVA

Cutaneous Manifestation of Endocrine Disorders

Cutaneous manifestation of:

- **Diabetes Mellitus**
- Hypothyroidism
- Hyperthyroidism
- Addison's disease
- Cushing syndrome

06 Scleredema Diabeticorum:



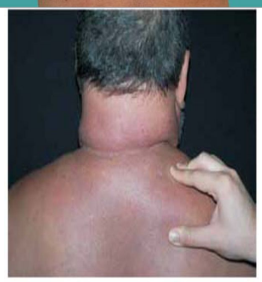


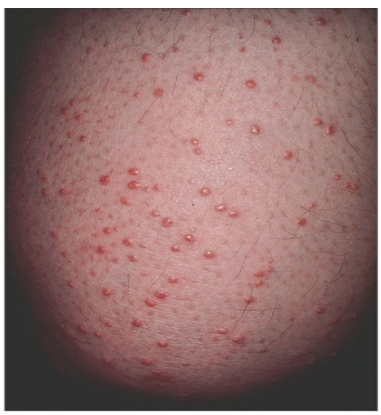
Figura 1. Nítida infiltração cutânea.

- Occurs diabetics with poorly controlled , long-standing disease, and obese men
- Painless, symmetric woody "peau d'orange" induration of the upper back and neck.
- It's Thinking of dermis (glycosamioglycan)
- No specific treatment is available
- Control of hyperglycemia does not improve the scleredema

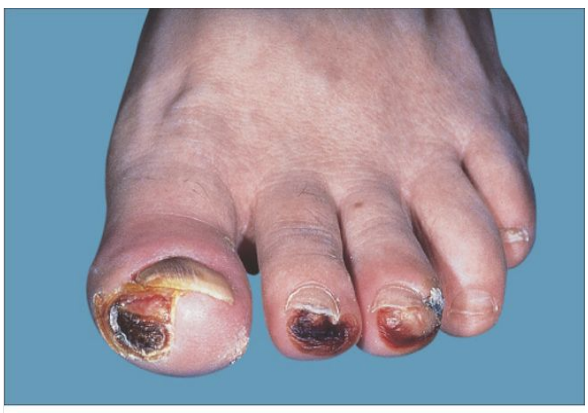
07 Cutaneous Infections:

- Diabetic patients are predisposed to develop cutaneous infections due to poor microcirculation (Bacterial, Fungal)

08 Other manifestation of DM:



Eruptive Xanthomas
Multiple legion in short period distributited all over the body (orange,red)



Diabetic neuropathy (peripheral) , Neuropathic ulcers

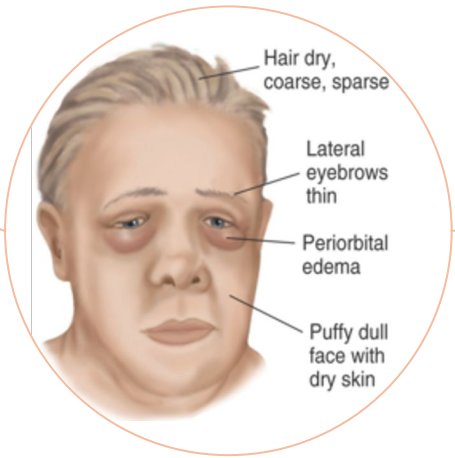
Cutaneous Manifestation of Endocrine Disorders

Cutaneous manifestation of:

Diabetes Mellitus | **Hypothyroidism** | Hyperthyroidism | Addison's disease | Cushing syndrome

Hair

- Dry, brittle, coarse hair
- Diffuse alopecia, Telogen effluvium
- Loss of lateral third of eyebrow (madarosis)



Skin

- Cool, dry, pale
- Xerosis
- Hypohidrosis
- Yellowish hue secondary to carotenemia
- Generalized myxedema: swollen waxy appearance
- Swollen lips, broad nose, macroglossia
- Purpura secondary to impaired wound healing
- Pruritus

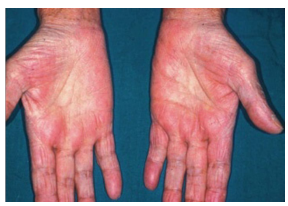
Cutaneous manifestation of:

Diabetes Mellitus | Hypothyroidism | **Hyperthyroidism** | Addison's disease | Cushing syndrome

Non-specific Manifestations of Hyperthyroidism:

Skin

- Warm, and moist
- Palmar erythema
- Flushing of head/neck, trunk
- pruritus



Nails

- Faster rate of growth
- Onycholysis
- Plummer nails: concave deformity with distal onycholysis

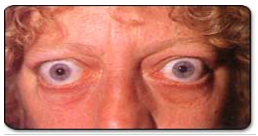
Hair

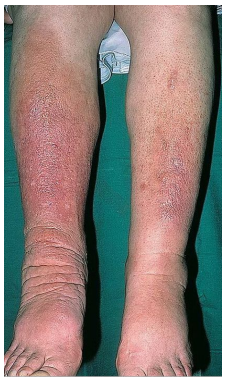
- Soft/fine/straight
- Diffuse reversible alopecia (Telogen effluvium)

Pigmentation

- Focal or generalized hyperpigmentation
- Vitiligo

Thyroid dermopathy (Pretibial Myxedema):





- Bilateral, non-pitting yellowish- brown to red waxy papules, nodules and plaques on the shins
- Occur in Graves disease triad (pretibial myxedema ,goiter,exophtalmus.

