Review

ARTICLE

DERMATOLOGICAL MANIFESTATIONS OF VENOUS DISEASE: PART I

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ABSTRACT

Skin changes are one of the earliest signs of venous hypertension. Some of these changes such as venous eczema are common and easily identified whereas other changes such as acroangiodrmatitis are less common and more difficult to diagnose. Other vein related and vascular disorders can also present with specific skin signs. Correct identification of these skin changes can aid in making the right diagnosis and an appropriate plan of management. Given the significant overlap between phlebology and dermatology, it is essential for phlebologists to be familiar with skin manifestations of venous disease. This paper is the first installment in a series of 3 and discusses the dermatological manifestations of venous insufficiency as well as other forms of vascular ectasias that may present in a similar fashion to venous incompetence.

Introduction

Patients with venous disease often exhibit dermatological changes. Sometimes these skin changes are the only clue to an appropriate list of differential diagnoses. Venous insufficiency is the most common venous disease which presents with a range of skin changes. Most people are familiar with venous eczema, lipodermatosclerosis and venous ulcers as manifestations of long-term venous insufficiency. The less familiar are changes such as acroangiodermatitis and pigmented purpuric dermatoses. Dilatation of venous structures is not always due to venous incompetence and could be primary or secondary to systemic disease, or environmental influences such as actinic damage and radiation. This paper will discuss the dermatological manifestations of venous insufficiency as well as other forms of vascular ectasias that may present in a similar fashion to venous incompetence. The second instalment will focus on dermatological manifestations of haematologic disease and in particular thrombo-inflammatory conditions. The third instalment will discuss vascular anomalies.

Skin Manifestations Of Venous Insufficiency

Common Manifestations

Common manifestations of venous insufficiency include oedema, corona phlebectasia paraplantaris (Figure 1), stasis dermatitis, pigmentory changes (hyper- and hypo-), atrophie blanche, lipodermatosclerosis, and skin ulceration. Less common manifestations include pigmented purpuric dermatoses, and acroangiodermatitis. Superficial thrombophlebitis (STP) can also occur in association with venous incompetence but will be discussed in the second instalment of this paper (Figure 2).

Venous hypertension is responsible for many manifestations of chronic venous insufficiency (CVI) including oedema, red cell extravasation, perivascular fibrin deposition, impaired arterial inflow, and other locally mediated disturbances. The local tissue sequelae of CVI are due to impaired clearance of cellular metabolites secondary to concurrent damage to the lymphatic system.

One of the earliest manifestations of venous hypertension is lower limb oedema. Oedema is defined as a clinically apparent increase in the interstitial fluid volume. Oedema that retains an indentation when pressed with a finger is called pitting oedema. Systemic causes of oedema include congestive cardiac failure, hepatic insufficiency, renal failure, pretibial oedema in myxoedema, hypoproteinemia, and other systemic disorders. Lymphoedema may be a sign of primary lymphatic outflow obstruction, or it may be secondary to the overproduction of lymph due to severe

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venous hypertension (the so-called phlebolymphoedema). Usually, lower leg oedema is symmetrical in patients with systemic disorders.

Oedema of venous insufficiency is quite often associated with skin changes. Venous eczema (Figure 3) is usually the earliest cutaneous manifestation of venous insufficiency, and it may be a precursor to hyperpigmentation and lipodermatosclerosis. The medial ankle is most frequently involved revealing erythematous, scaling, and sometimes exudative, weeping patches and plaques (Figure 4). Secondary bacterial infection with staphylococcus aureus can cause typical crusting. Pseudomonal colonisation is a common complication. In long-standing cases, lichenification may occur as a consequence of chronic scratching and rubbing.

The development of contact dermatitis is especially problematic in the treatment of patients with venous eczema. Topical treatments, including neomycin, bacitracin and some topical corticosteroids, have been reported to cause contact sensitization in patients with venous eczema.[1] Contact dermatitis should be considered when venous eczema becomes clinically worse or does not improve despite appropriate topical treatment. Patch testing will be indicated. Topical antibiotics such as neomycin and popular preparations such as Kenocomb should never be used on open venous ulcers.

Venous hypertension leads to red cell extravasation. Breakdown of red cells leads to haemosiderin deposition and cutaneous pigmentation (Figure 5). Hyperpigmentation secondary to red cell extravasation is difficult to treat and not responsive to hydroquinone or other standard bleaching agents used to treat hypermelanosis. Treatment of the underlying venous hypertension can help in lightening the pigmentation but in general once haemosiderin pigmentation is present, it is mostly irreversible. Post-inflammatory hypopigmentation may occur which can even result in depigmentation. This should be differentiated from atrophie blanche.

