Surgical management of the hand in scleroderma

Charles P. Melone Jr, MD*, James C. McLoughlin MD, SM,† Steven Beldner MD‡

Disabling deformity of the hand is a hallmark feature of the person afflicted with scleroderma. However, existing literature provides little guidance to operative treatment for the wide spectrum of hand derangement. Although arthrodesis is generally recommended for severe flexion contractures of the interphalangeal joints, other surgical procedures such as arthroplasty, excision of painful calcinosis, and digital sympathectomy have been employed sparingly, undoubtedly due to potentially hazardous soft tissue conditions. Based on experience with 70 scleroderma patients requiring 272 hand operations, this article provides further insight as to the role of surgical treatment for the scleroderma hand. The favorable results in this relatively large series of cases support the efficacy of precisely timed and skillfully executed surgery in the alleviation of pain, prevention of tissue loss, preservation of function, and improvement in aesthetics. For the ischemic tissues of the scleroderma hand the prerequisite for uncomplicated surgery is a tension-free wound, often requiring judicious skeletal shortening and healing by secondary intention. Curr Opin Rheumatol 1999, 11:514–520

*Chief, Division of Hand Surgery, Beth Israel Medical Center, New York, NY, USA; Professor of Clinical Orthopaedic Surgery, Albert Einstein College of Medicine, Yeshiva University, Bronx, NY, USA; †Chief Resident, Department of Orthopaedic Surgery, Montefiore Medical Center, Bronx, NY, USA; ‡Attending Hand Surgeon, Beth Israel Medical Center, New York, NY, USA

Correspondence to Charles P. Melone, Jr., MD, Division of Hand Surgery, Beth Israel Medical Center, 321 East 34th Street, New York, NY 10016, USA

Current Opinion in Rheumatology 1999, 11:514–520

Abbreviations

CREST syndrome	calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia
MP	metacarpophalangeal
PSS	progressive systemic sclerosis

ISSN 1040–8711

Scleroderma, or, as correctly termed, *progressive systemic sclerosis* (PSS), is one of the most disabling disease processes encountered by both rheumatologists and hand surgeons. Scleroderma has been categorized into specific types, such as PSS and CREST (calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia) syndrome, and this differentiation has a profound significance regarding visceral damage and ultimate prognosis [1]. Regardless of the type, scleroderma is primarily a disease of ischemic fibrosis, with the widespread deposition of collagen invariably affecting the hands at an early stage. The frequent result is extensive skin atrophy, subcutaneous fibrosis, subcutaneous calcinosis, vascular compromise with Raynaud's syndrome, painful ulcerations (often with infection), and the classical hand deformities comprising flexion contractures of the interphalangeal joints, extension contractures of the metacarpophalangeal (MP) joints, and adduction contracture of the thumb (see Figure 1). Rarely is a case reported in which the diagnosis of scleroderma is made without these characteristic features [2]. While surgery of the hand has assumed an increasing role in patient management, a basic premise must be emphasized: medical management must be optimal before consideration of operative treatment.

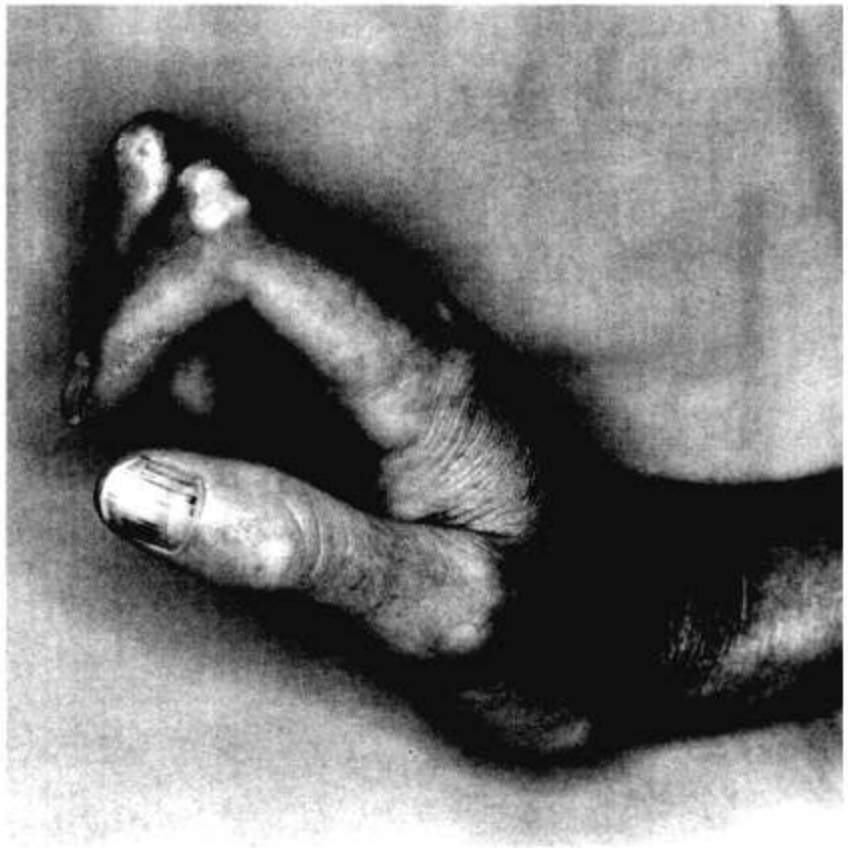
To assess the role of surgical treatment, one must carefully evaluate the type of scleroderma as well as the success of medical management, the potential surgical correction, the preoperative medical and surgical assessment, the types of surgery available, and the anticipated result. Goals for surgery should be realistic and specific, the principle objectives being alleviation of pain, preservation of hand function, prevention of ulceration, infection, and ultimately tissue loss, and enhancement of aesthetics.