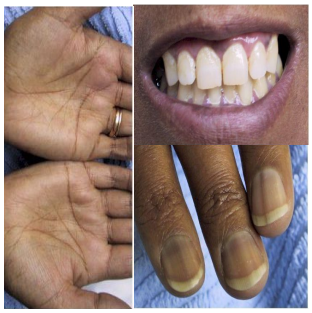
The clinical findings are due to an increase in (mucin) hyaluronic acid in dermis.
Treatment regimens include high potency topical steroids intralesional steroid.

Cutaneous Manifestation of Endocrine Disorders

Cutaneous manifestation of:

Diabetes Mellitus | Hypothyroidism | Hyperthyroidism | **Addison's disease** | Cushing syndrome

Hypocorticism (Addison disease):

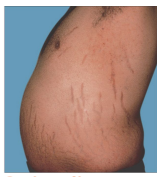


- Generalized hyperpigmentation that is more prominent in light exposed areas, scars, genitalia, palmar and finger creases, and under the nails. The pigmentation characteristically affects the mucous membranes areola.
- Loss of pubic and axillary hair in females.
- Improvement of acne
- Low steroid >increase ACTH >high MSH>hyperpigmintation

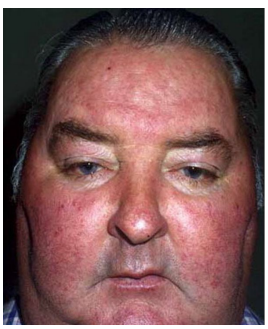
Cutaneous manifestation of:

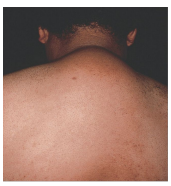
Diabetes Mellitus | Hypothyroidism | Hyperthyroidism | Addison's disease | **Cushing syndrome**

Cushing syndrome:



Striae distensae





Buffalo hump

- endogenous or exogenous
- Deposition of fat over the clavicles and back of the neck" Buffalo hump"
- Rounded erythematosus face with telangiectasia "Moon face"
- Truncal obesity with slender wasting limbs.
- Striae distensae -Hirsutism, acneform rash, androgenetic alopecia.
- Easy bruising of the skin on simple trauma.

Manifestations of endocrine diseases:

Table 53.6 Dermatologic manifestations of hypothyroidism.

DERMATOLOGIC MANIFESTATIONS OF HYPOTHYROIDISM	
Skin changes	Dry, rough, or coarse; cold and pale, boggy and edematous (myxedema) Yellow discoloration as a result of carotenemia Easy bruising (capillary fragility)
Cutaneous diseases	Ichthyosis and palmoplantar keratoderma Eruptive and/or tuberous xanthomas
Hair changes	Dull, coarse, and brittle Slow growth (increase in telogen hair phase) Alopecia of the lateral third of the eyebrows
Nail changes	Thin, brittle, striated Slow growth Onycholysis (rare)

Table 53.5 Dermatologic manifestations of hyperthyroidism.

DERMATOLOGIC MANIFESTATIONS OF HYPERTHYROIDISM	
Cutaneous changes	Fine, velvety, or smooth skin Warm, moist skin due to increased sweating Hyperpigmentation – localized or generalized
Cutaneous diseases	Vitiligo Urticaria, dermatographism Pretibial myxedema and thyroid acropachy
Hair changes	Fine, thin Mild, diffuse alopecia
Hair disease	Alopecia areata
Nail changes	Onycholysis Koilonychia Clubbing from thyroid acropachy

Table 53.7 Dermatologic manifestations of Cushing's disease (syndrome). *This same change is indicative of insulin resistance and occurs in HIV-associated lipodystrophy.

DERMATOLOGIC MANIFESTATIONS OF CUSHING'S DISEASE (SYNDROME)
Altered subcutaneous fat distribution*
• Rounded facies • Fullness of the cheeks ('moon' facies) • Dorsal cervical vertebral fat deposits (buffalo hump) (Fig. 53.20) • Pelvic girdle fat deposition • Reduced fat in the arms and legs
Skin atrophy
• Global atrophy with epidermal and dermal components affected • Multiple striae on abdominal flanks, arms, and thighs (Fig. 53.21) • Cutaneous fragility and prolonged wound healing • Purpura with minor trauma due to reduced connective tissue support
Cutaneous infections
• Pityriasis (tinea) versicolor • Dermatophytosis and onychomycosis • Candidiasis
Appendageal effects
• Corticosteroid-related acne • Hirsutism

Table 53.8 Selected dermatologic manifestations of Addison's disease. MSH, melanocyte-stimulating hormone.

SELECTED DERMATOLOGIC MANIFESTATIONS OF ADDISON'S DISEASE

• Hyperpigmentation (MSH-like effect due to secretion of ACTH)
  • Diffuse with sun-exposed accentuation
  • Sites of trauma
  • Axillary, perineum, and nipples
  • Palmar creases
  • Nevi
  • Mucous membranes
  • Hair
  • Nails
• Loss of ambisexual hair in postpubertal women
• Fibrosis and calcification of cartilage including the ear (rare)
• Vitiligo
• Chronic mucocutaneous candidiasis

Cutaneous Manifestation of Endocrine Disorders

Hyperlipoproteinemia

Types:

Type I
- Familial lipoprotein lipase deficiency (AR) or apoprotein CII deficiency
- Increased chylomicrons
- Associated with hepatomegaly, pancreatitis

Type IIa
- Familial hypercholesterolemia, common hypercholesterolemia (AD)
- Increased LDL