Atrophie blanche is a specific type of dermal scarring usually presenting as a small reticulated porcelain white patch. Dermoscopic examination of atrophie blanche demonstrates dilated capillary loops interspersed within the dermal scars. Atrophie blanche is not specific for venous hypertension and can be a manifestation of livedoid vasculopathy as discussed below. Atrophie blanche is caused by obstruction of dermal arterioles and infarction of the skin supplied by these vessels (Figures 6-8). This obstruction can be secondary to venous hypertension or thrombo-occlusive vasculitis involving dermal arterioles. Skin in this condition is avascular and prone to ulceration and necrosis. Vasculitis can also lead to identical white scars of atrophie blanche on the legs. Other systemic diseases, such as systemic lupus erythematosis (SLE), rheumatoid arthritis, and Klinefelter syndrome can result in skin ulcerations that heal with atrophie blanche-like lesions.[2]

Lipodermatosclerosis (LDS) is the induration of skin and the underlying fat in the lower legs (Figure 9). It usually extends from the ankles to the lower border of the calf muscle. Histologically, there is a septal panniculitis and sclerosis of the overlying skin.[2] A less-known form is the so called 'acute lipodermatosclerosis' which is primarily an acute panniculitis. Acute lipodermatosclerosis can be confused with acute cellulitis or even acute venous eczema. Eventual induration of skin and the underlying adipose tissue leads to the more familiar clinical picture of chronic lipodermatosclerosis. Progression of disease leads to the classic inverted champagne bottle appearance. The induration and lack of perfusion of the tissue makes it prone to ulceration.

Most chronic lower leg ulcers have a venous aetiology. [3] Most venous ulcers are caused by superficial venous incompetence. [4] Only a minority are caused by deep vein incompetence or obstruction. Non-healing ulcers of the medial ankle are most likely due to underlying venous hypertension. These areas are especially prone to venous hypertension because their drainage largely depends on the competence of the great saphenous system and its associated perforating veins. The incompetent Cockett perforators along the posterior arch tributary of the great saphenous vein (GSV) play a crucial role in the altered haemodynamics of this area.

Venous ulcers are usually larger but shallower than other ulcers, have a moist granulating base and an irregular border (Figure 10). Calcification is a common finding. The tissue surrounding these ulcers may exhibit signs of stasis dermatitis and other changes such as atrophie blanche and hyperpigmentation.

Non-healing, nodular areas of venous ulcers should be biopsied to exclude neoplastic change. The most common tumour is squamous cell carcinoma, however rarely basal

cell carcinomas may develop too. [3] Malignant melanoma may present as a lower leg ulcer. Other important differentials include arterial, neuropathic, traumatic, pyoderma gangrenosum, vasculitis, infection, and self-induced artefact. Biopsies should be taken from the edge of the ulcer and not from its necrotic base. Multiple biopsies may be required for the diagnosis. Care must be taken in providing haemostasis. It is usually not possible to close the biopsy site with suture as the tissue is friable. Steri-strips can be applied but compression should be used to prevent bleeding from the biopsy site.

Livedoid Vasculopathy

Atrophie blanche is not always a manifestation of CVI but can be a sign of livedoid vasculopathy (LV). This condition is characterized by ulceration associated with a segmental pattern of reticulate purpura and atrophie blanche affecting the lower limbs (Figure 11). It has been falsely referred to as livedoid vasculitis, although true vasculitis is not a feature of this condition. It has also been described as 'livedo reticularis with summer ulcerations'. LV is a thrombo-occlusive process. Histologically, it shows fibrin deposition within both the wall and the lumen of affected vessels but there is no evidence of a true vasculitis. [5]

LV has been associated with Raynaud's phenomenon and acrocyanosis. Patients with LV may have a history of recurrent leg ulcerations. Such patients may have thrombophilic abnormalities such as factor V Leiden mutation, protein C deficiency, hyperhomocysteinemia, abnormalities in fibrinolysis, and increased platelet activation. [6]

Pseudo-Kaposi's Sarcoma (Acroangiodermatitis)

Another unique feature of CVI is the development of violaceous plaques and nodules on the legs and dorsal aspects of the feet (Figures 12-13). These lesions frequently undergo painful ulceration and can be clinically indistinguishable from classic Kaposi's Sarcoma (KS). This clinical appearance has led this entity to be

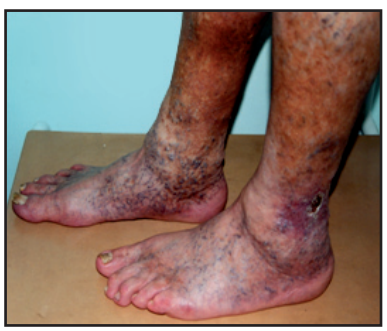

Figure 1: *Corona phlebectatica para-plantaris.*

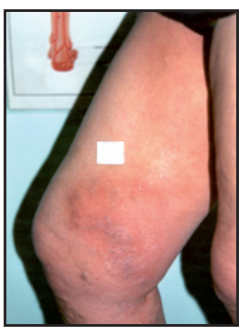

Figure 2: *Superficial Thrombophlebitis (STP) of a Great Saphenous vein tributary.*

