

CUTANEOUS MANIFESTATIONS OF SYSTEMIC DISEASES

Dr. Saleh Alrasheed

Objectives

To highlight the relation between skin manifestations and common systemic disorders.

To understand various skin clues and their importance in investigating and managing different systemic diseases

Contents:

- Introduction:

- u Endocrinological Diseases

- v Diabetes Mellitus

- v Hyperthyroidism

- v Hypothyroidism

- v Cushing's Syndrome

- v Addison's disease

● GIT:

- u Chronic Liver Disease (CLD)
- u Acrodermatitis entropathica
- u Peutz Jeghers Syndrome
- u Pyoderma Gangrenosum
- u Hereditary hemaorrhgic Telangectasia

● Metabolic:

- u Hyperlipidemia

- Neurocutaneous diseases

1. Neurofibromatosis
2. Tuberous Sclerosis

- Behcet's Syndrome

- Nutritional deficiency disorders

- 1 Scurvy
- 2 Pellagra

- Causes of Generalized pruritus without skin lesions

● Cutaneous manifestations of internal malignancy.

- u Acanthosis Nigricans
- u Dermatomyositis

● Nails:

- u Clubbing
- u Koilonychia
- u Splinter haemorrhages

● When to do HIV testing for skin Disease ?

Skin and endocrine system

Diabetes mellitus.

Thyroid diseases.

Cushing's syndrome.

Addison's disease.

Diabetes mellitus

- Skin tags.
- Acanthosis nigricans.
- Diabetic dermopathy.
- Bullous diabeticorum.
- Thickening of skin.
- Necrobiosis lipoidica diabeticorum.
- Bacterial and fungal infections.
- Perforating dermatosis.

Skin tags

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

Acanthosis nigricans

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

Diabetic dermopathy

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

Bullous diabeticorum

- ❑ Rare cutaneous complications of diabetes; M > F, long standing diabetics.
- ❑ Trauma and microangiopathy may play a role.
- ❑ Clinical picture: Rapid onset of painless tense blisters on the hands and feet.
- ❑ Pathology: Intraepidermal and/or subepidermal split without acantholysis. DIF is negative, because its not an inflammatory bullae.
- ❑ Treatment: Spontaneous healing in two to five weeks

Granuloma Annulare:

- ❑ Clinical picture: Asymptomatic red-purple dome shaped papules arranged in annular configuration.**
- ❑ Association between granuloma annulare and diabetes is controversial. Usually when there is multiple granuloma annulare there is more risk of association with DM.**
- ❑ Generalized form of GA is the most closely associated with DM, about 20%.**
- ❑ It has a chronic and relapsing course.**
- ❑ Treatment :**
 - ❑ Intralesional steroid.**
 - ❑ Systemic steroid.**
 - ❑ PUVA and many other options.**
 - ❑ Controlling blood sugar WILL NOT improve GA.**

Thickening of skin

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

Scleredema Diabeticorum:

- ☒ Occurs more commonly in type 2 diabetics, long-standing disease, and obese men (M>F).**
- ☒ Clinical picture: Painless, symmetric woody “peau d’orange” indurations and thickening of the upper back and neck (Interscapular area), and usually it affect the shoulder motility.**
- ☒ No specific treatment is available**

Necrobiosis lipoidica diabeticorum

- Yellow atrophic plaques on the shins.
- Sometimes they ulcerate.
- Histopathology shows tiered granulomatous reaction.
- Treatment with topical, intralesional steroids, tacrolimus, phototherapy, cyclosporine, and rarely surgery.

Bacterial and fungal infections

- Pyodermmic infections such as impetigo, folliculitis, carbuncles, furunculosis, ecthyma, and erysipelas can be more severe and widespread in diabetic patients.
- Erythrasma, caused by *Corynebacterium minutissimum* mostly on axillae and groin.
- malignant otitis externa, often caused by *Pseudomonas aeruginosa*.
- Tinea pedis and onychomycosis.
- Candidal infections like perleche on corners of mouth, and on vulva.
- Rare infections like mucormycosis by Phycomycetes and anaerobic cellulitis by *Clostridium* species.

Perforating dermatosis

- Pruritic hyperkeratotic papules on the legs and trunk.
- Histopathology shows transepidermal elimination of collagen and/or elastin.
- Common in patients with diabetes and renal failure.
- treatments include topical keratolytics, phototherapy, topical and systemic retinoids, topical and intralesional steroids, oral antihistamines, and cryotherapy.

