Chapter 7
Telangiectasias

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Definition

Mucocutaneous telangiectasias are dilatation of the capillaries, arterioles, and/or venules of the dermis. They appear as erythematous or bluish macules (that can be confluent into patches) that are blanchable on diascopy. They can be linear, lacy, or matted. If the blood vessels involved are deep (and thus not as compressible from external forces), they can sometimes appear as non-blanching lesions and should still be considered in the differential diagnosis of purpura. They can occur almost anywhere on the surface of the skin and mucous membranes. Telangiectasias can be simply a cosmetic nuisance or they can be a sign of underlying disease. Their distribution, configuration, associated signs and symptoms, and the medical history of the patient are all important clues to being able to distinguish between isolated lesions and those which require consideration for underlying systemic pathology. It should be noted that telangiectasias can also be secondary findings in a number of dermatologic lesions (e.g., basal cell carcinoma), but those will not be discussed here.

Differential Diagnosis

Primary Telangiectasia Syndromes

Certain hereditary syndromes (e.g., genodermatoses) are associated with telangiectasias. These include ataxia telangiectasia, Bloom's syndrome, xeroderma pigmentosum, Sturge-Weber disease, Rothmund-Thompson syndrome, and Klippel-Trénaunay

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syndrome. The most common hereditary telangiectasia is actually a capillary malformation—the nevus flammeus. Also known as the port-wine stain, these lesions typically present as confluent patches (1–5 cm) on the posterior neck or occipital scalp. Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu) usually presents in the third or fourth decade with discrete, round telangiectatic macules on the face, lips, oral mucosa, trunk, arms, and fingers [1]. Palmar erythema can also be seen. The lesions are actually small arteriovenous shunts that can also occur in the liver, gastrointestinal tract, lungs, and brain. Clues to this diagnosis include a family history (with autosomal dominant pattern), mucous membrane lesions, recurrent epistaxis, and visceral involvement.

Generalized essential telangiectasia is one of the most common forms of idiopathic generalized telangiectasia [2]. In this disorder, lesions most commonly begin on the feet, ankles, and distal legs but are characterized by slow extension to other areas. The lesions can be associated with numbness or burning. Patients typically present in the fourth or fifth decade of life. Typically the lesions are bright red, and individual, discrete capillaries can be seen that often form lacy, syncytial networks. Less commonly the lesions can be "matted" (e.g., present as a nearly confluent macule). Histopathology demonstrates only dilatation of the superficial dermal vasculature with no inflammation or abnormality in the surrounding connective tissue.

Cutaneous collagenous vasculopathy is a rare disorder that can clinically be confused with generalized essential telangiectasia [3]. Histologically, however, the lesions are characterized by a thick perivascular wall of collagen surrounding the dermal capillaries.

Unilateral nevoid telangiectasia syndrome (UNTS) is a rare disorder (both congenital and acquired) that can occur at any age (peak onset in the third decade of life) characterized by focal telangiectasias in a dermatomal distribution most commonly in the distribution of the trigeminal, cervical, or upper thoracic nerves [4]. A possible etiologic relationship to estrogens has been debated, as it occurs frequently in pregnancy and with liver disease.

Secondary Telangiectasias

Venous hypertension can be a common cause of telangiectasias on the legs and feet. These lesions are secondary to high pressure in the venous system that is most commonly due to poor valve function, but any chronic impairment of venous return can result in these lesions. Because they are dilations in the venous system, the lesions are typically more bluish than the erythematous capillary dilations that are seen in other disorders. There will often be associated, palpable bluish venous varicosities and a history of leg edema.

Telangiectasia can be the natural result of response to many types of trauma, including ulceration. Chronic UV exposure can result in telangiectasias, as can radiation. These irradiation-driven telangiectasias can be identified as they are often associated with pigmentation abnormalities and/or epidermal atrophy and usually

are bound by the limits of the exposure (e.g., photodistributed or in the field of radiation), and this can be helpful in the diagnosis.

Telangiectasias can also be induced by certain medications. The most commonly associated oral medications include lithium, oral contraceptives, thiotixene, interferon alpha, and isotretinoin. These can occur anywhere on the body. Calcium channel blockers (namely, amlodipine and nifedipine) can induce telangiectasias with a photodistribution, as can rarely other medications such as bosentan or venlafaxine [5]. Chronic use of topical corticosteroids can result in telangiectasia—helpful associated findings include epidermal atrophy, striae, and/or acneiform lesions. Exposure to aluminum has been associated with the appearance of characteristic, asymptomatic, erythematous, matted, 0.2–3 cm telangiectatic lesions primarily on the upper chest and back [6]. Lesions are characterized histologically by elastic degeneration of the stroma and a mononuclear cell infiltrate.