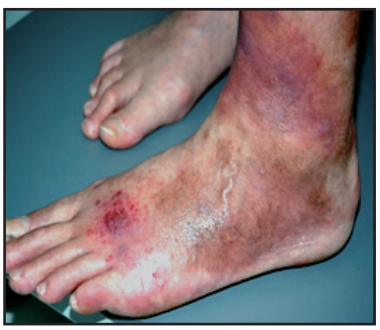

Figure 3: *Venous eczema over the dorsum of the foot. Venous hyperpigmentation over the lateral aspect of the foot.*

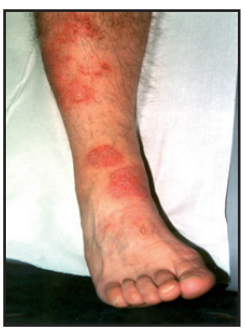

Figure 4: *Discoid eczema associated with venous hypertension.*

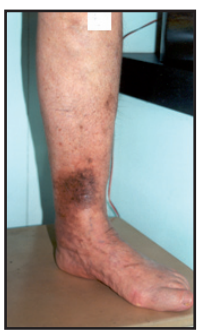

Figure 5: *Typical venous hyper-pigmentation of the medial gaiter area.*

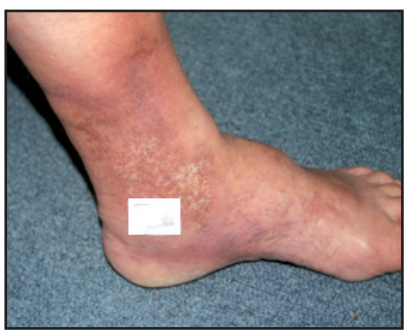

Figure 6: *Porcelain white scars of atrophie blanche in an area of lipoderma-tosclerosis.*

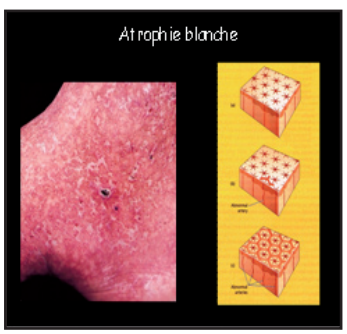

Figure 7: *Diagram demonstrating the arteriolar supply of the skin corresponding with cutaneous infarcts of atrophie blanche.*

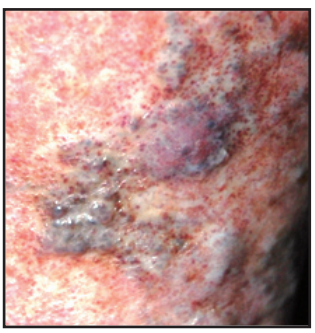

Figure 8: *Close-up image of atrophie blanche, interspersed punctate telangiectasias and underlying varicose veins.*

called pseudo–KS or acroangiodermatitis. Pseudo-KS is a hyperplasia of pre-existing vasculature. In contrast, KS is a vascular tumour associated with Human Herpes Virus type 8 (HHV 8). Vascular proliferation in KS is independent of the existing vessels.

Pseudo-KS is usually seen as a complication of severe CVI. Less commonly, congenital or acquired arteriovenous malformations, and arteriovenous fistulas can result in localised venous hypertension and the development of pseudo–KS. Pseudo-KS can also occur distal to the site of arterio-venous fistulas for haemodialysis. These lesions may resolve after closure, thrombosis or surgical elimination of the shunt. Pseudo-KS has also been described in association with hepatitis C and in some vascular malformations such as Klippel-Trenaunay syndrome (KTS). Bilateral lesions are usually associated with CVI, whereas unilateral lesions suggest an underlying arterio-venous malformations or a high flow lesion. Patients occasionally experience pruritus and pain. Ulceration and bleeding may also occur.

Pigmented Purpuric Dermatoses

Pigmented purpuric dermatoses (PPD) are a group of conditions characterized by extravasation of red cells and marked haemosiderin pigmentation of the skin. This condition has been wrongly called 'capillaritis', although there is no evidence of a true vasculitis. The hallmark of PPD is its characteristic orange-brown, speckled, cayenne pepper–like discoloration. The aetiology of PPD is currently under investigation. Venous hypertension, contact dermatitis, and certain drugs have been implicated in the pathogenesis of PPD. Other associations are hypersensitivity to food dyes and preservatives such as tartrazine, and clothing dye dermatitis (especially khaki dyes). Drugs such as aspirin, glipizide, thiamine and interferon-alfa have been implicated. [7-10] The author's recent research has discovered coagulation abnormalities and in particular platelet function abnormalities in this group of patients (in press). Differential diagnoses include angiocentric mycosis fungoides, venous hyperpigmentation, leukocytoclastic vasculitis, petechial

