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Scleroderma: considerations for dental hygienists

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Abstract: Scleroderma, the general name of a group of progressive diseases affecting the connective tissues is the most deadly of the varying connective tissue disorders. Characterized by abnormal thickening of the skin, this collagen-vascular disease is associated with immune dysfunction. Hallmark signs of scleroderma include fibrosis, vascular instability and initial inflammation resulting from excessive collagen deposition. Oral facial involvement is considerable, necessitating adaptations in patient oral self-care and influencing oral hygiene. Appropriate dental hygiene management of patients with this autoimmune disorder requires an understanding of clinical characteristics, the recognition of oral facial involvement, treatment considerations and pharmacological interventions. With this information, dental hygienists will be better prepared to provide compassionate, safe and effective dental hygiene management and care to patients with scleroderma.

Key words: campaigns; care; dental hygienist; institutionalized; knowledge; mentally retarded; oral health; patients; problems; special care; status

Introduction

Scleroderma results from an excess production and accumulation of collagen, resulting in chronic hardening and thickening of the skin. While the prevalence of scleroderma is far less than arthritis, it still affects up to two of 10 000 persons (1, 2). As with rheumatic diseases, scleroderma affects women more than men (5:1), with onset generally between 30 and 50 years of age (3, 4). The worldwide incidence of scleroderma is most likely greater than estimates as this disorder is often misdiagnosed and confused with other autoimmune disorders. Much variation in clinical manifestations and rate of progression add to the confusion, making diagnosis difficult.

However, early diagnosis is critical as early therapeutic protocols may improve patient lifestyles, slow disease progression and assist with better disease management. Several patterns of symptoms assist with diagnosis and initial patient complaints often focus on the changes in the skin of the face and hands.

Types of scleroderma

There are two major forms of scleroderma: localized scleroderma and systemic scleroderma (SSc). Localized forms of scleroderma are characterized by asymmetrical skin indurations and thickening (5). Internal organs are not affected. By contrast, the systemic forms involve the internal organs and skin lesions manifest with symmetric cutaneous induration and thickening. Because the connective tissue fibrosis affects the internal organs, SSc can be life threatening and interstitial lung disease is the most common cause of death.

Localized scleroderma is divided into two morphologic variants; morphea and linear. While this form is not fatal, the pain as well as the hand and facial deformities that result often compromise the patient's quality of life (6). The linear type is most commonly found in children and is characterized by thickened and abnormally coloured skin. The skin changes are typically linear or band like and commonly affect the lower limbs and torso and to a lesser extent the arms and face (7). Morphea scleroderma is characterized by thickened sclerotic skin lesions involving the hands, face, neck, arms and torso. These benign patches of dry, thickened skin are usually painless but itchy and can be successfully managed simply with emollients for dry skin (5).

Systemic scleroderma also known as systemic sclerosis (SSc) targets the vasculature of both the skin and multiple organ systems including the pulmonary, cardiac, gastrointestinal and renal systems. Chronic hardening and shrinking of the connective tissue of any organ in the body may occur. As the fibrosis progresses, life-threatening organ failure manifests (7). Subclassification of SSc includes limited and diffuse scleroderma. Individuals with either subtype are commonly afflicted with Raynaud's phenomenon. The hands as well as the feet are frequently affected by this episodic circulatory disorder. Typically occurring with temperature changes especially cold, Raynaud's phenomenon is characterized by ischemia to the fingers, toes, ears and nose. Hallmark signs include extremity blanching of the skin, cyanosis and redness. In addition to the triphasic colour change, numbness, tingling, discomfort, severe pain, ulceration and problems with hand function are common sequelae (8, 9).

The thickened sclerotic skin lesions that characterize limited scleroderma primarily affect the hands, torso and extremities. Manifestations of the skin lesions include calcinosis, oedema and telangiectases. Painful tightening of the skin occurs with disease progression. The skin eventually binds with underlying structures and painful ulcerations occur at the joints. Other symptoms include severe itching related to skin dryness, digital ulcerations and infection. Gastrointestinal problems and pulmonary hypertension are the most common systemic manifestations and, although rare, other organ involvement can occur (8, 9).

