

Case Report

Spinal Cord Stimulation Utilization to Treat the Microcirculatory Vascular Insufficiency and Ulcers Associated with Scleroderma: A Case Report and Review of the Literature

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Abstract

Objective. To report a case of scleroderma with associated Raynaud's phenomenon and its successful treatment with spinal cord stimulation. To demonstrate the use of transcutaneous oxygen pressure monitoring to guide the progression from trial to implantation and to assess post-implantation microcirculatory recovery.

Design. Case report and literature review.

Patient. A 51-year-old female with scleroderma, associated Raynaud's phenomenon, and a non-healing 3.7-cm lower extremity ischemic ulcer. Ankle-brachial indexes demonstrated normal macrocirculation, but transcutaneous oxygen pressures demonstrated significant microcirculatory insufficiency.

Intervention. Treatment was a spinal cord stimulator implantation after a successful trial. Transcutaneous oxygen pressures were interpreted during the trial and post-implantation stages.

Results. Based on a 5-day trial that documented improvements in transcutaneous oxygen pressures and pain relief, the patient underwent implantation. At 4 months, the ischemic ulcer had healed. The patient had significant improvement in pain control and reduced Raynaud's phenomenon signs and symptoms. At 18 months, the patient continued to have improvement with no associated complications. A literature review demonstrated only four published reports, including a total of 18 patients, on spinal cord stimulator treatment for scleroderma and associated Raynaud's phenomenon.

Conclusions. We report the healing of a greater than 3-cm ischemic ulcer in an individual with normal macrocirculation but severe microcirculatory insufficiency from scleroderma. Improvements in microcirculation correlated with wound healing. Spinal cord stimulation may be considered for select individuals with microcirculatory reserves that can be modulated with treatment.

Key Words. Spinal Cord Stimulation; Scleroderma; Raynaud's Phenomenon; Transcutaneous Oxygen Pressure

Introduction

Scleroderma is a degenerative autoimmune disease associated with significant microvascular alterations and deficiencies [1]. The microcirculatory deficiencies may cause tissue necrosis and painful ulcerations that are often unresponsive to conservative treatment. Individuals with scleroderma may also experience Raynaud's phenomenon, a vasospastic disorder classified by prolonged cyanosis and ischemia of the toes or fingers in response to a cold stimulus or emotional stress [2,3].

Spinal cord stimulation (SCS) may have value in the treatment of the microcirculatory deficiency associated with scleroderma. SCS is widely used in Europe to treat inoperable atherosclerotic peripheral vascular disease. However, its use in the United States for this condition is limited [4]. Only a small number of reports exist on the use of SCS for microcirculatory vascular abnormalities linked to scleroderma. We describe the successful use of SCS to

mediate pain, increase tissue perfusion, and treat a non-healing skin lesion in a patient with Raynaud's phenomenon secondary to scleroderma. Transcutaneous oxygen pressure (TcPO₂) monitoring was used to guide decision-making and monitor microcirculatory function during the SCS trial and postoperative stages. In addition, we reviewed the pertinent literature regarding the use of SCS treatment for the vascular complications associated with scleroderma.

Case Report

A 51-year-old female with scleroderma and associated Raynaud's phenomenon was referred for assistance with pain control, wound healing, and sympathetic blocks. She had a non-healing ischemic ulcer on her left lower extremity measuring 3.7 cm long by 2 cm wide by 0.1 cm deep (Figure 1). The ulcer had not responded to 8 months of treatment including multiple debridements and topical collagen dressings. Her left lower extremity pain was an 8/10 on a 0 to 10 numerical rating scale (NRS), and she was taking short-acting oxycodone for pain control. She

reported coldness, color changes, and intermittent edema in her lower extremities. Cold ambient temperatures significantly intensified her symptoms.

Physical examination demonstrated bilateral 3+ femoral pulses and non-palpable dorsalis pedis and posterior tibial pulses in her left foot. The patient presented with evidence of pallor, hyperalgesia, allodynia, and dystrophic nails in her left lower extremity. Medical history was significant for tobacco use, chronic obstructive pulmonary disease, hypothyroidism, gastroesophageal reflux disease associated with esophageal dysmotility, and sclerodactyly.