Type IIb
- Familial hypercholesterolemia (AD)
- Increased LDL and VLDL

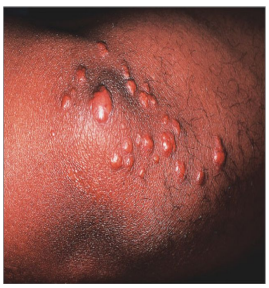
Type III
- Familial Dysbetalipoproteinemia (AR)
- Increased IDL

Type IV
- Familial hypertriglyceridemia (AD)
- Increased VLDL

Type V
- Familial type V hyperlipoproteinemia, familial lipoprotein lipase deficiency (AD)
- Increased chylomicrons and VLDL

01 Tuberous Xanthoma

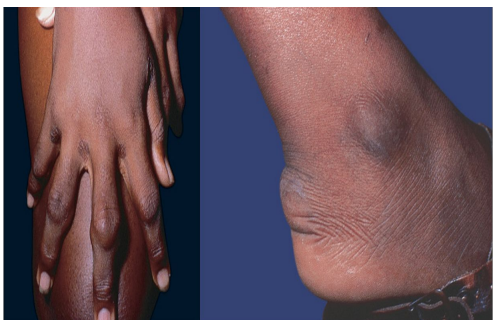
- Flat or elevated, rounded, grouped, yellowish- orange nodules over joints (particularly elbows and knees)
- Types II, III, and IV
- Biliary cirrhosis



Tuberous xanthomas of the knee.
Note the yellowish hue

02 Tendinous Xanthoma

- Papules or nodules over tendons (extensor tendons on dorsum of hands, feet, and achilles)
- Types II, III

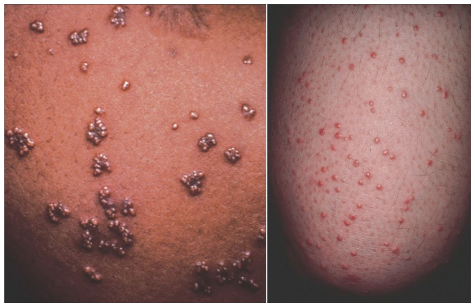


Tendinous xanthomas of the fingers in a patient with homozygous familial hypercholesterolemia

Linear swelling of the Achilles area representing a tendinous xanthoma in a patient with dysbetalipoproteinemia.

03 Eruptive Xanthoma

- Small yellow/orange/red papules appearing in crops over entire body → buttocks, flexor surfaces, arms, thighs, knees, oral mucosa and may koebnerize
- Associated with markedly elevated or abrupt increase in triglycerides (elevated chylomicrons)
- Types I ,III , IV , and V
- Diabetes, obesity, pancreatitis, chronic renal failure, hypothyroidism, estrogen therapy, corticosteroids, isotretinoin increase TG, acitretin



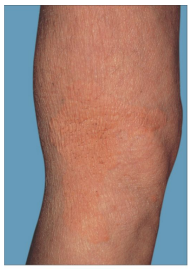
Eruptive xanthomas.
Note the yellowish hue

Cutaneous Manifestation of Endocrine Disorders

Hyperlipoproteinemia

04 Planar Xanthoma

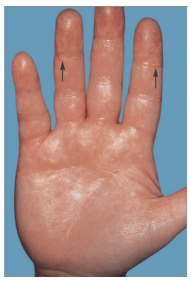
- Flat macules or slightly elevated plaques, yellow/ tan color
- Associated with biliary cirrhosis, biliary atresia, myeloma, monoclonal gammopathy, lymphoma.
- Characteristically around eyelids, neck, trunk, shoulders, or axillae
- Types II,III



Plane xanthoma in a patient with a monoclonal IgG gammopathy

05 Palmar Xanthoma

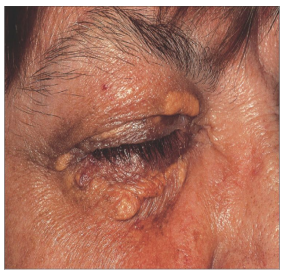
- Nodules and irregular plaques on palms and flexural surfaces of fingers
- Type III



Plane xanthomas of the palmar creases in a patient with dysbetalipoprotenemia (arrows)

06 Xanthelasma

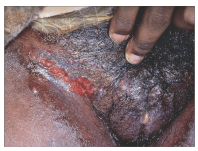
- Most common type of xanthoma
- Eyelids
- Usually present without any other disease dont be surprised if you get normal lipid profile, but can occur in types II and III
- Common among women with hepatic or biliary disorders, also seen in myxedema, diabetes
- Best treated with surgical excision



Xanthelasma palpebrarum with typical yellowish hue.

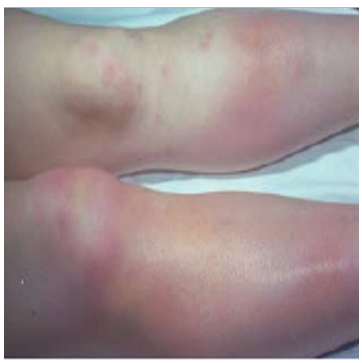
Cutaneous Manifestation of Gastrointestinal Disorders

Manifestations of Inflammatory Bowel Disease (IBD)

