Systemic Sclerosis: "Scleroderma"

BY Paul Chapman

Systemic sclerosis (SSc) is a relatively rare condition but is an important entity that wound-care practitioners should be aware of. Data from the United States report an incidence of 10 to 20 new cases per million people per year. This would equate to 300 to 600 new cases per year in Canada. The disease is three to four times more common in females than in males. It is rare before age 30 but can occur at any age. Most cases are seen between 40 to 80 years of age.

Systemic sclerosis, as the name implies, is a disease that involves sclerotic (fibrotic) changes in many organ systems throughout the body. The disease is categorized as limited or diffuse, based on the degree of cutaneous involvement. The number of organs involved generally parallels the cutaneous distribution. With limited disease, fewer organ systems are involved, and the skin changes are mostly in distal extremities. With diffuse disease, there is a greater degree of organ involvement, and skin changes include the extremities, face and trunk.

Prognosis

In a meta-analysis, Ioannidis et al. conclude that there

TABLE 1

Systemic Sclerosis Involvement

Organ System	Manifestations
Vascular	Raynaud's phenomenon: Triphasic response to cold; hands turn white, blue, then red. Vasospasm may be so severe as to cause digital ischemia with eventual necrosis and autoamputation.
Heart	Fibrosis and thickening of vessels → occlusion of vessels with associated infarcts of cardiac tissue. May then develop arrhythmias, pericarditis, congestive heart failure.
Skin	Dermal fibrosis, loss of subcutaneous fat, epidermal atrophy → loss of sweat glands, hair → thick, tight, dry skin with decreased joint mobility that is more prone to damage/infection Characteristic "mask-like" face with beak nose and radial perioral furrows.
Gastrointestinal	Atrophy and fibrosis of GI-tract → decreased motility, dysphagia, gastroesopageal reflux (with heartburn), possible ulcers/strictures. Often have diarrhea or constipation with malabsorption and bacterial overgrowth.
Muscluloskeletal	Affects joints and muscles. Sclerosis of synovium, which may lead to resorption of underlying bone (seen often in terminal phalanges), fibrosis and atrophy of muscle with tendonitis, arthritis.
Renal	Fibrosis of small arteries may lead to focal areas of necrosis of renal tissue and possible malignant hypertension. If untreated → renal failure.
Lung	Fibrosis and thickening of lung tissue and vessels resulting in non-productive cough, shortness of breath, fatigue and pulmonary hypertension.

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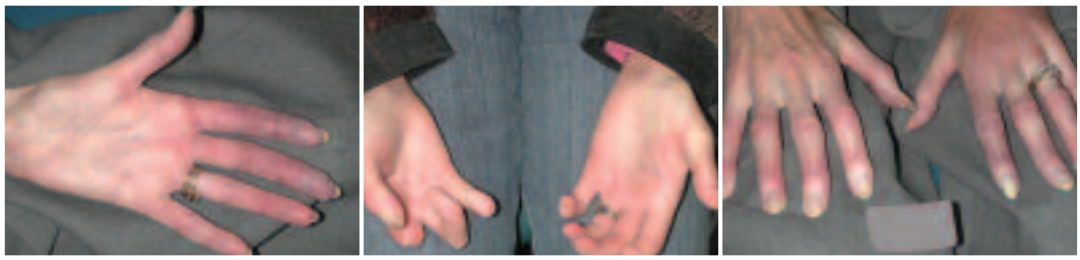

Skin damage resulting from sclerosis.

is a high mortality risk (from 1.5–7.2) for persons with SSc. Rates increase with internal organ involvement and anti-topoisomerase antibodies.[1]

Causation

Arnett et al. report that "A positive family history of SSc is the strongest risk factor yet identified for SSc."[2] There is also evidence to suggest links to environmental agents; occupational exposure to solvents may be a risk factor for developing SSc.[3,4] However, the cause(s) of this disease remains unknown. Whatever the cause, three components to the mechanism causing damage are typical: fibroblasts over-produce collagen and other extracellular matrix proteins; damage to, and subsequent thickening of, vessel walls with narrowing (and possible obliteration of the vessel lumen) manifests as vasculopathy; and an element of autoimmunity develops with auto-antibodies directed against cellular nuclear elements such as the centromere, topoisomerase I and RNA polymerases.