Diffuse scleroderma is associated with visceral involvement of many organ systems as well as the skin changes associated with limited scleroderma. Skin changes may start as a painless swelling or pitting oedema distal in the extremities in the early course of the disease. Eventually, the skin becomes indurated, smooth, atrophic and bound to the subcutaneous tissues and affected with telangiectases. Progressive painful skin tightening results in a significant reduction in the range of motion affecting all parts of the body with facial features becoming mask like (8). The severe fibrosis of the skin especially in the fingers and hands results in a claw-like hand deformity causing major disability (10). Severe skin dryness and pruritus experienced by those afflicted with diffuse SSc is related to the disabled sebaceous and sweat glands from the collagen deposited in the upper dermis. In addition, most individuals with diffuse SSc will present with Raynaud's phenomenon as well as joint and muscle pain. Renal problems, cardiac involvement and interstitial lung disease are common systemic manifestations associated with the diffuse form which have life-threatening consequences (11).

Pathogenesis

The pathogenesis of SSc involves three findings: (i) vascular dysfunction that manifests as injury to the endothelial cells; (ii) immunological activation of T cells, cytokines and inflammation; and (iii) fibrosis. Inflammation manifests initially as a perivascular macrophage infiltrate (9, 11, 12). Injury to the endothelial cells causes prominent thickening of vessels and two of the most serious complications, pulmonary arterial hypertension and renal crisis (13). Increased deposition of fibroblasts causes destruction of normal tissues which forms the basis for the tissue and organ dysfunction that occurs in the skin, lungs, kidneys and GI tract (7). All three disease pathways are most likely linked. Research suggests that fibroblasts and myofibroblasts that over-produce collagen and other extracellular matrix products are stimulated via the by-products

of the functional vasculopathy. B cells also are involved in the pathogenesis as there are multiple scleroderma-specific antibodies; however, no correlation with specific pathological damage has been found (13).

Treatment

Currently, most pharmacological interventions are aimed at suppressing the immune system, dilating blood vessels and increasing blood flow in addition to treating individual organ problems (14–16). Because of the complicated pharmacological interventions associated with SSc as well as possible organ dysfunction impacting on immune functioning, consultation with the patient's rheumatologist may be prudent prior to dental hygiene treatment. Several of the drug regimens have side effects that may require adaptation in dental hygiene treatment (17). Methotrexate and cyclosporine are common immunosuppressive drugs prescribed for patients with SSc. Antibiotic premedication prior to dental hygiene care may be necessary for patients taking these drugs due to the immune suppression that results. An increased incidence of fungal infections may also be encountered in individuals taking these as well as a slower recovery from periodontal debridement therapy due to prolonged wound healing. Nifedepine and amiodipine are prescribed for treating Raynaud's phenomenon. Patient's gingival condition may be affected by nifedepine causing drug-influenced gingival enlargement.

Patients may also be on corticosteroid therapy and would need to be medically evaluated for possible premedication due to immune suppression. Patients should also be evaluated for adrenal insufficiency which would require supplemental glucocorticoid medication prior to treatment. Because of problems with renal function in advanced cases, medications prescribed to manage dental pain and infection should be closely evaluated as to their effect on the kidneys and alternative mediations not secreted by the kidneys may be warranted.

Oral concerns

The promotion of health and the prevention of complications is the treatment paradigm for patients with scleroderma (6). Regular oral care is essential if the goal of optimal health and well-being is to be met. Due to the many vascular, inflammatory and fibrotic changes associated with scleroderma, patients are more prone to oral disease. Early treatment interventions are extremely important to prevent systemic complications from oral infection as well as complications when implementing dental hygiene care. As the disease progresses, patients may not tolerate long chair time and will experience more difficulty opening their mouth limiting practitioner's access. In an effort to be more empathetic to the patient, oral healthcare professionals must be cognizant of the psychological toll this disease takes on many patients due to the disfiguring facial changes as well as its overall debilitating and painful effects (18, 19).