Telangiectasias are seen in certain systemic or metabolic diseases. Liver disease or pregnancy can commonly be associated with cutaneous telangiectasias, due to the effects of high levels of estrogen. Palmar erythema is often seen, as are discrete, lacy, red telangiectasias, often found over the chest and back. Other metabolic abnormalities including thyroid disease can also result in telangiectasias.

Red fingers syndrome is characterized by painless erythema and telangiectasia of the dorsal tips of the fingers and toes and is classically associated with HIV, HBV, or HCV [7]. It is thought that the association with these viruses might be ultimately due to the liver dysfunction that ensues in these patients.

Certain neoplasms can result in the appearance of telangiectasias. Atrial myxomata have been reported in association with telangiectasias of the chest [8]. The most common malignancies that are associated with telangiectasias are carcinoid, intravascular (angiotropic) lymphoma, and mycosis fungoides—however, they have also been reported in association with ovarian, bronchogenic, and breast carcinoma.

Telangiectasias are associated with certain inflammatory diseases that are not necessarily autoimmune in nature. Cutaneous mastocytosis can present with telangiectatic macules, mostly commonly with the subtype known as telangiectasia macularis eruptiva perstans (TMEP). These lesions are usually hyperpigmented, pruritic, truncal, and often urticate (become raised) when rubbed. Acne rosacea is associated with telangiectasias of the face, especially the cheeks and nose. The presence of inflammatory papules or pustules helps the clinician make this diagnosis, although these are often absent (in the so-called erythrotelangiectatic form of acne rosacea), and can often be confused with the malar rash of acute cutaneous lupus erythematosus or dermatomyositis (see below).

Finally, telangiectasias can be a sign of underlying rheumatic disease. The erythema of dermatomyositis and cutaneous lupus erythematosus is often telangiectatic on close examination (Fig. 7.1). Violaceous color, photodistribution (e.g., face, upper back, chest, upper arms), and itch or burning (especially in the case of dermatomyositis) are all clues to an underlying autoimmune inflammatory disease. The inflammatory lesions of dermatomyositis and cutaneous lupus often resolve with lacy telangiectasia (Fig. 7.2). Dermatomyositis patients can also present with telangiectatic patches adjacent to white, avascular appearing macules that are often found in a

Fig. 7.1 Telangiectatic papules and plaques in a patient with subacute cutaneous erythematosus