With so many systems affected by this disease (nutrition, circulation, oxygenation, cardiac output, etc.) and with tight, dry, thin skin, the person with SSc can easily be wounded and experience difficulty in healing.

Globally, one's ability to perform activities of daily living is detrimentally affected by SSc.

Raynaud's Phenomenon

Virtually all persons affected with SSc develop Raynaud's phenomenon. The resultant vasospasm limits its distal circulation and hinders wound healing; in some cases it is so severe that autoamputation results. Many pharmacological treatments for Raynaud's have been studied. Some have been found to be beneficial, others not. Calcium channel blockers or prostacyclin (or synthetic derivatives such as Iloprost) have been used to "relax" the vasculature. ACE-inhibitors, ARBs or alpha-blockers have been used to decrease peripheral vascular resistance. Side effects are common, as with any medication, and may outweigh the benefit derived.

In a study of leg ulcers in patients with collagen vascular disease, Hafner et al. report that five of their six SSc subjects had concomitant arterial and venous disease.[5]

Treatment

As the disease is not well understood, treatments are, at times, of an experimental nature based on clinical experience. A number of studies have used the agents listed below with varying results. They are listed here by way of example, rather than as treatment suggestions. The reader is urged to study the literature on particular agents prior to using any of them.

Regarding wounds: local wound care employing the principles of wound bed preparation of moist, interactive healing is the mainstay of treatment.[6] Patients often avoid cold and wear hats and mittens to prevent distal extremities from entering into vasospasm. Emolients may be beneficial in maintaining a certain moisture level in the skin in an effort to reduce the likelihood of skin drying and breakdown.

Some newer studies may have practical wound-care implications:

- A pilot study (17 patients with SSc) by Sandqvist et al.[7] investigated the effects of hand exercises and the use of warm (50°C) paraffin baths in persons with SSc. Treatments were carried out daily for one month. Measurements were made at baseline and at one month. Participants experienced significantly increased mobility and a decrease in perceived stiffness of the treatment hand (the opposite hand was used as the control).
- Another study investigating the effects of stretching exercises on mouth-opening (n=10) demonstrated a significant increase in opening after the 18-week period in all patients. Patients also reported clinical improvements in eating, speaking, oral hygiene and denture insertion.[8]
- In a pilot study of 26 patients Pfizenmaier et al.[9] investigated whether Intermittent Pneumatic Compression would aid in the healing of ulcers in the upper extremity, as it had been shown to do with lower extremity ulcers. Compression treatments were for five hours per day. They report a 96 per cent (26/27 ulcers) healing rate with a mean healing time of 25 weeks.

TABLE 2

Agents and Interventions to Treat Systemic Sclerosis

System	Agent/Intervention	Desired Effect
Vascular	Calcium channel blocker	Vasodilation
	ACE-inhibitor/ARB	Vasodilation
	α-1 blocker	Vasodilation
	Prostecyclin (and analogues)	Vasodilation
	Smoking cessation	Vasodilaton
	Nitroglycerin patch	Vasodilation
Gastrointestinal	Octreotide	Increase in gastric motility
	Antibiotics	Prevention of bacterial overgrowth
	Proton pump inhibitors, Histamine H2 blockers	Decrease in gastric acid production
Renal	ACE-inhibitors	Prevention of renal crisis
Cardiac	Antiarrhythmics	Antiarrythmic activity if heart so affected

Summary

SSc is a relatively rare disease that ranges from mild to extremely debilitating. With extensive involvement, manifestations are protean and wound-healing is difficult. Wound care may involve systemic agents, surgery, exercise, and augmentative therapy, as well as local wound care. A patient-centred, multidisciplinary approach will best serve the patient as this is a complicated and multifaceted disease process.

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