Cutaneous findings	Association	
Commonly involves Perineum associated with edema and inflammation	CD>UC	Fissures and Fistulas
Edema, cobblestone, ulceration, nodules	CD	Oral Crohn's
Nodules, plaques, ulcerations; commonly on extremities or intertrigenous regions mimics Erythema Nodosum	CD	Metastatic Crohn's 
Tender red nodules on anterior lower legs; precedes or occurs simultaneous with IBD flare	UC>CD	Erythema nodosum
Papules, pustules, hemorrhagic blisters → enlarge, ulcerate with dusky undermined edges; exacerbated by trauma; frequently on legs	UC>CD	Pyoderma Gangrenosum (PG)
Vegetating plaques, vesiculopustules of intertrigenous areas; heal with hyperpigmentation; when process involves mucosa =Pyostomatits vegetans	UC	Pyoderma Vegetans
Identical to common aphthous ulcers; develop with IBD flares	UC>CD	Chronic Apthous Ulcers

Other less common manifestation: Epidermolysis bullosa acquisita, erythema multiforme, urticaria, clubbing, psoriasis, vitiligo. Note: CD = Crohn's disease ,UC = Ulcerative Colitis

Erythema Nodosum:



- Erythematous, tender nodules on anterior shins; also seen on thighs, lateral aspects of lower legs, arms, and face , bilateral , symmetrical.
- Often accompanied by fever, chills, malaise, and leukocytosis
- 70% have associated arthropathy
- Occurs at any age, but most prevalent between 20 and 30 years of age
- Nodule in subcutaneous fat

Causes : MNEMONIC SHOUT BCG
S=Sarcoid, Sulfa drugs, Strept commonest cause. H=Histoplasmosis. O=Oral contraceptives, pregnancy. U=Ulcerative colitis. T=TB. B=Bechet's. C=Crohns. G=GI (Yersinia, salmonella)

Work up:
- Hx (exclude drugs , hx of infection & GI symptoms)
- CBC with diff, ESR
- Throat swab , ASO titre
- CXR -PPD
- Stool for occult blood

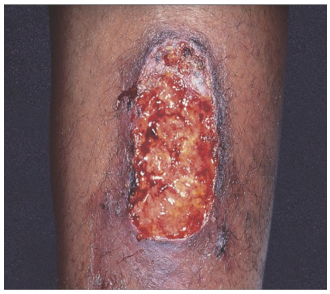
Histology: Septal panniculitis without vasculitis

Treatment:
- Spontaneous resolution usually occurs within three to six weeks without scarring
- NSAIDs such as indomethacin or naproxen
- Systemic steroids effective in
- severe cases and can be dangerous if infection is etiology
- Potassium iodide if chronic and recurrent

Cutaneous Manifestation of Gastrointestinal Disorders

Pyoderma gangrenosum:

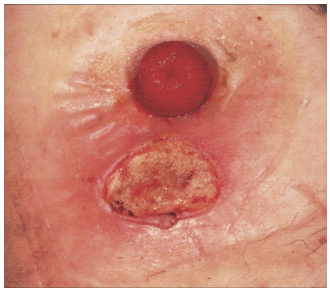
- 1.5-5% of patients with IBD develop PG
- Associated with leukemia, myeloma, monoclonaL gammopathy (IgA), polycythemia, chronic active hepatitis, HCV , HIV , SLE & pregnancy
- Associated with PAPA syndrome → pyogenic arthritis, pyoderma gangrenosum, severe cystic acne
- May be associated with arthritis



Distinct rolled edges and show satellite violaceous papules that break down and fuse with central ulcer

Types:

- Ulcerative
- Pustular end up with ulcer
- Bullous end up with ulcer
- Vegetative



Peristomal Pyoderma Gangrenosum

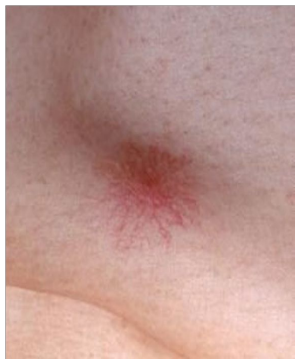
Histology: Septal panniculitis without vasculitis

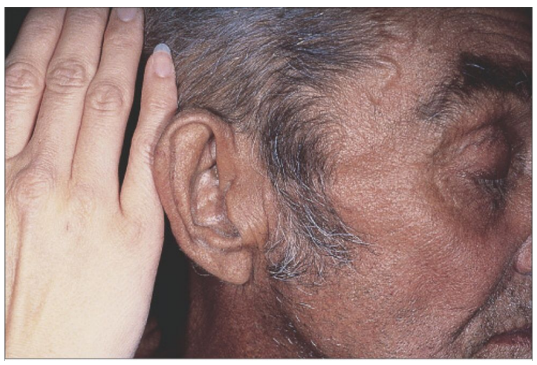
Treatment:

- Treat underlying cause
- Potent topical steroids or IL steroids
- Topical tacrolimus
- Systemic steroids
- Cyclosporine ,Sulfapyridine, sulfasalzine, and dapsone
- Infliximab
- Other agents: thalidomide, SSKI, azathioprine, cyclophosphamide, chlorambucil

Cutaneous Manifestation of Liver Diseases

- Pruritus: generalized itching especially in the presence of biliary obstruction or jaundice.
- Jaundice. -Spider naevi: small telangeictatic blood vessels especially on the face and upper chest.
- Palmar erythema.
- Thinning of the hair and sometime loss of sexual hair in the axillae and pubic areas. Because of sex hormon binding protien
- Porphyria cutanea tarda.
- Xanthoma





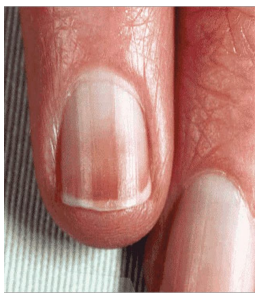
Diffuse bronzing of the skin in Hemochromatosis