Vascular studies were performed. Ankle-brachial indexes (ABIs) indicated no significant atherosclerotic disease or macrocirculatory deficiency in either extremity (R: 1.0, L: 1.04). Waveform analysis demonstrated normal multiphasic waveforms throughout both lower extremity arterial systems. TcPO₂ monitoring evaluated the current status of the microcirculatory function. Baseline TcPO₂ data were collected with an electrode at 44°C on the dorsum of the respective foot at a room temperature of 21°C in both the

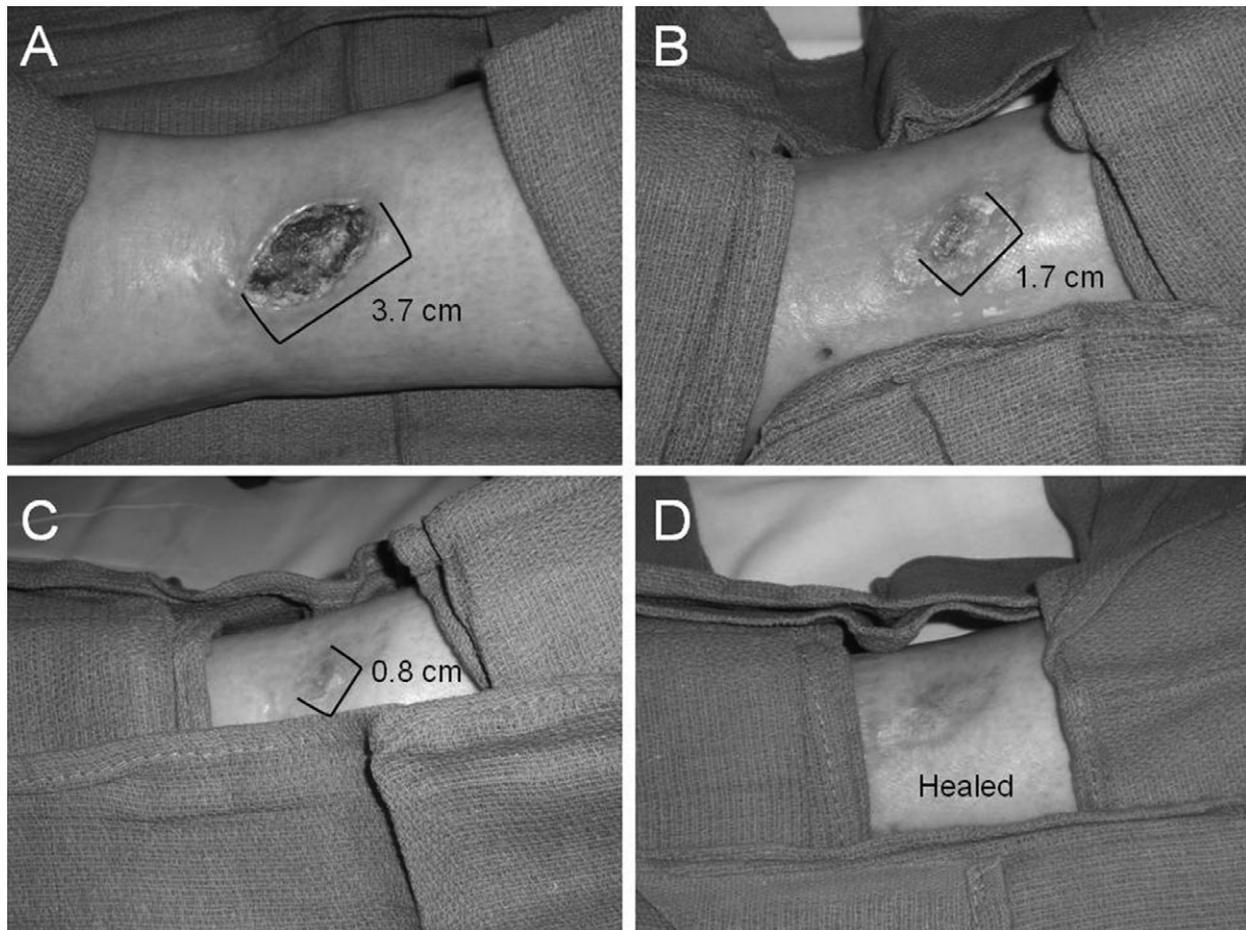


Figure 1 Photographs displaying ulcer size at different time points during healing: (A) prior to SCS implantation; (B) 1 month post-implantation; (C) 3 months post-implantation; (D) 4 months post-implantation.