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.

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

Hypothyroidism

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

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)

Cushing's syndrome

- caused by prolonged exposure to high levels of plasma glucocorticoid.
- Adrenocortical hyperplasia.
- Benign or malignant adrenal tumours.
- Ectopic ACTH syndrome – secretion of ACTH by malignant or benign tumours arising in structures other than the pituitary or adrenal glands.
- Exogenous steroid administration
- Acne and hirsutism.
- Clitromegaly and male pattern alopecia.
- Striae.
- Easy bruising and purpura.
- Moon face and buffalo hump with fat redistribution.
- Telangectasia on face.
- Poor wound healing.

● Addison's disease:

Hyperpigmentation:

at Sun exposed skin, sites of trauma, axillae, palmar creases, old scars ,nevi and mucous membranes

Adrenocortical hypofunction.

Diffuse pigmentation on skin and mucous membranes.

Melanocytes stimulation by ACTH

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

Gastrointestinal disease

- Dermatitis herpetiformis.
- Acrodermatitis enteropathica.
- Pyoderma gangrenosum.
- Peutz Jeghers syndrome.
- Porphyria cutanea tarda.
- Hemochromatosis.
- Liver cirrhosis.

Dermatitis herpetiformis

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

Acrodermatitis enteropathica

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

Peutz Jeghers syndrome

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

Pyoderma gangrenosum

- a painful, ulcerative lesion with a well-defined, undermined violaceous border.
- start as small pustules, which subsequently burst and expand to form the larger noninfectious ulcer.
- Positive pathergy test.
- Mostly associated with ulcerative colitis. Also with Crohn's disease, rheumatoid arthritis, and leukemia.
- Surgery is contraindicated.

Porphyria cutanea tarda

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

Hemochromatosis

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

Renal diseases

- **Xerosis** occurs in 50-92% of the dialysis population.
- Some patients may develop acquired ichthyosis.
- the exact cause of xerosis in ESRD remains unknown.
- Many patients respond to routine use of emollients.
- **Pruritus** affects 15-49% of patients with chronic renal failure and 50-90% of the dialysis population.
- Uremia is the most common metabolic cause of pruritus.
- Cutaneous manifestations of pruritus include excoriations, prurigo nodularis, and lichen simplex chronicus.
- Pruritus typically resolves after transplantation.
- Treatment include sedating antihistamines, emollients, phototherapy, thalidomide, and gabapentin .

Renal diseases

- **Half and half nails** occur in around 40% of patients on dialysis.
- Kidney transplant usually resolve this sign.
- Usually involve fingernails.

Renal diseases

- **Nephrogenic systemic fibrosis** mostly seen in ESRD and dialysis patients.
- Presents as thick, indurated plaques on the extremities and the trunk similar to scleroderma.
- gadolinium might have a role in the pathogenesis of this condition.
- Treatment includes immunosuppressive agents, phototherapy, topical steroids, retinoids, and photophoresis.

hyperlipidemia

- **Xanthelasma palpebrarum** is the most common of the xanthomas.
- asymptomatic and usually bilateral and symmetric.
- Can be associated with any type of primary hyperlipoproteinemia. And could be without hyperlipidemia.
- often treated with topical trichloroacetic acid, electrodesiccation, laser therapy, and surgical excision.

Hyperlipidemia

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

hyperlipidemia

- **Tuberous xanthomas** are firm and nontender cutaneous and subcutaneous yellowish nodules on extensor surfaces.
- Mostly associated with familial dysbetalipoproteinemia.
- May resolve after months of treatment with lipid lowering agents.

Hyperlipidemia

- Eruptive xanthomas are painless, yellowish papules on an erythematous base that present as grouped lesions on trunk, elbows and buttocks.
- Usually associated with hypertriglyceridemia.
- Could be seen in poorly controlled diabetes and acute pancreatitis.
- Usually resolve in few weeks after therapy.

Hyperlipidemia

- Planar xanthomas are elevated cutaneous yellowish-orange deposits on palmar creases.
- Usually associated with familial dysbetalipoproteinemia.