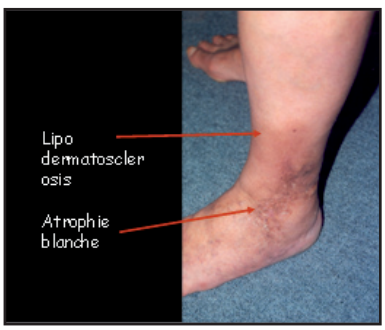

Figure 9: *Lipodermatosclerosis associated with atrophie blanche.*

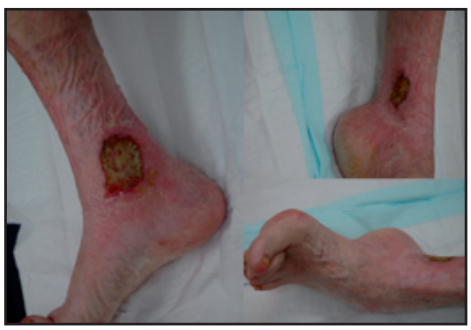

Figure 10: *Advanced venous ulcers causing fixed foot deformity. This patient received UGS and all ulcers healed. However, she eventually required a left below knee amputation to facilitate mobility.*

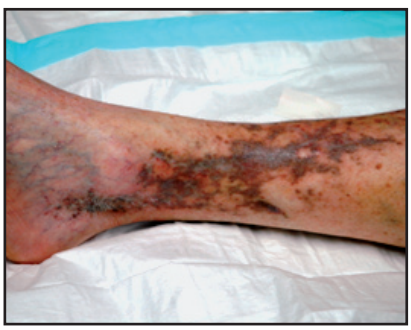

Figure 11: *Livedoid vasculopathy secondary to protein S deficiency.*

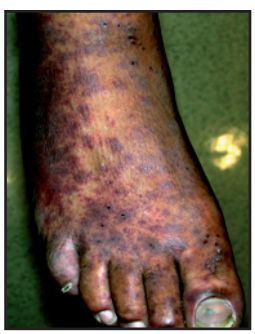

Figure 12: *Acroangio-dermatitis secondary to venous hypertension can resemble Kaposi's sarcoma.*

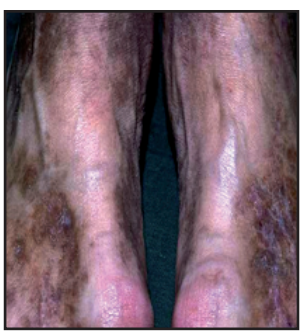

Figure 13: *Hyperpigmentation secondary to acroangiodermatitis can resemble venous hyperpigmentation.*

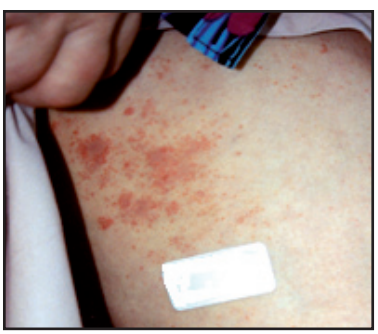

Figure 14: *Schamberg's Disease (taken from Schamberg J.F, A peculiar progressive pigmentary disease of the skin, British Journal of Dermatology, 1901, 13:1-5).*

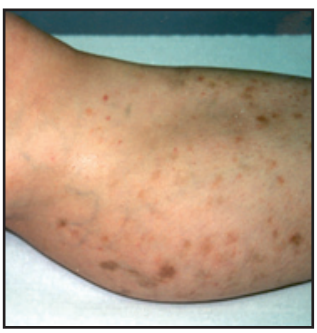

Figure 15: *Itchy purpura.*

haemorrhage and scurvy.

A number of clinical patterns are recognized all demonstrating a similar histologic appearance. These include Schamberg's disease (or progressive pigmented purpuric dermatosis) (Figure 14), itchy purpura of Lowenthal (Figure 15), lichen aureus (Figures 16-17), purpura annularis telangiectodes (Majocchi disease) (Figure 18), pigmented purpuric lichenoid dermatosis of Gougerot and Blum (Figures 19), unilateral PPD (Figures 20-21) and eczematoid-like purpura of Doucas and Kapetanakis. A granulomatous variant of PPD has also been reported.

These subclass all share common histopathologic features of red cell extravasation, haemosiderin deposition and a lymphocytic infiltrate. A lichenoid inflammatory infiltrate is present in the Gougerot-Blum sub-type. Spongiosis is present in the Doucas and Kapetankis sub-type. Older lesions tend to be less inflamed and extravasated red cells may no longer be present. In the author's experience, these sub-classifications are somewhat artificial and one patient may exhibit multiple sub-types. Lichen aureus is commonly found in association with venous disease and sometimes directly over an underlying incompetent perforator. Treatment of the underlying localised venous hypertension can help in the resolution of lichen aureus (Figures 16-17). Topical steroids are possibly helpful only if pruritus is present but otherwise have a limited role. The underlying condition and in particular coagulation abnormalities should be investigated and excluded. Patch testing, other forms of allergy testing and food elimination diets may be appropriate. Aspirin, vitamin E, fish oil and other forms of drugs and supplements interfering with platelet function should be stopped, if appropriate.