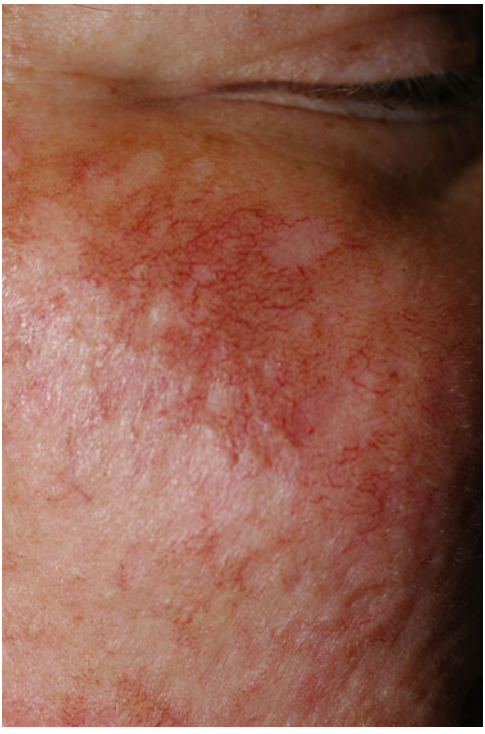


Fig. 7.2 Postinflammatory telangiectasia in a patient with dermatomyositis

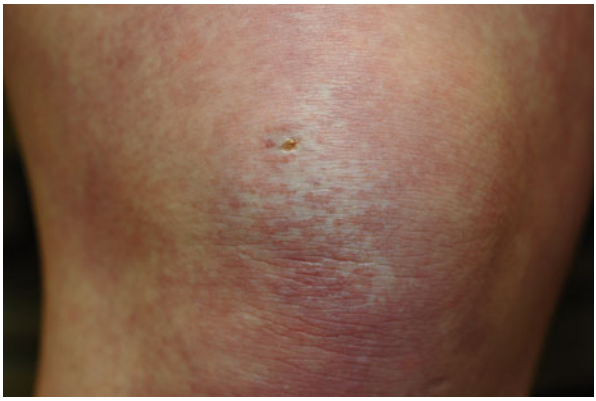


Fig. 7.3 Telangiectatic and avascular patches typical of dermatomyositis

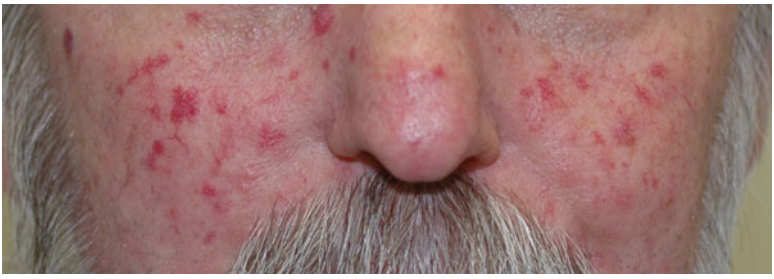


Fig. 7.4 Matted telangiectasias of systemic sclerosis

reticulate pattern (Fig. 7.3). In contrast, the telangiectasias seen in patients with systemic sclerosis are typically matted, discrete 1–5 mm lesions (Fig. 7.4). These typically occur on the hands and digits (palmar greater than dorsal surface) and the face and lips but can also be found on the chest and arms. Some data suggest that the presence of these lesions might correlate with the risk of developing pulmonary hypertension [9]. Patients with systemic sclerosis can also have discrete (non-matted) telangiectasias over the interphalangeal and metacarpophalangeal joints (Fig. 7.5), which are usually associated with prior digital ischemic ulcerations in these regions. Patients with SLE, dermatomyositis, and systemic sclerosis all can present with periungual capillary telangiectasias, which are often visible to the naked eye (Fig. 7.6). The periungual lesions of dermatomyositis are often associated with tenderness, while those of systemic sclerosis are associated with acrosclerosis or puffy fingers as well as Raynaud's. Periungual telangiectasias are considered to have high specificity for the presence (or future development) of connective tissue disease.

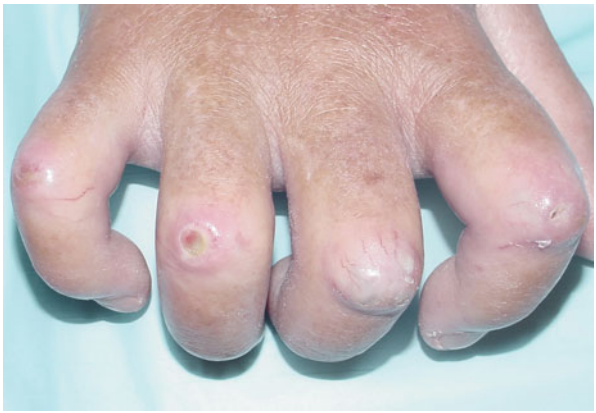


Fig. 7.5 Lacy telangiectasias overlying the proximal interphalangeal joints in a patient with systemic sclerosis. Note active digital ischemic lesion which is associated with this pattern of telangiectasia

