Chapter 5
Ischemic Digital Ulcers

Fredrick M. Wigley

Abstract Systemic sclerosis (scleroderma) is unique among the rheumatic diseases because a widespread obliterative vasculopathy exists involving the peripheral arteries and microcirculation. Various forms of digital ulceration and tissue breakdown are recognized. This chapter illustrates these different lesions by presenting representative examples of each type.

Keywords Raynaud's phenomenon • Digital ulcers • Tissue ischemia • Vasculopathy • Amputation • Macrovascular disease • Fissure

Introduction

Systemic sclerosis (scleroderma) is unique among the rheumatic diseases because a widespread obliterative vasculopathy exists involving the peripheral arteries and microcirculation [1, 2]. Vascular changes involving capillaries, arterioles, and small arteries are well documented. Macrovascular disease in larger peripheral arteries is also common [3]. Pathological specimens from digital vessels demonstrate striking intimal thickening with marked luminal narrowing and evidence of thrombi, while the smooth muscle of the media is usually normal. The involved vessels normally are important in both tissue nutrition and body thermoregulation. Raynaud's phenomenon is often the initial clinical manifestation of this peripheral vascular disease (Figs. 5.1 and 5.2). The abnormal response to cold and stress is caused by the abnormalities in the regulation of regional blood flow in the skin and digits.

F.M. Wigley (✉)
Department of Medicine/Rheumatology, The Johns Hopkins University School of Medicine, 5200 Eastern Avenue, Suite 4100, Mason F. Lord Building, Center Tower, Baltimore, MD 21224, USA
e-mail: fwig@jhmi.edu

M.D. Mayes (ed.), *A Visual Guide to Scleroderma and Approach to Treatment*,
DOI 10.1007/978-1-4939-0980-3_5,

Fig. 5.1 The Cyanotic phase of Raynaud's phenomenon due to vasospasm and reduced blood flow in the cutaneous arterioles and capillaries of the fingers

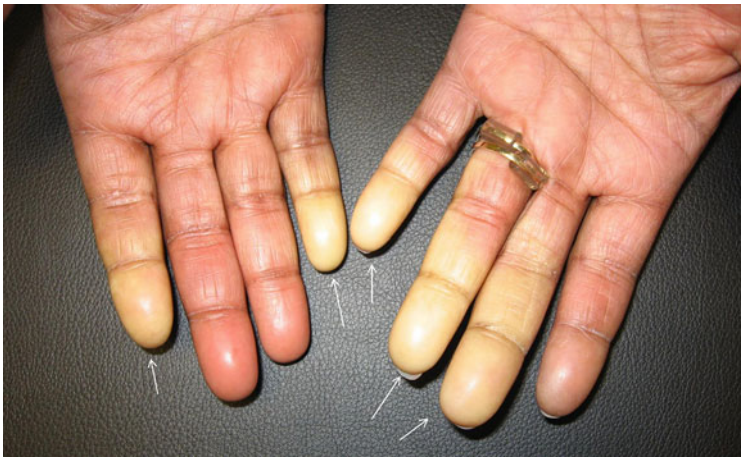

Fig. 5.2 The Pallor phase of Raynaud's phenomenon due to reversible vascular closure of digital arteries and cutaneous arterioles in involved fingers (see *arrows*)

Scleroderma is also associated with fibrotic skin, especially of the fingers. Fibrotic skin changes in association with the peripheral vascular disease lead to skin hypoxia, critical ischemia, and eventual tissue injury. Digital ulcers are reported in about 25–50 % of patients with scleroderma [4]. Prolonged critical ischemia can cause deep tissue injury and digital loss. Digital amputation secondary to occlusion of digital arteries occurs in a subset of (about 11 %) patients, usually with limited skin disease with the presence of anti-centromere antibody [5].

Various forms of digital ulceration and tissue breakdown are recognized (Figs. 5.3, 5.4, 5.5, 5.6, 5.7, 5.8, 5.9, 5.10, 5.11, 5.12, 5.13, 5.14, 5.15, 5.16, 5.17,

Fig. 5.3 Cutaneous ulcer at the site of a contracture of a proximal interphalangeal (PIP) joint in a patient with diffuse cutaneous scleroderma. These lesions occur due to trauma at the site of a joint contracture due to overlying fibrotic and avascular skin and soft tissue. In late stages of scleroderma, the skin is fragile due to secondary thinning and atrophy of subcutaneous tissue