Classification

Scleroderma has been categorized into two major clinical types and several less frequent variants. The two major types are classic (or diffuse) PSS and CREST syndrome. CREST syndrome refers to the constellation of calcinosis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly and telangiectasia, and is considered the more benign type of the two major clinical presentations. This syndrome often involves only the hands and face; visceral organ involvement may occur, but usually late in the disease process [3]. Surgical intervention is employed for both major types as each may severely compromise hand function.

Figure 1. Characteristic deformities of the hand in systemic scleroderma

Flexion contractures of the interphalangeal joints, extension contractures of the metacarpophalangeal joints, and adduction contracture of the thumb. Despite the guarded prognosis resulting from visceral involvement of the disease process, persons with optimal medical management often survive for many years. In such cases reconstructive hand surgery can prevent progressive tissue loss and considerably improve function and aesthetics.



When surgery is contemplated, the type of scleroderma is of critical importance in preoperative medical evaluation and treatment. Patients with PSS often have visceral changes early in the disease course, whereas these changes are usually delayed or absent in patients with the CREST form. Classic scleroderma often affects the lungs, kidneys, and gastrointestinal tract [3]; therefore, a complete medical evaluation, including analysis of both pulmonary and renal function, is essential before the administration of any anesthetic agent. Additional measures, which can be taken to formulate a complete assessment of the patient's disease process, include measurement of anticentromere and anti-Scl-70 antibodies. Severe peripheral ischemia is strongly and positively correlated with anticentromere antibodies, and a less significant association is seen with anti-Scl-70 antibodies. The presence of either of these antibodies suggests that the patient is "at risk of digital loss"[4]. The correlation between peripheral ischemia and the presence of anticardiolipin antibodies is less clear, due to conflicting study results [4,5]. Antiphospholipid antibodies are also positively correlated with severe peripheral ischemia in patients with PSS [6]. Each of these laboratory values can contribute to an increasingly accurate assessment of potential surgical risk.

General management considerations

Optimal treatment begins with comprehensive medical management of the scleroderma patient. Clearly, medical modalities should be the primary focus of treatment for scleroderma, and patients considered candidates for hand surgical treatment should preferably be managed by a rheumatologist. Only after complete rheumatologic evaluation and comprehensive medical treatment should these patients be referred to the hand surgeon for corrective surgery.

Critical to precise medical management is the differentiation between PSS and CREST syndrome, due to the variable visceral involvement. Surgical implications of this distinction relate primarily to the increased anesthetic risk for patients with PSS. In contrast, the two groups show no significant difference in hand changes such as ulcerations on fingers, sclerodactyly, and multiphasic digital color changes [7].

Strategies for medical management include the use of many behavior modification and pharmacologic methods.