Cutaneous Vascular Anomalies

Cutaneous vascular anomalies may be classified into the following categories of tumours, malformations and ectasias. Tumours may be congenital or acquired, benign or malignant and hyperplastic or neoplastic. The

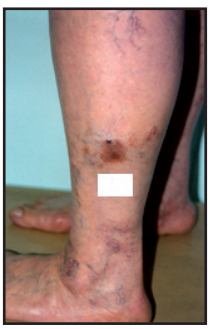

Figure 16a: *Lichen aureus before treatment.*

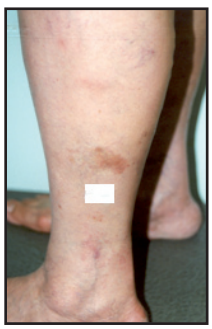

Figure 16b: *Lichen aureus after treatment.*

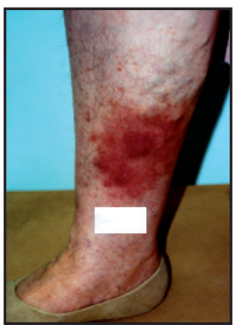

Figure 17a: *Lichen aureus before treatment.*

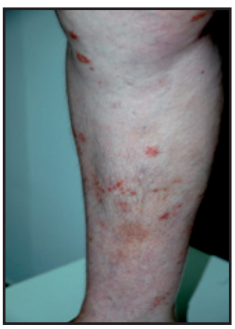

Figure 17b: *Lichen aureus after treatment.*

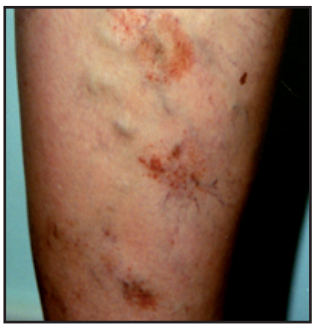

Figure 18: *Majocchi disease.*

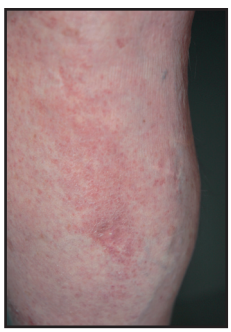

Figure 19: *Pigmented purpuric lichenoid dermatosis of Gougerot and Blum.*

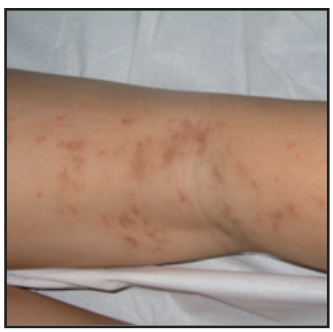

Figure 20: *Unilateral PPD.*

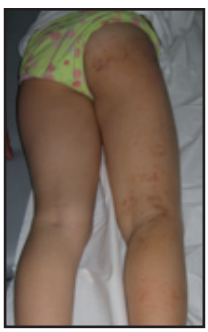

Figure 21: *Unilateral PPD.*

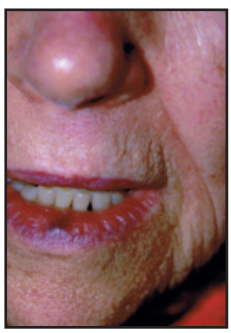

Figure 22: *Venous lake on the lower lip.*

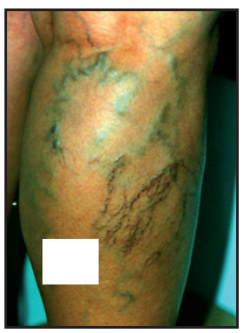

Figure 23: *Telangiectasias in association with underlying varicose veins.*

most common tumour of childhood is haemangioma. Haemangiomas are not present at birth and typically undergo involution. Malformations are errors of morphogenesis and are present at birth. They do not spontaneously involute. Vascular ectasias are dilatations of preexisting normal vessels. This paper will discuss vascular ectasias not necessarily associated with venous incompetence. The third installment of this paper will discuss vascular tumours and malformations.

Vascular Ectasias

Dilatation of blood vessels can be due to increased intravascular pressure or loss of elasticity of the vessel wall. Varicose veins are dilated vessels demonstrating increased muscularity and loss of elastic fiber. Vascular ectasia could also be due to actinic damage resulting in elastolysis of the vessel wall.

Venous Lake

Venous lakes are probably the most common vascular ectasias. These are dark blue phlebectasias that tend to occur on sun-exposed skin, especially on the lower lips or ears of elderly patients (Figure 22). Chronic actinic damage injures the vascular adventitia and the dermal elastic tissue, causing dilatation of normal venous structures. Treatment with sclerotherapy for larger lesions and vascular laser therapy for smaller lesions is usually successful. Malignant melanoma is a differential diagnosis and should be excluded. Venous lakes, as against melanomas, are compressible and empty with compression. Venous lakes are benign lesions but can bleed or thrombose causing pain.