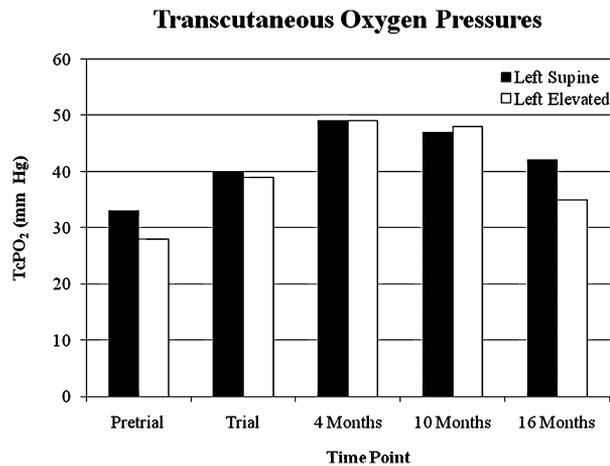


Figure 2 During the SCS trial stage, TcPO₂ increased in both the supine and 45° elevated positions. Following SCS, supine TcPO₂ of the lower left extremity was maintained at levels greater than 40 mm Hg for a period of 16 months.

supine and 45° elevated positions and revealed poor microcirculation in the left lower extremity (supine: 33 mm Hg, 45° elevated: 28 mm Hg, Figure 2).

Initially, the patient underwent a series of lumbar sympathetic blocks in conjunction with continued wound debridement. During the duration of the local anesthetic, the patient had substantial pain relief, evidence of vasodilation, and elevation in limb temperature. However, the ulcer neither healed nor decreased in size. The patient then underwent a SCS trial with a percutaneous octad lead with the tip placed at the superior portion of T10, left of midline. During the 5-day trial, the patient was instructed to keep the SCS on for 24 hours each day. During the trial, improvements in microcirculation occurred as demonstrated by 21% and 39% increases in supine and 45° elevated TcPO₂ values, respectively (Figure 2). She noted decreased cyanosis in her distal lower extremities.

Based on the improvements in TcPO₂ data and reported pain relief of greater than 60%, the patient progressed to the implantation of two octad leads and a rechargeable generator (Boston Scientific, Natick, MA, USA). At 4 months post-implantation, the patient's ulcer had healed (Figure 1). TcPO₂ monitoring documented improvements in microcirculation with a 48% increase in the supine value (Figure 2). The patient's global impression of change score displayed a selection of very much improved on a 7-point categorical scale with a range of "very much worse" to "very much improved." Her NRS decreased from 8/10 to 0/10. She discontinued her use of oral opioids, reported fewer Raynaud's episodes, and decreased sensitivity to cold ambient temperature in her lower extremities. The degree of pallor improved in her lower extremities. Supine and elevated TcPO₂ measurements were repeated at 16 months and demonstrated sustained improvement of

microcirculation at 27% and 25% above baseline, respectively. At 18 months post-implantation, the patient had not developed any ischemic wounds in her lower extremities and reported continued pain reduction (NRS 0/10). She utilized the stimulator for 24 hours each day.

Discussion

The vascular and inflammatory alterations associated with scleroderma involve small arteries, capillaries, and, particularly, arterioles. Initially, injury to the endothelial cells and basal lamina of blood vessels causes the intima to thicken and the lumen to narrow by as much as 75% to 80% [1]. Eventually, small vessels are completely obliterated. As this vascular disease progresses, microvascular beds in the skin diminish, resulting in localized areas of hypoxia, which are prone to tissue necrosis and in some cases, ultimately require amputations.

The results of medical and surgical management of the microvascular insufficiency associated with scleroderma are often disappointing. Sympathectomy for the treatment of Raynaud's phenomenon has provided mixed results [5–8]. de Trafford et al. reported the results of sympathectomies performed for 140 patients with Raynaud's phenomenon [8]. In this series, less than 20% had prolonged benefit, and 66% reported benefit lasting less than 1 year following the procedure.

By modulating microcirculation, SCS may be beneficial to individuals with scleroderma and non-healing wounds that are refractory to treatment. Although the exact mechanism of action for SCS on the microvascular system is unknown, several theories have been postulated and demonstrated in animal studies including modulation of the autonomic nervous system, activation of the descending inhibitory system, and antidromic activation of sensory nerves (A-delta and C fibers) and the subsequent release of vasodilators, such as calcitonin gene-related peptide and nitric oxide [9–11]. Calcitonin gene-related peptide is a potent microvascular vasodilator [11]. Improvement in oxygen levels with SCS may also reduce the proliferation of fibroblasts, assisting in the reduction of endothelial and skin damage [12].