The dental hygienist has a key role in assisting patients to maintain oral hygiene, identifying oral manifestations, providing self-care education as well as modifying treatment to ensure safe, compassionate and effective dental hygiene care. Common oral manifestations associated with scleroderma are listed in Table 1. The dental hygienist must be aware of the oral manifestations to provide better care and lower the risk of oral disease. Identification of the oral manifestations is also important as these can be precursors to life-threatening systemic involvement.

Oral manifestations

Several oral findings may be predictors of systemic involvement. The most frequent oral finding to precede systemic involvement appears to be trigeminal neuropathy followed by enlargement of the periodontal ligament (PDL) space (20, 21). The trigeminal neuropathy is characterized by slow and gradual facial muscle inactivity followed by pain, sometimes severe, and parasthesia. Dental professionals not finding a dental cause of this pain should suspect SSc as these symptoms may occur several years before a diagnosis has been made. The neuropathy is associated with deposition of collagen in the perineurium and or reduced vascularity to the trigeminal nerve itself (22, 23). In addition, enlargement of the PDL space is a common manifestation of SSc and is related to the fibrotic thickening of the PDL. This manifestation typically affects all teeth. With no known cause, such as occlusal trauma, SSc should be considered as unexplained PDL widening may be pathognomonic for SSc (22).

Table 1. **Common oral findings in patients with scleroderma**

Microstomia
Xerostomia
Dysphagia
Fibrosis at the hard and soft palate
Increased risk of periodontal disease and caries
Widening of the periodontal ligament space
Enamel erosion
Mandibular resorption
Trigeminal neuropathy
Facial and mucosal telangectasis

Resorption of the mandible is another common oral manifestation. The mandibular resorption results from facial skin tightening, vessel constriction and the underlying taut musculature exerting continuous pressure on the mandible (23–25). Often asymptomatic, the areas of mandibular resorption are of concern as they increase the risk of mandibular fractures and painful oral conditions such as trigeminal neuropathy and osteomyelitis (24). The angle of the mandible, the condyles and coronoid process may be affected. Dental hygienist should plan for regular panoramic radiographs as an important screening mechanism for the early detection of oral bone resorption in patients with SSc.

Another oral finding of significance is the increased risk of oral cancer that has been reported for some individuals with scleroderma. In particular, a recent study found an increased risk of squamous cell carcinoma of the tongue (26). Hence, providing regular and thorough oral cancer examinations is extremely important in patients with SSc.

Of particular importance to oral healthcare professionals is the skin involvement affecting the hands and oral facial structures. Both impact the patient's ability to perform the activities of daily living including oral self-care, movement of oral structures and increased risk of oral disease (18, 27, 28). One of the most prevalent oral findings is the excess collagen deposition affecting the peri-oral structures (27). Many of the debilitating resultant functional problems relating to scleroderma result from microstomia. Due to the loss of elasticity and subsequent tightening of the lips and checks, both self-care and professional care can be challenging. Having realistic expectations about what a patient can accomplish and being encouraging at all times is an important strategy for providing compassionate, quality care. Instructing patients on simple stretching exercises of the mouth (see Table 2) can significantly improve flexibility and opening and assist with a more productive dental hygiene appointment (29–31). Likewise sharing with the patient some simple stretching exercises involving the fingers (Table 3) have been found to improve function and could improve self-care measures (32–34).

The major extremity discomfort from Raynaud's phenomenon and scleroderma, along with oral-facial and hand functional limitations result in oral self-care being compromised. The dental hygienist will need to discuss with the patient ways to maintain their oral hygiene and at the same time have realistic expectations about what can be accomplished when function is compromised and discomfort is high. Due to hand deformities and functional limitations, enlargement and or extension of the toothbrush handle may facilitate improved plaque biofilm removal via tooth brushing (22, 34).