● Types of Xanthomas

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

Neurocutaneous Disorders

■ Neurofibromatosis:

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

● Tuberos Sclerosis (Epiloia) :

- √ Epi = Epilepsy
- √ Loi = Low intellegence
- √ a = adenoma sebaceoum

● Skin features

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

● Behcet's Syndrome:

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

● Scurvy :

- u Vitamin C deficiency
- u Bleeding gums
- u Can cause teeth loss (permanent complication)

● Scurvy(cont'd)

- u Easy bruising
- u Diagnosis : Low ascorbic acid (Vit-C) level in Leukocyte

● Pellagra:

- u Nicotinic acid deficiency
- u 4 “D”s

Dermatitis (Photodermatitis)

Diarrhea

Dementia

Death (if not treated)

● Causes of generalized pruritus without skin lesions:

1. Endocrine: DM, hypo& hyperthyroidism
2. Haematological: polycythemia rubra vera, iron def anemia
3. Malignancy; e.g. Lymphoma
4. Hepatic: primary biliary cirrhosis
5. Renal: CRF

The commonest manifestation of CRF is pruritus

6. Neurological : e.g. Tabes dorsalis
7. Others:
 - ✓ Psychogenic
 - ✓ Drugs
 - ✓ Idiopathic

Nails

● Clubbing :

- ✓ Exaggeration of the normal nail curve associated with loss of the normal angle between nail and posterior nail fold
- ✓ Causes:
 1. Thoracic: Lung abscess, Lung CA
 2. CVS: Congenital cyanotic heart disease
 3. GIT:GI carcinoma, Inflammatory bowel disease
 4. Endocrine: Thyroid disease
 5. Idiopathic:

Splinter Haemorrhages :

u Causes :

1. Bacterial endocarditis
2. Septic emboli
3. CTD
4. Trauma
5. Idiopathic

Koilonychia :

- u Spoon- shaped appearance

- u Causes:

1. Iron deficiency anemia
2. Thyroid disease
3. Physiological; early childhood
4. Dermatoses: Lichen planus

Alopecia Areata and others

Purpura and Vasculitis

Objectives

- To differentiate between different types of purpura.
- To know the difference between inflammatory and non-inflammatory purpura.
- To have an approach to diagnose purpuric lesions.
- To be familiar with serious and non-serious conditions and when to refer to a specialist.

Purpura

- Reddish-purplish skin lesions from extravasation of RBCs into the skin.
- Non palpable purpura: classified according to size to Petechiae less than 2 mm, and Ecchymosis or bruises for larger lesions.
- Palpable purpura is the inflammatory type of different sizes, the main sign of vasculitis.

Abbreviations

ESR - erythrocyte sedimentation rate

CRP - C reactive protein

ANCA - antineutrophil cytoplasmic antibody

p-ANCA - perinuclear ANCA

ELISA - enzyme linked immunosorbent assay

AAV - ANCA associated vasculitides

IF - immunofluorescence

Purpura

Causes

```
graph TD; A[Causes] --> B[Platelet disease]; A --> C[Coagulation defect]; A --> D[Blood vessel wall pathology];
```

Platelet disease

Coagulation defect

Blood vessel wall pathology

Causes

1-Platelet Disorders

Thrombocytopenia

Platelet Dysfunction

2-Coagulation Factor Deficiency

Congenital

Factor VIII Deficiency

Factor IX Deficiency

Von Willebrands disease

Acquired

*Disseminated Intravascular
Coagulopathy*

Liver disease

Uremia

Vitamin K deficiency

3-Vascular Factors

Congenital

Hereditary Hemorrhagic

Telangiectasia

Ehlers-Danlos Syndrome

(Type IV)

Acquired:

Inflammation(Vasculitis)

Trauma

Vitamin c deficiency (scurvy)

Definition

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

Aorta and the great vessels (subclavian, carotid)

Claudication, blindness, stroke

-Medium-vessel vasculitis

Arteries with muscular wall

*Mononeuritis multiplex (wrist/foot drop), mesenteric ischemia,
cutaneous ulcers*