Angiokeratomas

Angiokeratomas are a group of vascular ectasias that involve the papillary dermis. Angiokeratomas are not true angiomas but rather ectasias of pre-existing vessels. Overall, altered haemodynamics typically caused by trauma appear to produce telangiectatic vessels of the papillary dermis with an overlying reactive hyperkeratosis to the epidermis. These lesions may also produce papillomatosis, and acanthosis of the epidermis. [11]

Angiokeratoma of Fordyce. These are typically asymptomatic, 2-5 mm, blue-red papules with a scaly surface located on the scrotum, shaft of penis, labia majora, inner thigh, or lower abdomen. They are dilated ectatic capillaries in the superficial dermis with overlying epidermal hyperplasia. These lesions bleed easily. Angiokeratomas occur in the presence of varicoceles or vulvar varices and venous hypertension has been implicated in its pathogenesis. [12] If treatment is needed, then locally destructive methods have been used. Light hyfrecation may not provide adequate haemostasis and bleeding may occur. Vascular laser photocoagulation usually provides adequate treatment.

Angiokeratoma Circumscriptum (AC). This is a rare type of angiokeratoma with a unilateral distribution of discrete papules and nodules localized to a small area of the leg or trunk. Due to its unilateral and truncal distribution, AC has also been called angiokeratoma corporis neviform. AC has been classified as a capillary malformation. It has been reported to coexist with angiokeratoma of Fordyce and caviar spots (angiokeratomas of the tongue). It can be found in association with other vascular malformations and haemangiomas. Recurrent bleeding can occur. Women are affected more commonly than men, in a ratio of approximately 3 to 1. [13] These lesions respond well to vascular laser treatment.

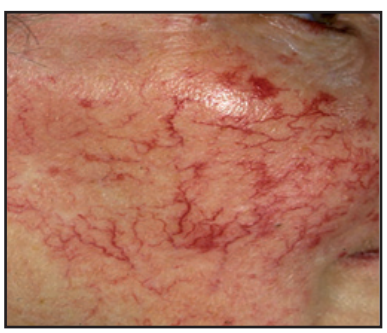

Figure 24: *Facial telangiectasias.*

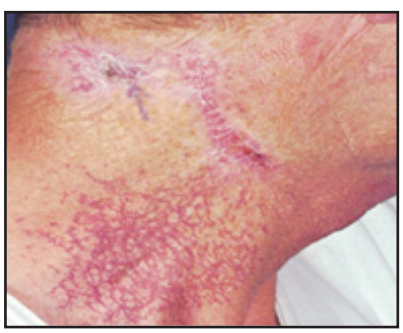

Figure 25: *Post-radiation telangiectasias.*

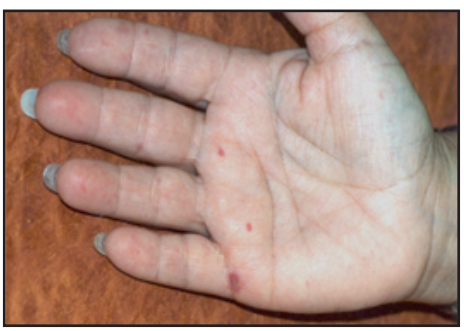

Figure 26: *Mat telangiectasia of scleroderma.*

Angiokeratoma Corporis Diffusum (Fabry's disease). This is an X-linked disorder caused by a deficiency of the lysosomal enzyme alpha-galactosidase. This inborn error of metabolism results in deposition of neutral glycosphingolipids in the lysosomes of most cells. The most affected cell is the vascular endothelium. [14] Angiokeratomas form and tend to concentrate between the umbilicus and the knees but usually spare the head. Lymphoedema and varicose veins are present. Renal failure, cardiac failure and cerebrovascular disease are the major causes of death. Heterozygous females may develop angiokeratomas but experience a milder clinical course.

Telangiectasias

Telangiectasias are dilated venules, arterioles or arteriovenous malformations visible on the skin or mucosal surfaces. Telangiectasias that develop in the absence of any preceding or coexisting cutaneous or systemic disease are considered to be primary or essential. Telangiectasias resulting from or in association with a known disease are classified as secondary

Leg telangiectasias (spider veins). These vessels are dilated post-capillary venules demonstrating backflow from incompetent reticular (feeder) veins or directly from incompetent tributary or even truncal veins (Figure 23). Doppler examination may demonstrate reflux in even very small telangiectatic vessels. Telangiectasias associated with venous incompetence are usually dark red. Larger bluish vessels are termed venulectasias. Not all leg telangiectasias are secondary to venous incompetence and essential telangiectasias are commonly found near the feet and ankles. Gold standard for treatment of leg telangiectasias is sclerotherapy. The underlying venous incompetence must be treated first, before any attempt is made at sclerosing the telangiectasias. Vascular laser therapy is unlikely to be effective if underlying venous incompetence is present.