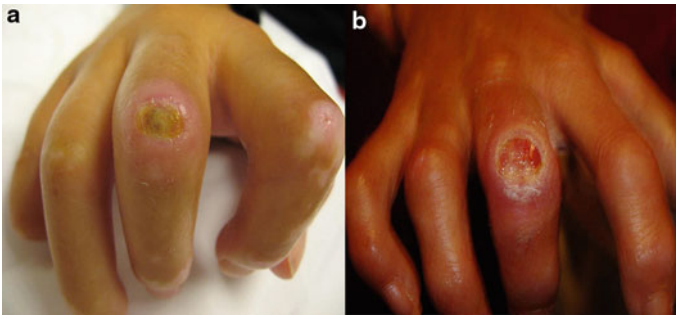

Fig. 5.4 Cutaneous ulcer at site of a contracture of the proximal interphalangeal (PIP) joint in patients with late diffuse cutaneous scleroderma. Lesion (**a**) is superficial with dry crusted exudate and no signs of secondary infection. Lesion (**b**) is deep and there is associated healthy granulation tissue in it, the ulcer bed

5.18, 5.19, 5.20, 5.21, 5.22, 5.23, 5.24, 5.25, and 5.26). These include distal digital ischemic ulcers secondary to digital artery and skin arteriole disease; breakdown of islands of skin on the digits due to microvascular occlusion in fibrotic skin; skin fissures due to drying of skin surface; ulcerations at the site of joint contractures where fragile skin is easily traumatized; deep tissue injury leading to gangrene; and loss of digits due to macrovascular occlusion of major peripheral arteries in the arm, palm, or fingers. Similar events occur in the lower extremities. This chapter illustrates these different lesions by presenting representative examples of each type of lesion.

Treatment of these lesions includes vasodilators, protection from repeated trauma, and attention to good wound care [6–8].

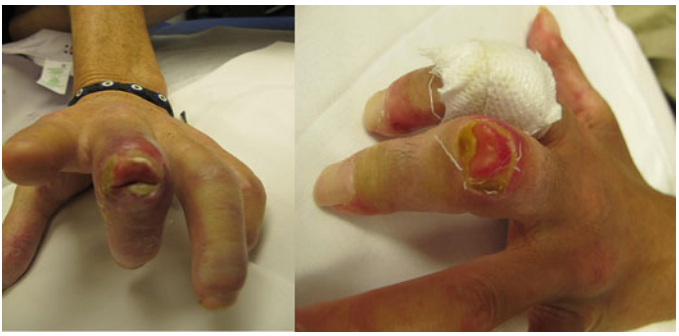

Fig. 5.5 Deep ulcerations at proximal interphalangeal (PIP) joint of a patient with late diffuse cutaneous scleroderma. Note that the joint capsule is exposed due to the large wound in the overlying avascular skin. Skin grafting or amputation is needed in such cases

Fig. 5.6 Ulcerations on the proximal interphalangeal (PIP) joint of the fingers of a patient with diffuse scleroderma. Note the crusted exudate without signs of secondary infection. These wounds are caused by injury to areas of fibrotic and avascular skin in joint contracture area subject to trauma

Fig. 5.7 Distal ischemic area of finger of patient with scleroderma. This lesion will likely develop into a shallow ulcer due to small vascular disease in the finger and skin arterioles

Fig. 5.8 Digital ulcer due to vascular compromise and tissue ischemia. This lesion is present in a patient with limited scleroderma. Note the deep ulcer has no signs of infection and is located on the distal finger typical of an ischemic ulcer due to closure of digital arteries and cutaneous arterioles

Fig. 5.9 Examples of relative small vascular disease secondary to scleroderma causing ischemic digital ulcers. These lesions will improve over several weeks and may be prevented with vasoactive drugs

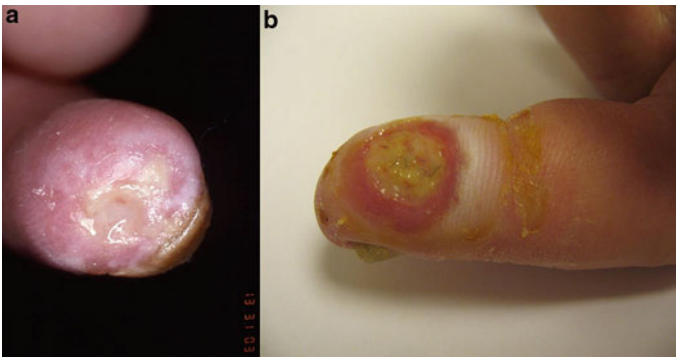

Fig. 5.10 Digital ulcers in a patient with limited scleroderma due to vascular compromise and tissue ischemia. Lesion (**a**) is deep with modest exudate, while lesion (**b**) has surrounding erythema suggesting expanding area of ischemia and/or superimposed infection. The location on the distal palmer finger is typical of an ischemic ulcer due to closure of digital arteries and cutaneous arterioles