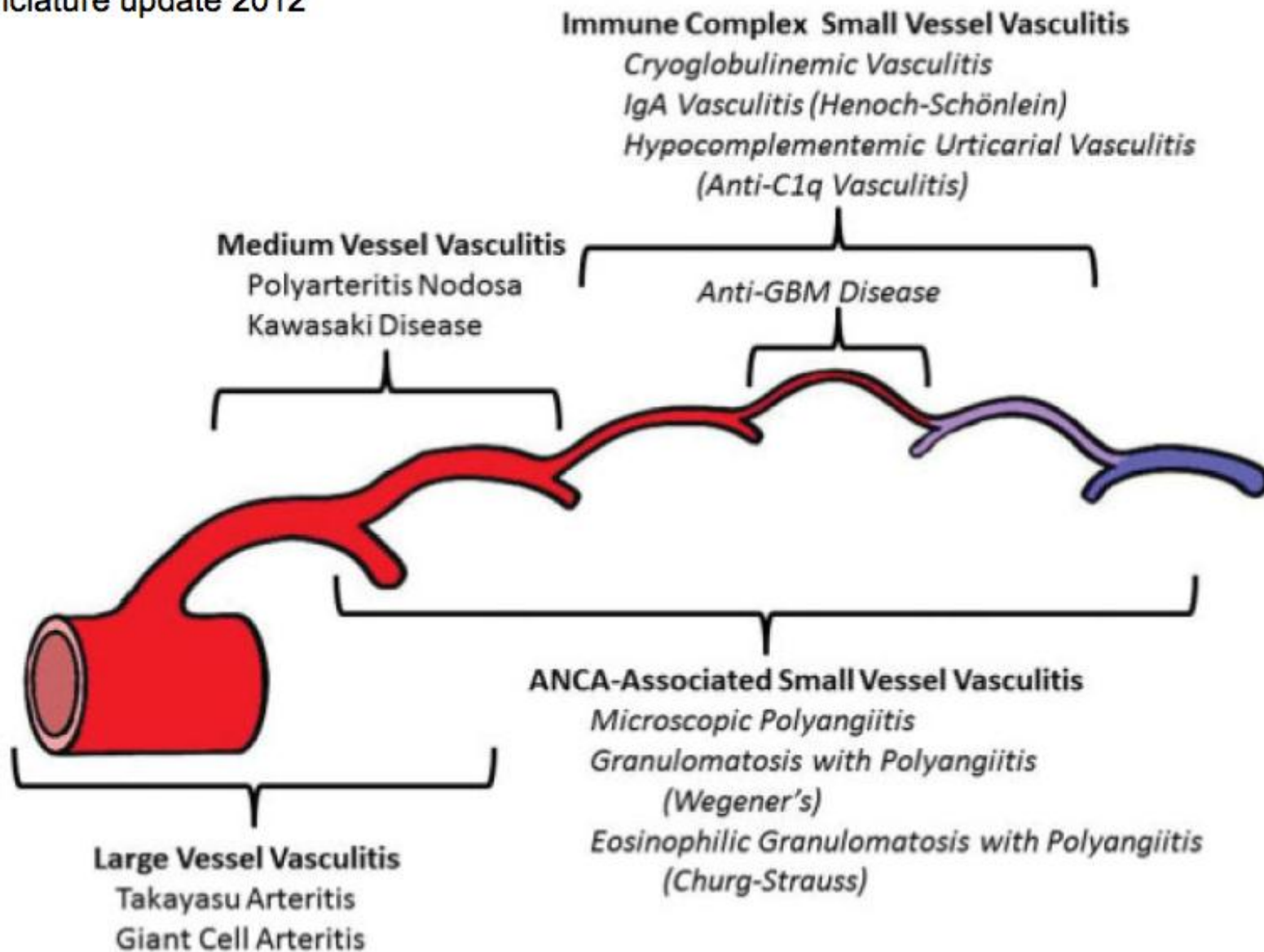
-Small-vessel vasculitis

Capillaries, arterioles, venules

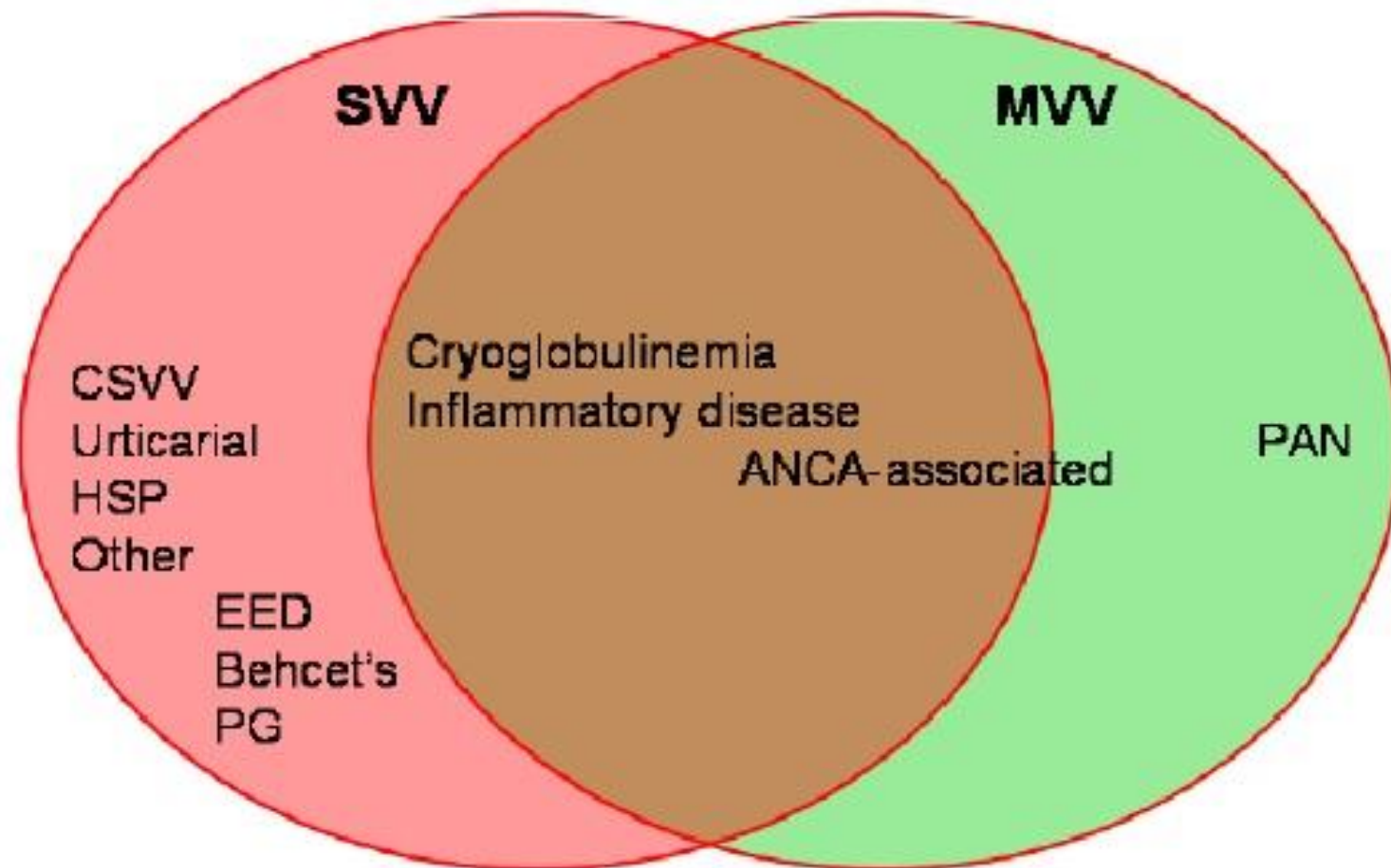
Palpable purpura, glomerulonephritis, pulmonary hemorrhage

Classification of Vasculitis

Chapel Hill Consensus Criteria
Nomenclature update 2012



Primary cutaneous vasculitis



Classification of Primary Cutaneous Vasculitis based on diseased vessel size

Abbreviations in legend:

SVV=small vessel vasculitis

MVV=medium vessel vasculitis

CSVV=cutaneous small vessel vasculitis

HSP=Henoch-Schonlein Purpura

EED=erythema elevatum diutinum

PG=pyoderma gangrenosum

PAN=polyarteritis nodosa

ANCA=anti-neutrophil cytoplasmic antibody

Cutaneous small-vessel vasculitis (CSVV)

- Also called Leukocytoclastic vasculitis.
- characterized by neutrophilic infiltration into the peripheral small dermal blood vessels.
- Purpura, urticaria, erythema-multiforme-like erythema, papules, nodules, pustules, blistering, erosion and ulceration occur, mainly in the lower extremities.
- An immune complex of an antigen (e.g., bacterium, virus, drug) and the antibody against that antigen deposit on the arteriolo-venular walls. These activate the immune system and cause vasculitis (type III allergic reaction). Penicillin, sulfa drugs, thiazides, NSAIDs and other drugs, chemical substances, hemolytic streptococcus bacteria, or viruses may be the foreign antigen. Collagen diseases and antibodies against malignant tumors can also be causes.