Facial telangiectasias. Telangiectasias seen on the face and trunk are usually due to actinic damage. Facial telangiectasias may also be associated with flushing and vascular instability as seen in rosacea or carcinoid syndrome (Figure 24). Best treatment for these lesions is vascular laser therapy. Peri-alar telangiectasias usually demonstrate higher flow volumes and are more difficult to treat. They are more common in people suffering with sinus disease and chronic nasal congestion. This can lead to recurrent peri-alar telangiectasias despite adequate treatment. Vascular laser therapy may need to be repeated a number of times for larger peri-alar telangiectasias or sclerotherapy can be attempted.

Post-radiation telangiectasias. Radiotherapy can lead to appearance of telangiectasias up to 20 years after the procedure. The radiation damage causes destruction of elastic fibers within vessels and leads to formation of the telangiectasias (Figure 25). Malignancy and in particular squamous cell carcinoma can develop in these areas.

Mat telangiectasias. These are patches of telangiectasias seen in patients with scleroderma and CREST syndrome (Figure 26). They present as flat and non-pulsatile vessels appearing as discrete mats. This should not be confused with telangiectatic matting which is a complication of surgery and sclerotherapy.

Telangiectatic matting. This is a form of neovascularisation which occurs as a complication of sclerotherapy or surgery (Figure 27). Patches of very small telangiectasias appear over the sites of recanalized vessels. Inflammation seems to play a role in its pathogenesis. Inflammatory cytokines with angiogenic properties probably play an important role

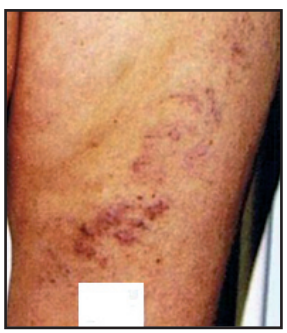

Figure 27a: *Telangiectatic matting – pre-sclerotherapy.*

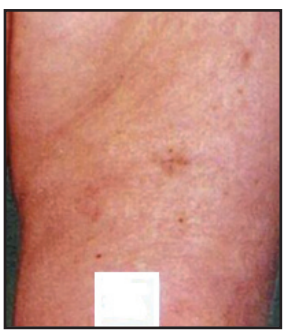

Figure 27b: *Telangiectatic matting – post-sclerotherapy.*

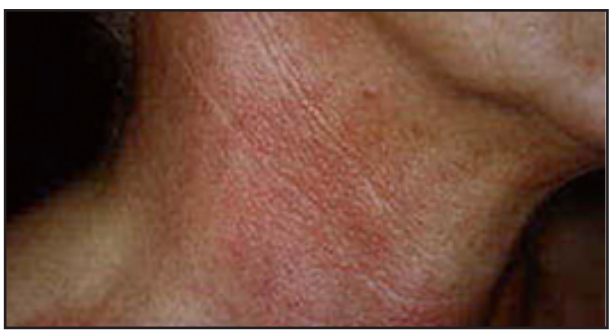

Figure 28: *Poikiloderma of Civatte.*

in the pathogenesis of this condition. Other associations include concurrent use of female hormonal supplements such as oral contraceptives and hormone replacement therapy. There are usually underlying small incompetence veins in direct association with these vessels.

Poikiloderma of Civatte. Poikiloderma of Civatte refers to a reticulate erythematous pigmentation seen on the sides of the neck (Figure 28). Areas shaded by the chin are spared. The term poikiloderma refers to the combination of atrophy, telangiectasias, and pigmentary changes (both hypo- and hyper-pigmentation). It occurs most commonly in fair-skinned middle-aged or elderly females. Chronic exposure to ultraviolet light appears to be a primary aetiologic factor but photosensitizing chemicals in perfumes or cosmetics have also been implicated. [15]

Periungual telangiectasias. These are telangiectasias along the proximal nailfold. They are visualized dermoscopically as dilated capillary loops. These are pathognomonic signs of connective tissue diseases, such as lupus, scleroderma, and dermatomyositis.[16]

Generalised Essential Telangiectasia (GET). This refers to the syndrome of widespread primary telangiectasias of unknown aetiology. There is no reported association with venous disease or actinic damage. Familial cases have been reported with an autosomal dominant pattern of inheritance.[16, 17] It is termed 'generalised' because of the widespread anatomic distribution of the telangiectasias. However, GET usually starts as light pink linear or punctuate telangiectasias on the feet, ankles, and distal legs and may gradually spread (Figures 29-30). When the lesions become confluent, the skin appears diffusely erythematous. Involvement of the oral mucosa and conjunctiva has been reported. GET is usually asymptomatic, but tingling, burning or numbness is occasionally reported.[18] The age of onset is usually in the fourth or fifth decade, but it may be observed in younger adults

Hereditary Benign Telangiectasias. This condition resembles GET. It commences in early childhood, and is usually present on sun exposed skin.