The myriad causes of vascular disease, such as smoking and diabetes, are carefully evaluated, and, if present, treated. Calcium channel blockers, sympathetic blocks, and iloprost infusions can all be used to reduce afferent vascular tone and therefore increase digital blood flow. Other agents, such as aspirin and pentoxyifyllin, may improve microcirculation, and ketanserin improves venous outflow [8]. Each of these agents has obvious conceptual advantages, but usage has been limited by adverse side effects. For example, iloprost infusion has been useful in treating imminent gangrene and ischemic ulcers of the digits with a prolonged physiologic improvement; however, side effects such as headache, nausea, and flushing have limited patient compliance [9,10]. Sympathetic blocks, especially stellate ganglion blocks, have demonstrated increases in temperature and pulse wave amplitude in the extremities blocked; however, and unfortunately, paradoxical decreases in temperature have been reported on the unblocked sides [11]. Recombinant tissue plasminogen activator, a known thrombolytic agent, has also been used both in bolus and infusion form to treat the digital ischemia in sclerodactyly, because digital vessels often show thrombosis and intimal hyperplasia. The subjective effects of recombinant tissue plasminogen activator clearly outlive the objectively measured effects, and cost is a significant concern [12]. Thus its role in management remains unclear.

Other drugs that have been used include vasoactive drugs such as phenoxybenzamine and alpha-methyldopa; anti-inflammatory drugs such as salicylates, antimalarials, corticosteroids, and antimitotic agents have also been used [13]. From a surgical perspective, the critical aspect of initial management is evaluation by a rheumatologist well-versed in the pathophysiology and the treatment of this complex disease process.

Surgical management: basic concepts

The wide range of hand disorders encountered, coupled with the varying degree of underlying digital vascular ischemia and visceral damage contribute to the challenge in treating scleroderma patients.

Patients with PSS classically demonstrate progressive contractures, digital tuft resorption, sclerodactyly, subcutaneous calcinosis, joint space narrowing, juxta-articular demineralization, and ankylosis [14]. Resorption of the distal phalanges is considered the most common bony change radiographically; however, flexion contracture of the hand is the most frequent overall radiographic finding [15]. The spectrum of hand involvement is wide and intra-articular calcification [16] and even carpal synostosis have been reported [17]. The typical patient has proximal interphalangeal joint flexion contractures and MP extension contractures of the fingers, with an adducted thumb (Fig. 1). Digital ischemia, a characteristic feature of the patient with PSS, is an obvious concern of the hand surgeon. Ninety percent of these patients are apt to experience digital vasospasm on exposure to cold, even when there are no apparent signs of Raynaud's phenomenon [18]. Studies also suggest that this transient vasospasm is not limited to digital circulation, but has a more widespread systemic effect. This "Raynaud-like transient vasoconstrictive response to cold has been suggested from clinical studies of the cardiac, pulmonary and renal circulations" [8]. This phenomenon has profound implications on the surgical environment, which should be maintained at a relatively warm temperature.

Comprehensive preoperative medical evaluation also necessitates pulmonary function testing, electrocardiography, and urinalysis. Preoperative vascular analysis includes pulse volume recordings in conjunction with cold stress testing and digital nerve blockade. For example, if pulse volume recording waveforms are relatively flat, the patient's hands are immersed in warm water to stimulate vasodilation; the pulse volume recordings are then immediately repeated and compared with the prior result. If the waveforms improve significantly, digital sympathectomy should be considered for the ischemic digit. Similarly a digital nerve blockade that results in increased digital temperature and improved coloration also suggests a beneficial effect from digital sympathectomy. Although there is increased enthusiasm for magnetic resonance angiography, rarely is traditional angiography necessary for the evaluation of the scleroderma hand.

Preoperative planning extends to the operative facility. The operating room staff and the engineering facilities must be notified in advance regarding a patient with PSS. The previously discussed multi-organ sensitivity to cold must be recognized; and, as stated, the operating room should be maintained at a relatively warm temperature. Operative assistants and anesthesiologists must be fully cognizant of the special requirements of the patient with PSS and the need for a highly precise and expeditious procedure. Our postoperative results indicate that with thorough preoperative evaluation and skillful surgery, excellent clinical satisfaction is usually achieved with adherence to the aforementioned surgical caveats.

Surgical management: author's experience

Two hundred seventy-two procedures have been performed, including 211 interphalangeal joint arthrodeses, 28 MP implant arthroplasties, 2 thumb basal joint arthroplasties with concurrent thumb MP implant arthrodeses, 12 calcinosis excisions, and 10 digital sympathectomies. A tension-free wound closure was essential to success and for 30 cases no skin sutures were used. Operative parameters relatively unique to the scleroderma patient include specific incisions, types of

skeletal fixation, exacting fusion angles, wound management, and operating room preparation. With adherence to evolving surgical principles, favorable results can now be anticipated in the vast majority of cases [19,20].