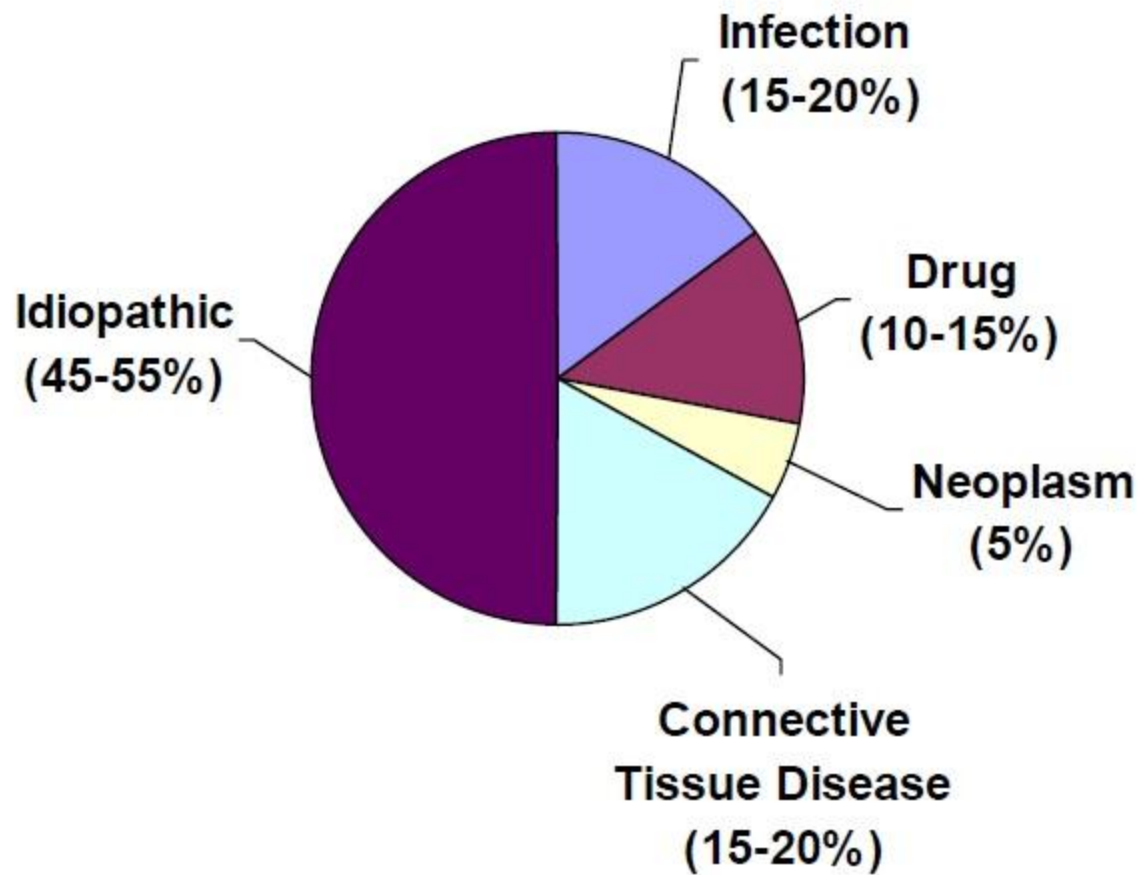
Cutaneous small vessel vasculitis

Pathogenesis:

- Many forms of small-vessel vasculitis are felt to be caused by circulating immune complexes*
- These lodge in vessel walls and activate compliment*

Cutaneous small vessel vasculitis

Causes of cutaneous vasculitis	
Infections	
Bacterial	<ul style="list-style-type: none">• Streptococcal, meningococcal, urinary tract infections
Viral	<ul style="list-style-type: none">• Hepatitis B and C, HIV
Mycobacterial	<ul style="list-style-type: none">• Tuberculosis
Connective tissue disorders	<ul style="list-style-type: none">• SLE and related conditions• Rheumatoid arthritis• Systemic sclerosis, Sjogren syndrome• Dermatomyositis• Medium vessel vasculitides (Wegener granulomatosis, polyarteritis nodosa, Churg-Strauss syndrome)
Malignancy	<ul style="list-style-type: none">• Haematologic<ul style="list-style-type: none">– myeloproliferative– lymphoma– monoclonal gammopathy– multiple myeloma
Drugs	Including antibiotics, antihypertensives
Idiopathic	Henoch-Schonlein purpura