Cutaneous Manifestation of Renal Diseases

End Stage Renal Disease (ESRD) and Dialysis:

01 Pruritus:

- The most common cutaneous manifestation of ESRD



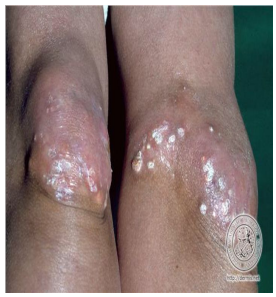
02 Half and half (Lindsay's) nails:

- Result from edema of the nail bed and capillary network and give the proximal half of the nail an opaque white appearance

03 Metastatic Calcification:

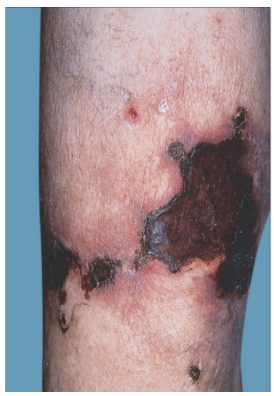
- Deposition of calcium within tissue secondary to abnormal calcium and or phosphate metabolism hyperparathyrodism
- It can manifest in the skin as:

1. benign nodular calcifications (calcinosis cutis)
2. Or as a more serious condition (calciphylaxis) CA in small blood vessele with an associated mortality rate between 60-80%



04 Calciphylaxis:

- Calciphylaxis presents as painful purpuric plaques and retiform pupura with progression to ulceration and necrosis. Distribution of the lesions may predict prognosis; patients with acral lesions have a better outcome than those with proximally located lesions



Histological finding: medial calcification/intimal hyperplasia of small arteries and arterioles

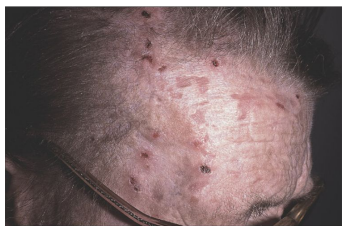
Management: includes

- total or subtotal parathyroidectomy (if PTH levels are elevated)
- wound care
- avoidance of precipitating factors.
- Mortality is related to Staphylococcal superinfection of ulcers with resultant sepsis
- Change the diet

05 Porphyria Cutanea Tarda (PCT):

- **Pathogenesis:** may be related to the suboptimal clearance of uroporhyrins (product of heme synthesis pathway) from the circulation which is a photosensitizer .
- Patients may present with:

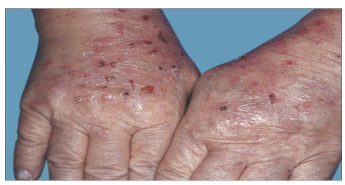
1. photodistributed bullae.
2. skin fragility
3. hyperpigmentation.
4. Hypertrichosis



Thy have enzymatic deficiency either acquired (renal and liver disease) inherited (gentaic deficiency)

05 Pseudo-PCT:

- Similar clinical and histological findings of PCT, in setting of normal porphyrin profile
- Usually due to certain medications such as furosemide, naproxen, tetracycline, nalidixic acid, or amiodarone



Cutaneous Manifestation of Renal Diseases

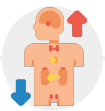
Generalized Pruritus

- Generalised pruritus in the absence of a rash requires investigation and exclusion of an underlying systemic disorder
- It is important to distinguish these from an underlying primary skin disease such as scabies or eczema

Conditions that Cause Pruritus:

Chronic renal disease

Cholestasis



Endocrine diseases

- **Thyrotoxicosis** – often due to increased skin blood flow which raises skin temperature
- **Hypothyroidism** – pruritus secondary to the dry skin

Malignancy

Most common association: Hodgkin's disease and polycythemia rubra vera

Infections:
1. HIV
2. Parasetic

Iron deficiency anemia

Workup of Generalized Pruritus:

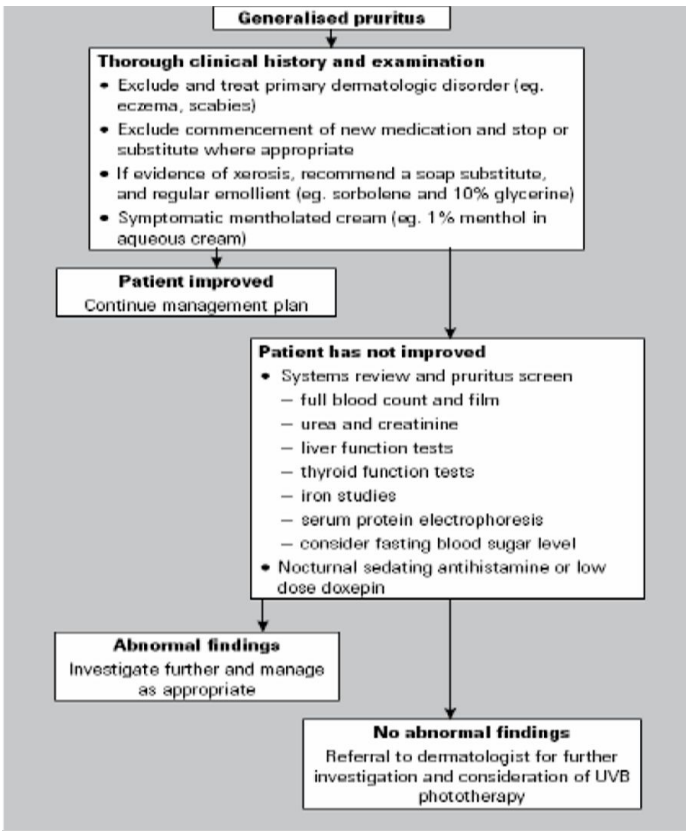
- History and Physical exam
- CBC, diff , Blood film
- Stool for O&P, occult blood
- CXR
- Thyroid, renal, and liver function tests