Follow-up evaluation ranging from 1–15 years has substantiated this concept and revealed: 1) uncomplicated wound healing by primary or secondary intention; 2) radiographic union of arthrodesis within 8 weeks of surgery, corroborating the report of Jones *et al.* [21]; 3) a 50-degree average arc of motion for the MP implant arthroplasties; 4) improved vascularity with decreased pain, increased warmth, and healing of ulcers following digital sympathectomy, supporting the experiences of others [22–24]; 5) eradication of painful ulcers after excision of subcutaneous calcinosis; and 6) overall functional and aesthetic improvement.

Specific operations

Interphalangeal arthrodesis

See Figure 2. Interphalangeal arthrodesis has proved the mainstay of surgical treatment for the scleroderma hand, accounting for 78% of total operations. Success is contingent on the following key surgical principles:

1) regional (axillary or wrist) anesthesia block is preferred due to its vasodilatory effect;

2) the use of a tourniquet expedites precision surgery, but owing to concern for the precarious circulation should be carefully monitored and, of course, employed in an expeditious fashion. In fact, tourniquet time rarely exceeds 1 hour for the vast majority of procedures on the scleroderma hand;

3) closure of wound margins must be tension free, thereby enhancing primary or secondary healing;

4) flap undermining should be minimal, thereby avoiding damage to the fragile digital circulation;

5) successful arthrodesis requires correction of severe deformity often characterized by subluxation of the interphalangeal joints, thereby necessitating complete capsulotomy in conjunction with judicious skeletal shortening;

6) stable surgical fixation is essential and is best achieved by employing fine Kirschner wires in a crossed pattern;

7) following tourniquet deflation, the digit may demonstrate ischemia owing to tension on vascular structures. In such instances the transfixion wires must be promptly removed to permit restoration of circulation. Once vascular perfusion is evident, stabilization of the arthrodesis is achieved by reinserting the Kirschner wires, often with the operative joint placed in a more relaxed position.

Dorsal transverse incisions over the contracted, destroyed joint are now routinely used to enhance preservation of the critical circulation, to facilitate flap coverage, and to promote primary wound healing. The final position for the fused joint is determined on an individual basis, with the principal consideration being the preoperative status of the deformed and destroyed joint and the functional needs of the patient. Attesting to the high level of satisfaction with interphalangeal arthrodesis, patients with PSS have consistently requested these procedures bilaterally.