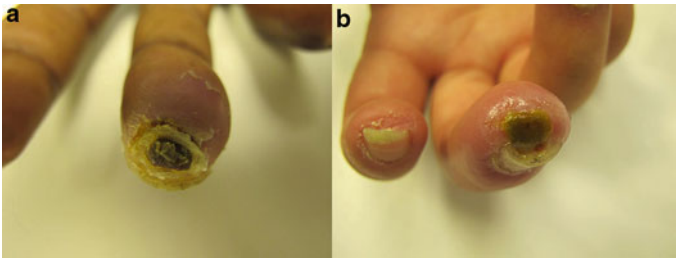

Fig. 5.11 Digital ischemic ulcers on distal fingers of patients with scleroderma. Note both ulcer lesions (**a**) and (**b**) are covered with cap-crusted dried exudate. There are no signs of infection

Fig. 5.12 A patient with limited scleroderma presents with digital lesion typical of superficial infarction of distal finger. Note the areas of dry gangrene without signs of secondary infection. Ulceration of the tissue found under this dry blackened tissue. These lesions are secondary to digital artery disease with vascular occlusion

Fig. 5.13 Healing ischemic digital ulcer on distal thumb in a patient with scleroderma. Raynaud's phenomenon and/or digital ulcers on the thumb are a manifestation of an underlying secondary vascular disease. The thumb is often spared in patients with primary Raynaud's phenomenon

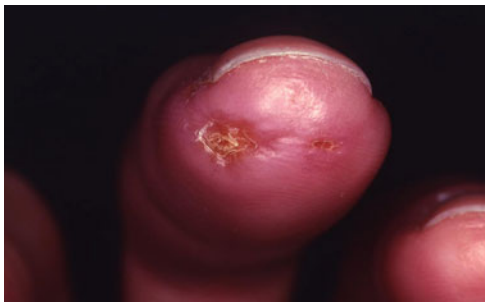

Fig. 5.14 This is an example of a digital pit typical of a healed ischemic event seen in patients with scleroderma. These events are secondary to small vessel disease in the digital and cutaneous circulation

Fig. 5.15 Healed distal ulcer in a patient with scleroderma. Superficial lesions often leave painless crusted dry material at site of lesion with small pitting of skin

Fig. 5.16 Note the shallow ulcer on the lateral finger of these patients with scleroderma. These lesions represent small vessel disease in the skin of the distal finger. Often these lesions expand along the shaft of the finger following areas of fibrotic and avascular skin

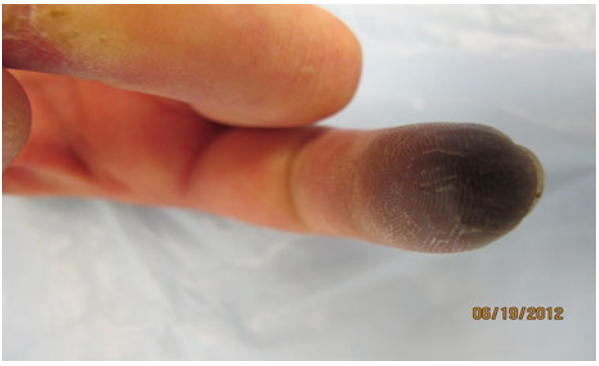

Fig. 5.17 Patient with acute distal finger ischemia due to digital artery vasospasm and impending occlusion. This type of presentation is seen prior to deep tissue infarction that will occur if blood flow to the finger is not improved

Fig. 5.18 Major digital artery occlusion has lead to distal infarction of finger in a patient with limited scleroderma. Note the sharp demarcation defining level of tissue perfusion. Patients with anti-centromere antibodies are at increased risk for these larger vessel occlusions and subsequent digital amputation

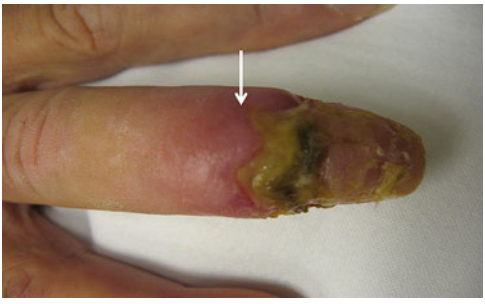

Fig. 5.19 Patient with limited scleroderma with digital artery occlusion leading to infarction of distal finger. Note area (see *arrow*) of hyperemia representing Ischemic area with vasodilated small cutaneous vessels attempting to limit further tissue injury