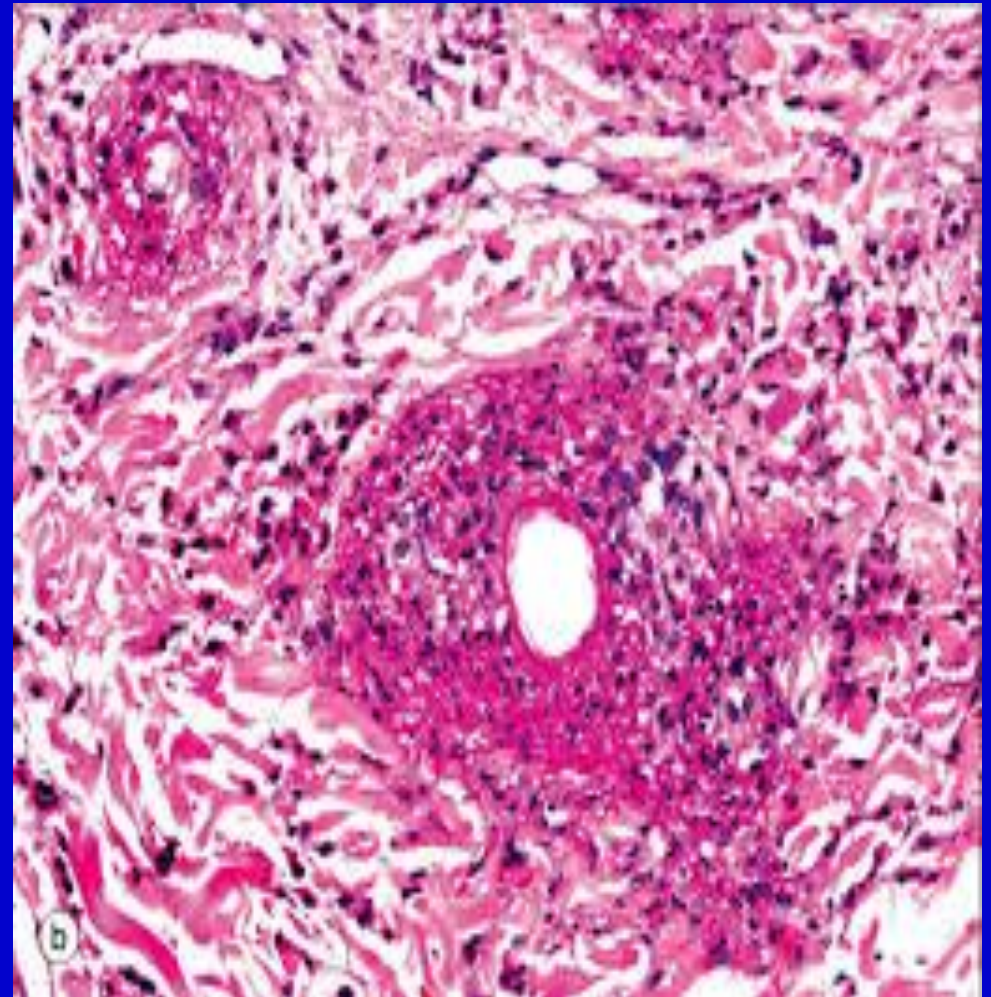


8 Etiologies of Cutaneous Vasculitis

Pathology

In the upper and middle dermal layer, fragments of nuclear debris and leakage of erythrocytes are found in the peripheral arteriola.

Neutrophilic infiltration occurs in the arteriolo-venous small blood vessels and capillaries. Thickening of the blood vessel walls and fibrinoid necrosis.



Investigations

- CBC, urea, creatinine.
- ESR (usually raised)
- Complements (decreased)
- Urinalysis (for protein and hematuria if kidneys involved)
- Occult blood in stool
- ANA, ANCA
- Cryoglobulins , hepatitis B, C, HIV.
- CRP, ASO titer and throat swab culture for streptococcal infection
- Skin biopsy

Investigations in a patient with cutaneous vasculitis

Vasculitis screen

All patients

- Full blood count and film
- Urea, electrolytes and creatinine (UE&C)
- Liver function tests (LFTS)
- Anti nuclear antibodies (ANA)
- Extractable nuclear antibodies (ENA)
- Antineutrophil cytoplasmic antibodies (ANCA)
- Rheumatoid factor (RF)
- Urinalysis
- Urine – phase contrast microscopy (glomerular red cells)
- Skin biopsy – histology and consider direct immunofluorescence