Our patient had substantial clinical improvement with SCS treatment. A non-healing wound that had existed for 8 months prior to SCS treatment had healed by 4 months post-implantation and correlated with documented improvement in microcirculation indicated by increasing TcPO₂. One of the tenets of wound healing is improved blood flow. Since implantation, the patient's pain, opioid requirements, edema, and Raynaud's episodes have decreased. Furthermore, this improvement has continued. Literature regarding critical limb ischemia suggests that wounds with dimensions greater than 3 cm do not respond well to SCS treatment [4,13,14]. In this case, an ulcer 3.7 cm in length healed with the assistance of SCS.

Few reports exist on the utilization of SCS for the treatment of the microvascular abnormalities associated with

scleroderma and secondary Raynaud’s phenomenon. Only four reports to date have been published with variable ranges of follow-up and patient sample size (Table 1) [12,15–17]. Clinical improvements were reported in each case report. Three of the case reports included one patient each [15–17]. Francaviglia et al. reported the largest case series with 15 individuals with symptoms in their upper extremities [12]. Raynaud’s phenomenon was present in all patients for more than 10 years prior to SCS treatment. Four of the 15 individuals had ulcers, but wound size was not documented.

To our knowledge, this is the first case report to document the utilization of TcPO₂ monitoring in an individual with secondary Raynaud’s phenomenon to guide decision-making and to monitor microcirculatory function in the SCS trial and post-implantation stages. TcPO₂ monitoring in individuals with critical limb ischemia assists in determining the existence of a microcirculatory reserve that can be captured by SCS prior to implantation. The TcPO₂ data (baseline, delta, and final values) obtained during the trial have been shown to provide important prognostic implications for implantation decision-making [13,18–21]. Our patient exhibited normal ABI values, suggesting adequate macrocirculation, but had significantly low pretrial TcPO₂ values (<40 mm Hg), indicating poor microcirculation, suboptimal oxygen delivery, and inadequately perfused tissues [22]. During the 5-day trial, we were able to demonstrate improvement in TcPO₂ data, suggesting the existence of a microcirculatory reserve that could be captured with SCS treatment. Critical limb ischemia studies have suggested that a 20% increase or an absolute increase of at least 10 mm Hg correlate with a positive clinical outcome [13,18–21]. Here, the patient satisfied both of these criteria. Her improvement has been maintained at 18 months follow-up.

Conclusions

In summary, we demonstrate the healing of a greater than 3-cm refractory skin ulcer in an individual with normal macrocirculation but severe microcirculatory insufficiency from scleroderma and Raynaud’s phenomenon. Improvement in microcirculation, as indicated by TcPO₂ data, correlated with wound healing and significant decreases in pain. SCS may be considered in select individuals with scleroderma, who possess a microcirculatory reserve that can be captured with SCS treatment. Future large-scale randomized controlled trials with long-term follow-up are warranted for SCS in comparison to conservative treatment options for secondary Raynaud’s phenomenon and non-healing wounds associated with scleroderma. However, this could be challenging due to the relatively small patient populations associated with this condition. Long-term follow-up is also necessary to determine if improved microcirculation affects disease progression for this subset of patients. Furthermore, particular attention should be paid to the prognostic value of microcirculatory monitoring (e.g., TcPO₂ monitoring) in the trial stage to predict the outcome of SCS implantation.

Table 1 Summary of previous case reports

Authors	No. of Patients	Range of Follow-Up (years)	No. Patients with Prior Sympathectomies†	Microvascular Monitoring to Assess SCS Effects	Anatomical Location	SCS Lead Position	Outcome‡	Complications‡	Success Defined by Author‡
Fiume, D. [15]	1	1‡	0	None	Right leg	T9	PC, U	IR	S (100)
Francaviglia, N. et al. [12]	15	1 to 6	5 (33)	None	Upper extremity	C4 to C7	E(86), F(86), M(83), PC(83), RE(93), U(100)	R(7)	S (100)
Neuhauser, B. et al. [16]	1	1.5	1	CM, LDA	Bilateral hand	C5–C6	PC, RE, U	None	S (100)
Sibell, D. et al. [17]	1	>1	0	None	Upper extremity	C4	F, M, PC, U§	I	S (100)

† Values in parentheses indicate percent of patients.

‡ SCS removed at 3 months due to infection, CM = capillary microscopy; E = reduced edema; F = functional improvement; I = treated infection; IR = infection requiring SCS removal; LDA = laser Doppler anemometry; M = reduced pain medication; PC = pain control; R = lead replacement; RE = Raynaud’s episode improvement; S = successful treatment; U = ulcer healing.

§ Patient experienced some healing, non-healing ulcers required distal phalanx amputation.

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