Table 3. Systemic Etiologies for Pruritus

Autoimmune	**Malignancy**
Dermatitis herpetiformis	Leukemia
Dermatomyositis	Lymphoma
Linear immunoglobulin A disease	Multiple myeloma
Sjögren syndrome	Solid tumors with paraneoplastic syndrome
Hematologic	**Metabolic and endocrine**
Hemochromatosis	Carcinoid syndrome
Iron deficiency anemia	Chronic renal disease
Mastocytosis	Diabetes mellitus
Plasma cell dyscrasias	Hyper/hypothyroidism
Polycythemia vera	Hyperparathyroidism
Hepatobiliary	**Neurologic**
Biliary cirrhosis	Cerebral abscess
Chronic pancreatitis with obstruction of biliary tracts	Cerebral tumor
Drug-induced cholestasis	Multiple sclerosis
Hepatitis, particularly hepatitis C	Stroke
Sclerosing cholangitis	**Other**
Infectious disease	Drug ingestion
AIDS	Eating disorders with rapid weight loss
Infectious hepatitis	Neuropsychiatric disorders
Parasitic disease (giardiasis, onchocerciasis, schistosomiasis, ascariasis)	Pregnancy
Prion disease	

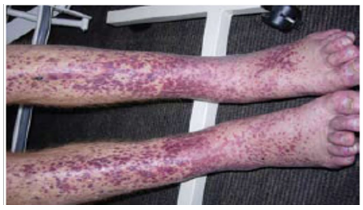
Information from references 2, 3, 5, 7, 9, and 10.

Approach:



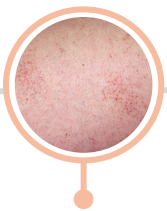
Purpura and vasculitis

Purpura and vasculitis

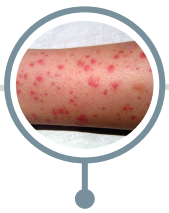


Definition:

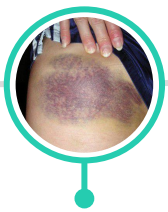
- Visible hemorrhage into the skin or mucous membrane subdivided as a follow:



Petechiae:
less than or equal 4 mm



Purpura:
(>4mm - < 1cm) which can be either Palpable or non palpable(macular)



Ecchymoses:
> or equal to 1 cm

Causes:

Platelet Disorders	Coagulation Factor Deficiency	Vascular Factors (Blood vessel wall pathology)
Thrombocytopenia	Congenital: - Factor VIII Deficiency - Factor IX Deficiency - Von Willebrands disease	Congenital: - Hereditary Hemorrhagic Telangectasia - Ehlers-Danlos Syndrome (Type IV)
Platelet Dysfunction	Acquired: - Disseminated Intravascular Coagulopathy - Liver disease - Uremia - Vitamin K deficiency	Acquired: - Inflammation(Vasculitis) - Trauma - Vitamin c deficiency (scurvy)

Vasculitis:

- A clinicopathologic process characterized by inflammatory destruction of blood vessels that results in occlusion or destruction of the vessel and ischemia of the tissues supplied by that vessel.

Classification:

- **Large-vessel vasculitis**

Anatomy: Aorta and the great vessels (subclavian, carotid)
Pathology: Claudication, blindness, stroke

- **Medium-vessel vasculitis**

Anatomy: Arteries with muscular wall EX.renal ,coronary artery
Pathology: Mononeuritis multiplex (wrist/foot drop), mesenteric ischemia, cutaneous ulcers
They present to dermatologist if it comes with small vessel

- **Small-vessel vasculitis**

Anatomy: Capillaries, arterioles, venules
Pathology: Palpable purpura, glomerulonephritis, pulmonary hemorrhage

CHAPEL HILL CONSENSUS CLASSIFICATION
Large-vessel vasculitis
• Giant cell arteritis • Takayasu's arteritis
Medium-vessel vasculitis
• Classic polyarteritis nodosa • Kawasaki disease
Small-vessel vasculitis
• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis (polyarteritis) • Henoch–Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis

Purpura and vasculitis

Cutaneous small vessel vasculitis

- Is the most common type of vasculitis and it primarily affect post-capillary venules

Pathogenesis:

- Caused by circulating immune complexes
- Tybe 3 immune Response
- These lodge in vessel walls and activate complement

Features:

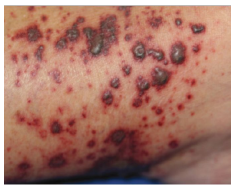
- Palpable purpura is the hallmark
- Pinpoint to several centimeters differentiated from Palpable vesicles in the picture
- Early on lesion may not be palpable,Papulonodular, bullous, pustular or ulcerated forms may develop
- Predominate on the ankles and lower legs i.e dependent areas
- HSV seen in pediatric

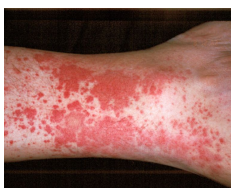
Table 3. Causes of cutaneous vasculitis[5,6]