Selected patients where appropriate

- Hepatitis B and C serology
- Serum protein electrophoresis
- Streptococcal serology – antistreptolysin O titre – anti-DNAse antibodies
- Throat swab – microscopy and culture
- Blood cultures
- Midstream urine – microscopy and culture
- Skin biopsy – gram stain, culture, special stains and cultures for other organisms (eg. fungal)
- HIV serology
- Other serology
 - cryoglobulins, serum complement levels
 - lupus anticoagulant anticardiolipin antibodies
 - faecal occult blood test (for Henoch-Schonlein purpura)
 - chest X-ray, Quantiferon gold, Mantoux test

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*

Henoch-Schönlein purpura (HSP)

- In children, the onset is mostly after upper respiratory infection; association with hemolytic streptococcus has been pointed out.
- Drugs (penicillin, aspirin)
- These antigens combine with antibodies (mainly IgA) in the body, and the immunocomplex deposits on the vascular walls. Immunoreaction is induced to cause vasculitis and purpura.
- Pathology shows leukocytoclastic vasculitis with IgA deposition is observed by direct immunofluorescence.
- The histology of the kidney in HSP patients often shows crescentic glomerulonephritis.

Henoch-Schönlein purpura (HSP)

- Treatment by bed rest, pain relief and antibiotics if strep throat infection is present.
- Systemic steroids for severe cases with systemic involvement.
- HSP generally has a good prognosis and resolves within several weeks in most cases; however, it may recur.
- Serious complications may occur in other organs, such as nephritis with IgA deposition in the mesangium area, enterorrhagia, intussusception, intestinal perforation, or cerebral hemorrhage.
- Adults may have prolonged kidney involvement.

- 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*
- IgA, C3 and fibrin depositions have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques*

Urticarial vasculitis

- Presents with urticarial weals that lasts more than 24 hours unlike urticaria. And usually leave hyperpigmentation after resolution.
- Could be associated with fever, arthralgia, abdominal pain and angioedema especially in hypocomplementaemic urticarial vasculitis that is associated with SLE.
- Causes include connective tissue diseases, viral and bacterial infections, drugs like ACE inhibitors, penicillin, sulfonamides, fluoxetine and thiazides. Also leukemia could cause it.
- Most cases are idiopathic and improve spontaneously after few months.
- Treatment is to remove the offending agents if present, treat the underlying diseases. Symptomatic therapy with antihistamines and NSAIDs.
- Dapsone, colchicine, hydroxychloroquine, steroids, azathioprine, and cyclosporine.

Cutaneous polyarteritis nodosa

- a rare form of vasculitis which involves small and medium sized arteries of dermis and subcutaneous tissue.
- tender subcutaneous nodules and livedo reticularis that may ulcerate on legs and feet.
- Pathogenesis is unknown.
- Systemic involvement is uncommon except for peripheral neuropathy, and progression to classical polyarteritis nodosa is an exception.
- may have neuromuscular involvement in the form of peripheral neuropathy that presents with tingling, numbness, sensory disturbances, weakness, and absent reflexes.

Cutaneous polyarteritis nodosa

- In cutaneous PAN, histopathological examination shows features of nodular arteritis with polymorphonuclear infiltrates involving medium sized arteries in deep reticular dermis. There is extensive fibrinoid necrosis. This is in contrast to classical PAN which rarely shows nodular arteritis and the picture is of small vessel leukocytoclastic.
- Cutaneous PAN runs a chronic course lasting months to years, and has a waxing and waning phenomenon. Patients are generally treated with non-steroidal anti-inflammatory drugs and oral steroids. Immunosuppressive drugs can also be used in low doses in more severe kinds of cutaneous PAN and as steroid-sparing drugs.

pigmented purpuric dermatoses

- a group of chronic diseases of mostly unknown etiology characterized by extravasation of erythrocytes in the skin with marked hemosiderin deposition.
- Mostly affect males.
- Venous hypertension, exercise, and gravitational dependency are important cofactors that appear to influence disease presentation.
- Histologically, a perivascular T-cell lymphocytic infiltrate is centered on the superficial small blood vessels of the skin, which show signs of endothelial cell swelling and narrowing of the lumen. Extravasation of red blood cells with marked hemosiderin deposition in macrophages is also found but no vasculitis.
- The lesions are chronic and persist for years. With time, many of the lesions tend to extend and may become darker brown in color.
- No effective treatment available. It is a cosmetic problem mostly.