Fig. 5.20 Major artery disease in a patient with limited scleroderma leading to deep tissue infarctions. This type of event leads to amputation of the distal finger. Patients with anti-centromere antibody are at increased risk for these larger vessel events

Fig. 5.21 Two examples of larger digital artery occlusion leading to deep tissue injury and dry gangrene. These lesions are completed and amputation beyond the area of the demarcation will result

Fig. 5.22 Deep and superficial digital tissue infarctions in patients with scleroderma. This type of presentation is secondary larger digital artery disease coupled with compromise in the cutaneous arterioles

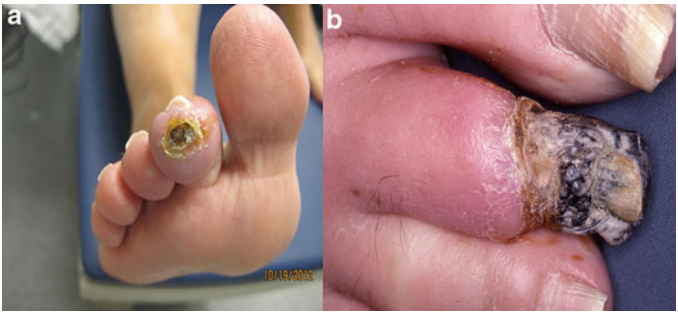

Fig. 5.23 Digital lesions of the toes in patients with scleroderma. Note lesion (**a**) is a small vessel-occlusive event typical of an ischemic ulcer on distal toe. Lesion (**b**) is due to larger digital artery disease and is often associated with macrovascular disease in the distal limb. In both cases Doppler ultrasound was used to rule out any correctable proximal larger vessel lesion

Fig. 5.24 Paronychia lesion due to small vessel disease in the nailfold. This is seen in this patient with diffuse scleroderma and is often associated with soft tissue infection

Fig. 5.25 Areas of subcutaneous calcinosis in distal fingers of patients with scleroderma mimicking digital ischemic ulcer. The X-ray (*left*) demonstrates a cluster of calcium hydroxyapatite in the subcutaneous tissue. The clinical picture (*right*) shows the hard white lumps of calcium under the skin. These lesions can spontaneously drain, cause local Inflammation, or they can ulcerate and get secondarily infected. Surgical removal can be done if problematic

Fig. 5.26 Fingers of patient with limited scleroderma with painful fissure (see *arrow*). Note the areas of cyanosis consistent with compromised blood flow due to ongoing vasospasm of cutaneous arterioles. Fissures are often mistaken for Ischemic ulcers but most commonly occur due to dry fibrotic skin

References

1. Rodnan GP, Myerowitz RL, Justh GO. Morphologic changes in the digital arteries of patients with progressive systemic sclerosis (scleroderma) and Raynaud phenomenon. Medicine. 1980;59:393–408.
2. Matucci-Cerinic M, Kahaleh B, Wigley FM. Review: evidence that systemic sclerosis is a vascular disease. Arthritis Rheum. 2013;65(8):1953–62.
3. Veale DJ, Collidge TA, Belch JJ. Increased prevalence of symptomatic macrovascular disease in systemic sclerosis. Ann Rheum Dis. 1995;54(10):853–5.
4. Steen V, Denton CP, Pope JE, Matucci-Cerinic M. Digital ulcers: overt vascular disease in systemic sclerosis. Rheumatology (Oxford). 2009;48 Suppl 3:iii19–24.
5. Denton CP, Krieg T, Guillevin L, Schwierin B, Rosenberg D, Silkey M, Zultak M, Matucci-Cerinic M, DUO Registry Investigators. Demographic, clinical and antibody characteristics of patients with digital ulcers in systemic sclerosis: data from the DUO Registry. Ann Rheum Dis. 2012;71(5):718–21.
6. Herrick AL. Management of Raynaud's phenomenon and digital ischemia. Curr Rheumatol Rep. 2013;15(1):303.
7. Tingey T, Shu J, Smuczek J, Pope J. Meta-analysis of healing and prevention of digital ulcers in systemic sclerosis. Arthritis Care Res (Hoboken). 2013;65(9):1460.
8. Lambova S, Müller-Ladner U. Connective tissue diseases: treatment of digital ulcers in systemic sclerosis. Nat Rev Rheumatol. 2011;7(1):5–6.