Infections	
Bacterial	• Streptococcal, meningococcal, urinary tract infections
Viral	• Hepatitis B and C, HIV
Mycobacterial	• Tuberculosis
Connective tissue disorders	• SLE and related conditions • Rheumatoid arthritis • Systemic sclerosis, Sjogren syndrome • Dermatomyositis • Medium vessel vasculitides (Wegener granulomatosis, polyarteritis nodosa, Churg-Strauss syndrome)
Malignancy	• Haematologic – myeloproliferative – lymphoma – monoclonal gammopathy – multiple myeloma
Drugs	Including antibiotics, antihypertensives
Idiopathic	Henoch-Schonlein purpura

Manifestations of the disease:

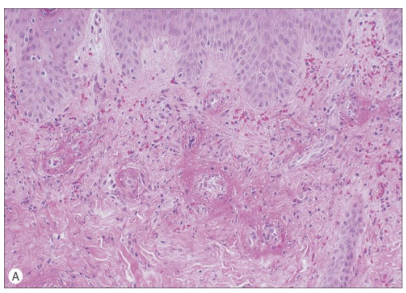
- Mild pruritis, fever, malaise, arthralgia and/or myalgia may occur
- Typically resolve in 3 to 4 weeks
- Residual postinflammatory hyperpigmentation may be seen
- Self-limiting
- May recur or become chronic
- Hemorrhagic vesicles or bullae may develop
- May be localized to the skin or may manifest in other organs.
- The internal organs affected most commonly include the joints, GIT, and the kidneys.
- Renal involvement present as glomerulonephritis
- The prognosis is good in the absence of internal involvement

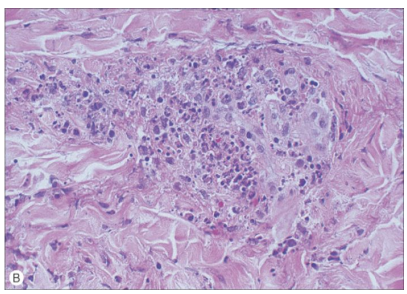




Histology:

Agiocentric segmental inflammation, endothelial cell swelling, fibrinoid necrosis of blood vessel walls(destruction of vessel wall and lumen)and a cellular infiltrate composed of neutrophil with RBC extravasation.fibrin deposition in vessel wall





Work up

- Detailed history and physical examination
- History should focus on possible infectious disorders, prior associated diseases, drugs ingested, and a thorough review of systems
- CBC, strep throat culture or ASO titer, Hep B & C serologies and ANA are a reasonable initial screen, renal profile
- **Urinalysis for RBC , proteins & cast**
- Skin biopsy

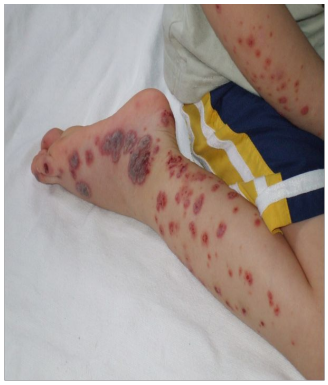
Treatment

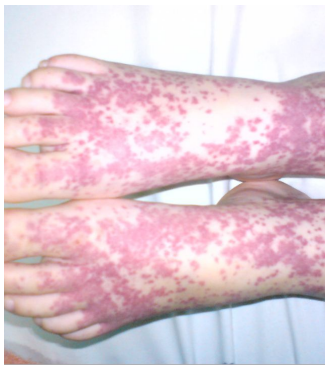
- Treatment of cause.
- Symptomatic treatment (if skin is only involved): rest , NSAIDS, Antihistamine
- severe visceral involvement may require high doses of corticosteroids with or without an immunosuppressive agent
- Immunosuppressive agents for rapidly progressive course and severe systemic involvement

Purpura and vasculitis

Henoch-Schönlein purpura (HSP)

- Primarily occurs in male children
- Peak age 4-8 years
- Adults may be affected
- A viral infection or streptococcal pharyngitis are the usual triggering event
- In about 40 % of the cases the cutaneous manifestations are preceded by mild fever, headache, joint symptoms, and abdominal pain for up to 2 weeks





Manifestations of the disease:

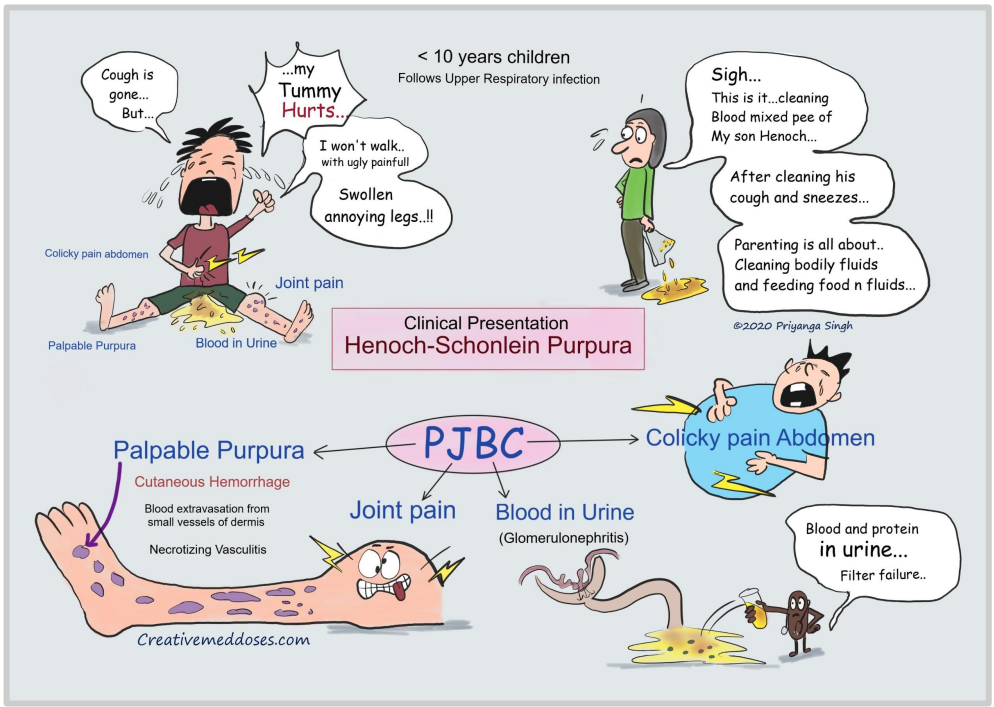
- Characterized by intermittent purpura, arthralgia, abdominal pain, and renal disease
- Typically purpura appears on the extensor surfaces of the extremities
- Become hemorrhagic within a day and fades in 5 days
- New crops appear over a few weeks
- May be associated with: pulmonary hemorrhage, Abdominal pain and GI bleeding
- GI radiographs may show “cobblestone” appearance
- Renal manifestations may occur in 25% or more but only 5% end up with ESRD