Table 2. **Stretching exercises to increase range of motion of the mouth**

Exercise 1

Instruct patient to:

1. Make an O with your mouth
2. Smile, grimace, smile, grimace
3. Open your mouth as wide as you can, and stretch as much as possible
4. Practice slowly 10 times a day

Exercise 2

Instruct patient to:

1. Stack four tongue blades together and secure with clean rubber band
2. Open mouth and slip stack between the upper and lower teeth
3. Determine the number of tongue blades that can comfortably fit between the teeth
4. Next add one additional tongue blade in the middle of the stack and let it gently stretch the mouth
5. Gradually increase the number of blades in the stack to stretch the mandibular opening
6. Repeat at least two times a day

Table 3. **Finger stretching exercises to improve function**

Instruct patient to:

1. Place arms at sides with fingers pointed toward the toes
2. Place palms facing your body
3. Curl fingers and attempt to touch your finger tips to your palm
4. Slowly uncurl and straighten fingers as much as possible
5. Repeat 10 times a day

In addition, powered toothbrushes and flosses may promote improved oral hygiene, although some patients may encounter difficulties fitting the device in the mouth due to their restricted opening. Most patients will need a compact head or pediatric toothbrush to gain maximum access. The buccal side of the teeth is often the most difficult to access even with a pediatric brush. Taking tweezers and removing two rolls of bristles has been suggested as a way to assist patient access to these areas (35). An end tuft brush could also be suggested due to the small head allowing for improved oral access.

Teflon-coated floss may be too slippery for some individuals with scleroderma. Dental tape may improve flossing techniques as this product has been reported to be easier to hold on to (31). Wrapping the floss around the fingers is problematic for many patients with Raynaud's phenomenon due to its cause of digital pain and ulceration. Most patients would therefore benefit from self-care education from the dental hygienists focusing on alternative ways to clean interdentally with flossing devices that are hand held or powered. In some cases, consultation with an occupational therapist will be necessary to meet the challenges of finding adaptive devices that may maximize oral self-care by the patient.

Due to the fibrosis and atrophy of major and minor salivary glands, patients with SSc experience a high incidence of xero-

stomia. The loss of salivary function is significant and similar to that afflicting patients after radiation therapy to the head and neck (36). Xerostomia adds other complications to the patients, diminishing quality of life by increasing the risk of caries, periodontal disease and fungal infections. Nutritional status, dietary habits, swallowing, taste, speech and tolerance to dental appliances may be adversely affected. Several strategies that the dental hygienist may recommend to minimize these adverse affects include use of salivary stimulants, fluoride therapy and antimicrobial mouth rinses. The use of a daily 1.1% sodium fluoride prescription rinse or a 4% sodium fluoride brush on gel are important for reducing caries risk (37). Many patients may benefit from the use of a custom fluoride mouth tray maximizing contact of fluoride gels with the teeth. Some patients, however, may not be able to fit the fluoride tray in the mouth. The application of fluoride varnish may be well suited for many of these patients as the small applicator brush allows for easier access into the oral cavity and provides excellent anticaries benefits (22).

Antimicrobial rinses are important for combating periodontal diseases, but only those without alcohol in the formulation should be recommended. Cetylpyridium chloride rinses from Procter and Gamble and alcohol-free chlorhexidine from Sunstar Butler are two good choices. Use of the chlorhexidine rinse may also lower the risk of oral fungal infections affecting individuals with scleroderma as it has proven broad spectrum activity against Gram-positive and Gram-negative organisms (36).

To assist patients with salivary dysfunction pharmacological options are strongly recommended. Swallowing function, diet, speech, taste and the incidence of oral disease may all show improvement through the use of cholinergic agonist agents, such as pilocarpine (Salagen®) and cevmeline hcl (Evoxac®) that significantly improve salivary flow in patients experiencing hypofunction and hence, provide many beneficial effects (37). Salivary substitutes, made of carboxymethylcellulose, are also important medicaments to recommend for scleroderma patients experiencing xerostomia as they possess chemical properties and a viscosity similar to saliva. In contrast to cholinergic agonist agents whose effects can last from 3 to 5 h, carboxymethylcellulose preparations only provide palliative, short-term benefits like oral lubrication that can improve oral function. Salivary substitutes that contain fluoride, calcium ions and phosphorous ions may provide an added benefit to the scleroderma patient due to their remineralization properties (22).