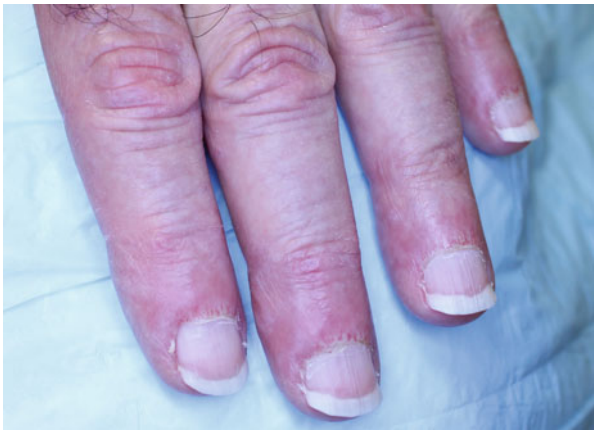
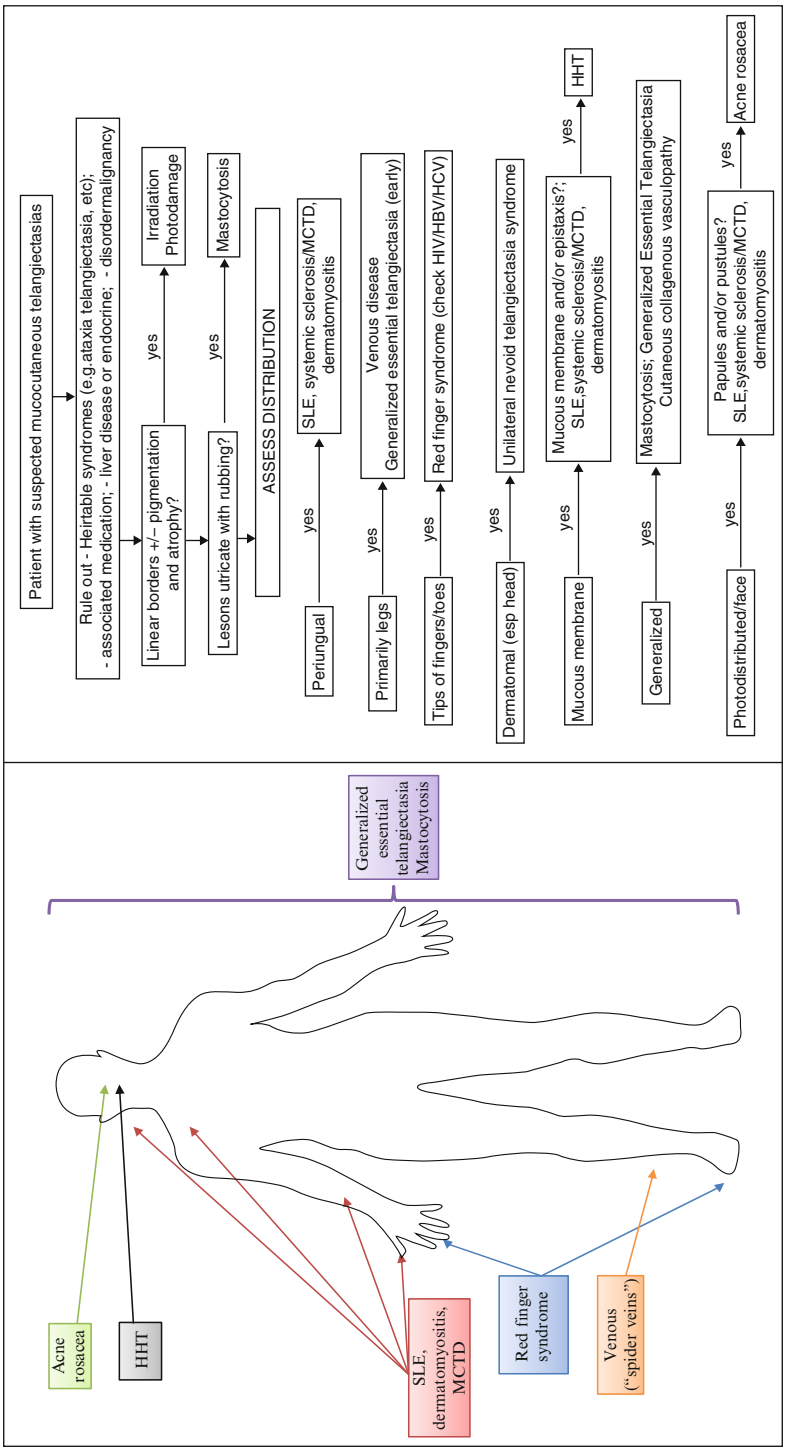


Fig. 7.6 Typical periungual telangiectasias in this patient with lupus erythematosus

Approach to the Patient

In summary, when presented with a patient with mucocutaneous telangiectasias, one should first rule out a primary, congenital syndrome as well as possible secondary causes such as medication or malignancy. The associated cause of the telangiectasias is then decided upon by both configuration and distribution of the lesions.


Patient with suspected mucocutaneous telangiectasias
Rule out - Heirtable syndromes (e.g.ataxia telangiectasia, etc);
- associated medication; - liver disease or endocrine; - disordermalignancy
Linear borders +/– pigmentation and atrophy?
yes
Irradiation
Photodamage
Lesons utricate with rubbing?
yes
Mastocytosis
ASSESS DISTRIBUTION
Periungual
yes
SLE, systemic sclerosis/MCTD, dermatomyositis
Primarily legs
yes
Venous disease
Generalized essential telangiectasia (early)
Tips of fingers/toes
yes
Red finger syndrome (check HIV/HBV/HCV)
Dermatomal (esp head)
yes
Unilateral nevoid telangiectasia syndrome
Mucous membrane
yes
Mucous membrane and/or epistaxis?;
SLE,systemic sclerosis/MCTD, dermatomyositis
yes
HHT
Generalized
yes
Mastocytosis; Generalized Essential Telangiectasia
Cutaneous collagenous vasculopathy
Photodistributed/face
yes
Papules and/or pustules?
SLE,systemic sclerosis/MCTD, dermatomyositis
yes
Acne rosacea
Generalized essential telangiectasia Mastocytosis
Acne rosacea
HHT
SLE, dermatomyositis, MCTD
Red finger syndrome
Venous ("spider veins")

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