Diagnosis:

- IgA, C3 and fibrin depositions around the blood vessel have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques
- When You take biopsy and give one sample for histo the other for IF in IF you found IGA and C3 only in the first 48 H

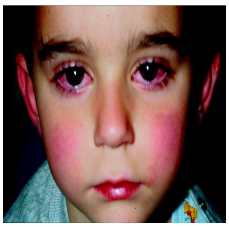
Prognosis:

- The long-term prognosis in children with gross hematuria is very good; however, progressive glomerular disease and renal failure may develop in a small percentage



Purpura and vasculitis

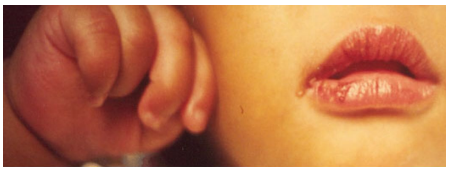
Mucocutaneous lymph node syndrome (Kawasaki's disease)



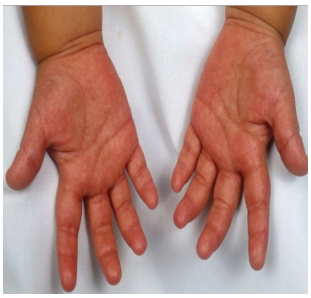
- Predominantly seen in children less than 5 years of age.
- Occurs most often in Japan
- It is coronary disease mainly

Diagnosis:

A patient should have a fever above 38.3 C for 5 days plus 4 of the 5 following criteria;



1. Edema of hands and feet
2. Polymorphous exanthema
3. Nonpurulent bilateral conjunctival injection
4. Changes in the lips and oral cavity
5. Acute, nonpurulent cervical adenopathy

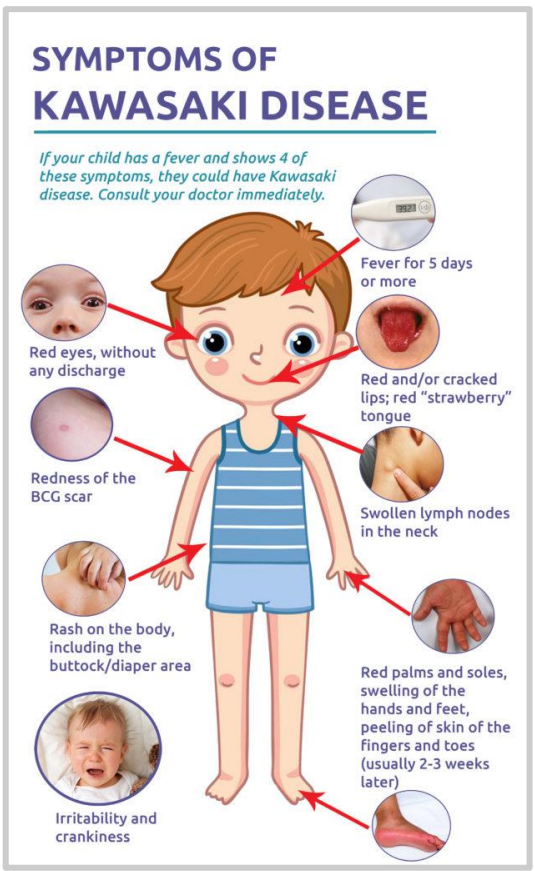


Complications:

- Coronary arterial disease occurs and thrombocythemia may occur
- In combination, vessel occlusion may occur and the subsequent MI, which occur as the child is recovering from the acute illness

Treatment:

- IVIG is the cornerstone of treatment
- Antiplatelet therapy with aspirin is recommended
- (high dose)



Questions

1- **Which of the following skin rashes is associated with diabetes mellitus?**

A) Acanthosis nigricans
B) Palpable purpura
C) Pretibial myxedema
D) Striae

2- **Which of the following is not an indication for HIV testing?**

A) Any STD
B) Oral hairy leukoplakia
C) Seborrheic Dermatitis
D) lichen planus

3- **What is the best treatment for Cutaneous small vessel vasculitis without systemic involvement?**

A) Methotrexate
B) Bed rest
C) Steroid

4- **What is pathognomonic sign for small vessel vasculitis?**

A) Vesicles
B) Bulla
C) Palpable purpura
D) Scar

Answers:
1 : A , 2 : D , 3 : B , 4 : C , 5: D, 6: D