A combination of oesophageal neural damage, vascular changes and fibrosis causes gastrointestinal reflux (GERD) and affects approximately 90% of SSc patients (12). Due to the acid from reflux, perimylolysis tooth erosion may occur resulting in hypersensitivity (20, 23). Use of sodium fluoride home rinses as well as fluoride dentrifices can assist with reduction in problems caused from hypersensitivity. Additionally, use of sodium fluoride products assists with rebuilding of enamel caused from acid reflux. Moreover, avoidance of acid-stimulating foods and regular use of anti-acids can help with disease management of GERD.

Due to the changes associated with the oesophagus, as the disease progresses many patient experience dysphagia in addition to GERD (4, 11). The hard and soft palate fibrosis leading to immobility and the dysphagia would contra-indicate use of air polishing and powered scaling devices in many patients with SSc.

Besides the avoidance of powered instrumentation devices, several other adaptations in dental hygiene care planning are necessary to ensure a safe and effective appointment. Scheduling the patient at the time of day they feel best and keeping the appointment as short as possible can facilitate more successful dental hygiene interventions. When providing dental hygiene care, due to tongue fibrosis, retraction might be difficult for the clinician. Having an assistant is helpful as they can retract and assist with more efficient delivery of care. To prevent the aspiration of oral debris, high-speed suction is imperative. A mouth prop will prove to be helpful in gaining oral access and assisting the patient in keeping the mouth open.

Patients may experience discomfort trying to open and sustain the opening long enough for care. Suggesting the use of a mild analgesic 1 h before the appointment such as Tylenol® may make the patient more comfortable (35). Exposing of radiographs when a patient is afflicted with microstomia can be problematic. Dental hygienist may be more successful with radiographs if size one or size zero film and or digital sensors over size two film or sensors are used (28).

To control oral infection, frequent re-care is highly advisable. Every 2- to 3-month re-care appointments are ideal to prevent the need for long, difficult appointments where more work and longer chair time may be needed. Dental hygienists encountering patients with the symptoms associated with Raynaud's phenomenon with no known reason should refer the patient for a prompt medical evaluation as this disorder is often a precursor to scleroderma and/or other autoimmune disorders. Importantly, early diagnosis can assist with more successful disease management. As the small blood vessels in the extremities are affected, patients with Raynaud's phenomenon often become overly sensitive to changes in ambient temperature (9). Therefore, dental hygienists may want to adjust the

temperature; so, it is warmer in the oral care setting, and avoid treatment rooms that are subject to drafts and have a blanket and gloves or hand warmer available for the patient in case they do not have their own. These additions can facilitate a more comfortable and less stressful appointment for the patient.

As stress exacerbates the painful effects of Raynaud's phenomenon, a stress-free appointment is helpful. Other suggestions to minimize stress during the appointment include facilitative guidance, aroma therapy, effective pain management and soothing music. Some patients may benefit from the use of anti-anxiety medications. Reminding patients to avoid smoking, alcohol, excessive intake of caffeine and second-hand smoke as they cause peripheral vasoconstriction and contribute to xerostomia are practical suggestions dental hygienists may wish to reiterate with patients as well. Because of the extreme dryness experienced by many patients with SSc, a room humidifier in the treatment area can be useful. The humidifier may be helpful in keeping the tissues hydrated. The dental hygiene appointment may be more successful as the hydration helps ease skin tension as well as improve movement of tight skin, which are potential problems during the appointment.

Conclusions

Scleroderma is a chronic connective tissue-vascular disease related to immune dysfunction. Hallmark signs of this disease involve excess collagen deposition affecting the connective tissues of the skin and some if not all of the internal organs of the body. Oral facial involvement is significant and affects the oral health status of the patient. Dental hygienist must be cognizant of the oral signs and symptoms as well as appropriate management strategies to provide optimal dental hygiene care in an empathetic, appropriate, comfortable and efficient manner.

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