Hereditary Haemorrhagic Telangiectasia (HHT- Osler-Rendu-Weber disease). These lesions are arteriovenous malformations (AVM) of the microvasculature. They usually appear as punctuate dark red elevated lesions on mucous membranes, face and distal limbs. Lesions may involve the tongue, retina, lung and the brain (Figure 31). HHT lesions appear during the third or fourth decade of life. Recurrent epistaxis is usually the presenting symptom. Coagulation factor deficiency such as Von Willebrand's disease may be present. Family screening may be indicated.

Unilateral Naevoid Telangiectasia (UNT). In this condition, patches of telangiectasia present in a unilateral linear distribution. The segmental pattern suggests a somatic mosaicism. Pregnancy, chronic liver disease and other states of oestrogen excess are implicated in its pathogenesis. The third and fourth cervical dermatomes are the most common sites, but the thoracic dermatomes and scattered distant sites may also be involved.[2]

Telangiectasia Macularis Eruptiva Perstans (TMEP). This is the telangiectatic form of cutaneous mastocytosis. The lesions appear as brown telangiectatic macules that may be generalized in distribution. When a lesion of other forms of mastocytosis such as urticaria pigmentosa or mastocytoma is stroked, it typically urticates, becoming pruritic, oedematous, and erythematous. This change is referred to as the Darier sign, which can be explained on the basis of mast cell degranulation induced by physical

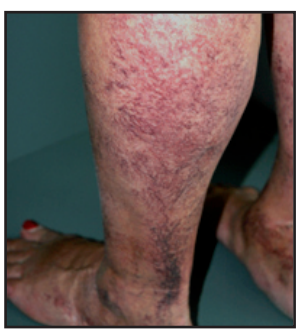

Figure 29: *Generalised Essential Telangiectasias (GET).*

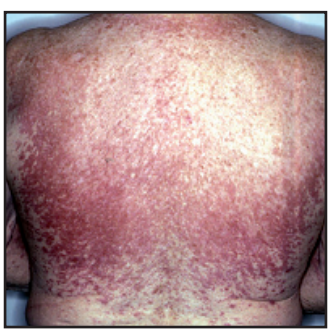

Figure 30: *Generalised Essential Telangiectasias (GET).*

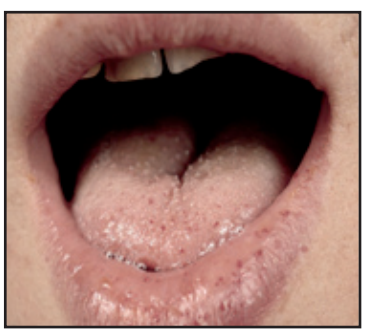

Figure 31: *Hereditary Haemmorhagic Telangiectasias (HHT).*

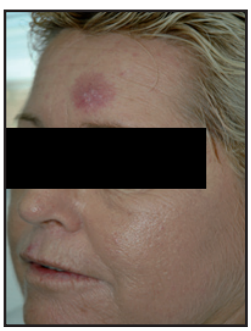

Figure 32: *Spider naevus of the forehead.*

stimulation. The Darier sign is usually negative in TMEP. This is because the lesions of TMEP are paucicellular and, therefore, mast cells may not be present in sufficient numbers for significant degranulation to occur.[19] Diagnosis of TMEP is made on skin biopsy. Anaesthetic should be injected without adrenalin adjacent to, and not directly into, the lesion to avoid mast cell degranulation, which would make histologic examination difficult.

Ataxia Telangiectasia (AT). This is an autosomal recessive immunodeficiency characterized by progressive neurologic impairment and cerebellar ataxia. Patients are susceptible to sinopulmonary infections, impaired organ maturation, and a predisposition to malignancy. Oculo-cutaneous telangiectasias have a later onset than ataxia, first noticed in childhood and sometimes not until adolescence. Ocular telangiectasias extend from the lateral canthus in the equatorial region of the conjunctivae towards the corneal limb. Telangiectasias may also involve the ears, eyelids, the cubital and popliteal fossae. The telangiectases are predominantly of venous origin and are not AV malformations.[20]

Spider Naevi. Spider naevi are dilated ascending arterioles. They usually exhibit radiating thin-walled vessels in association with a central arteriole (Figure 32). These lesions are also referred to as spider angiomas and naevus araneus. Spider naevi are not vascular proliferations but occur as a result of the dilation of preexisting vessels. They demonstrate arterial flow on Doppler examination. Compression of the central arteriole produces blanching. When released, the lesion quickly refills from the central arteriole. Spider naevi are acquired lesions present in 10-15% of healthy adults and young children. Lesions are found most commonly on the face, neck, chest and arms. In young children, spider naevi are common on the backs of the hands and fingers. Hormonal changes play a role in pathogenesis of these lesions. Many women develop lesions during pregnancy or while taking oral contraceptives. The lesions usually resolve spontaneously 6 to 9 months after delivery or after discontinuing oral contraceptives. Numerous lesions are seen in patients with significant hepatic disease such as advanced hepatitis C.

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