Metacarpophalangeal implant arthroplasty

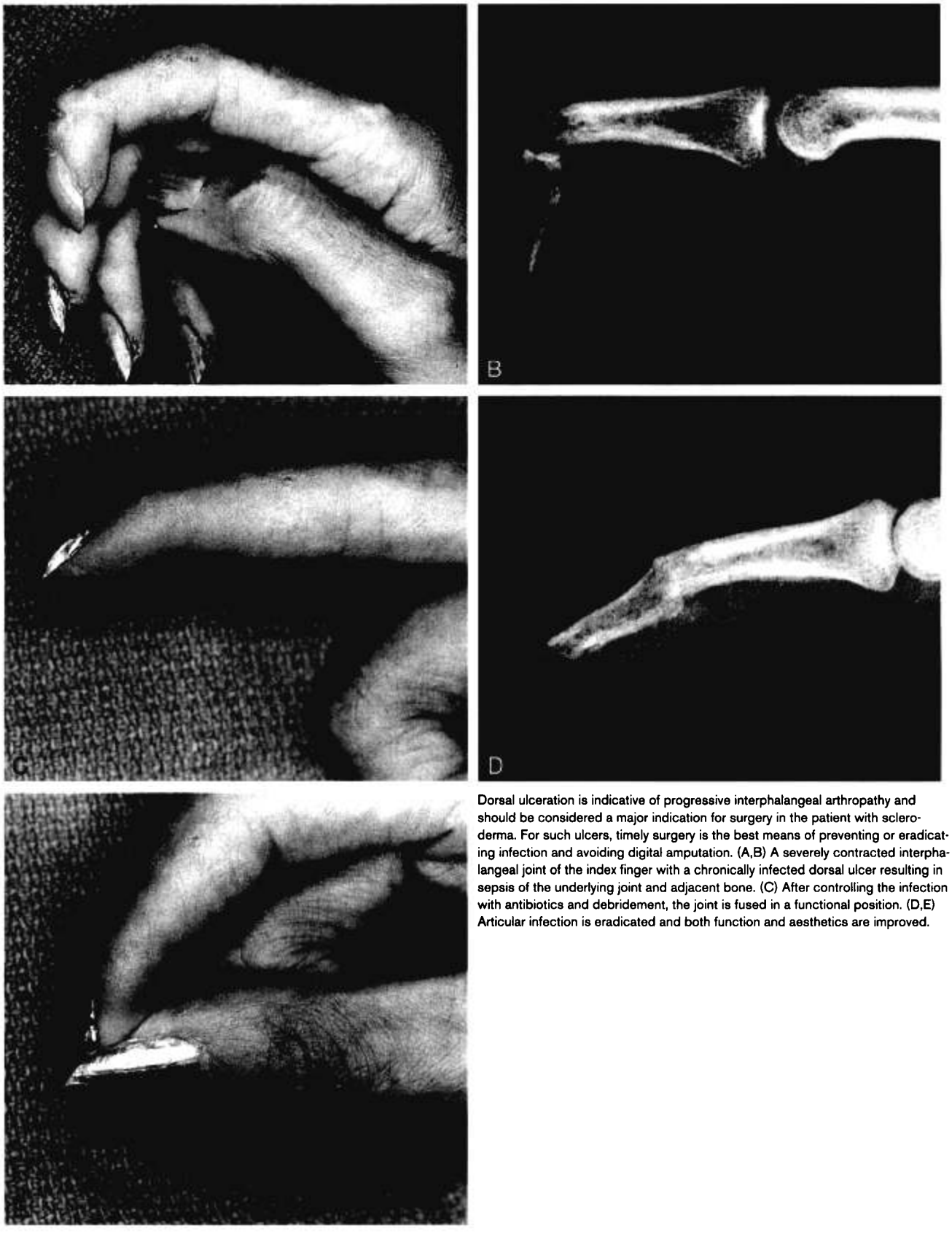
The destroyed MP joints of PSS are characterized by disabling and irreversible hyperextension contractures (Fig. 1) with narrowed joint spaces and flattened as well as moderately eburnated cartilaginous surfaces. In contrast to other arthropathies, such as rheumatoid arthritis, erosive changes and proliferative synovitis usually are not encountered. In the scleroderma (PSS) patient, a transverse incision with minimal soft tissue reflection and preservation of the critical dorsal veins is employed to approach all four finger MP joints. In contrast to MP resection arthroplasty for other conditions, no attempt is made to preserve the collateral ligaments; rather, a wide resection of the metacarpal head and condyles, including the collateral ligaments, is necessary to correct the deformity and to achieve relative lengthening of the chronically contracted extensor and flexor tendons. In our experience with these severe extension contractures, restoration of highly functional tendon excursion and joint mobility always requires substantial shortening of the metacarpal. The relatively generous skeletal resection also facilitates a tension-free primary wound closure.

To date, the outcome has been extremely gratifying to both surgeon and patient, and, significantly, on follow-up as long as 15 years, no adverse reaction to the implant has been noted.

Thumb basal joint arthroplasty

Progressive adduction contracture of the thumb is a disabling feature of PSS, owing to a resultant and severe articular imbalance of radial subluxation of the thumb carpometacarpal joint and fixed hyperextension deformity of the thumb MP joint (Fig. 1). In the thumb deranged by this dysfunctional zig-zag configuration, surgery comprises trapezium excisional arthroplasty and MP arthrodesis in mild flexion, thereby permitting realignment of the metacarpal in abduction and restoration of articular balance. No longer are silicone implants used as joint spacers; an improved joint is restored by

Figure 2. Disabling flexion contracture of index finger, typical of patient with scleroderma, surgically corrected by arthrodesis



Dorsal ulceration is indicative of progressive interphalangeal arthropathy and should be considered a major indication for surgery in the patient with scleroderma. For such ulcers, timely surgery is the best means of preventing or eradicating infection and avoiding digital amputation. (A,B) A severely contracted interphalangeal joint of the index finger with a chronically infected dorsal ulcer resulting in sepsis of the underlying joint and adjacent bone. (C) After controlling the infection with antibiotics and debridement, the joint is fused in a functional position. (D,E) Articular infection is eradicated and both function and aesthetics are improved.

excising the deformed trapezium coupled with reconstructing ligament restraints. This creates a highly mobile yet stable basal joint without the need for silicone or other foreign substances.

Digital sympathectomy

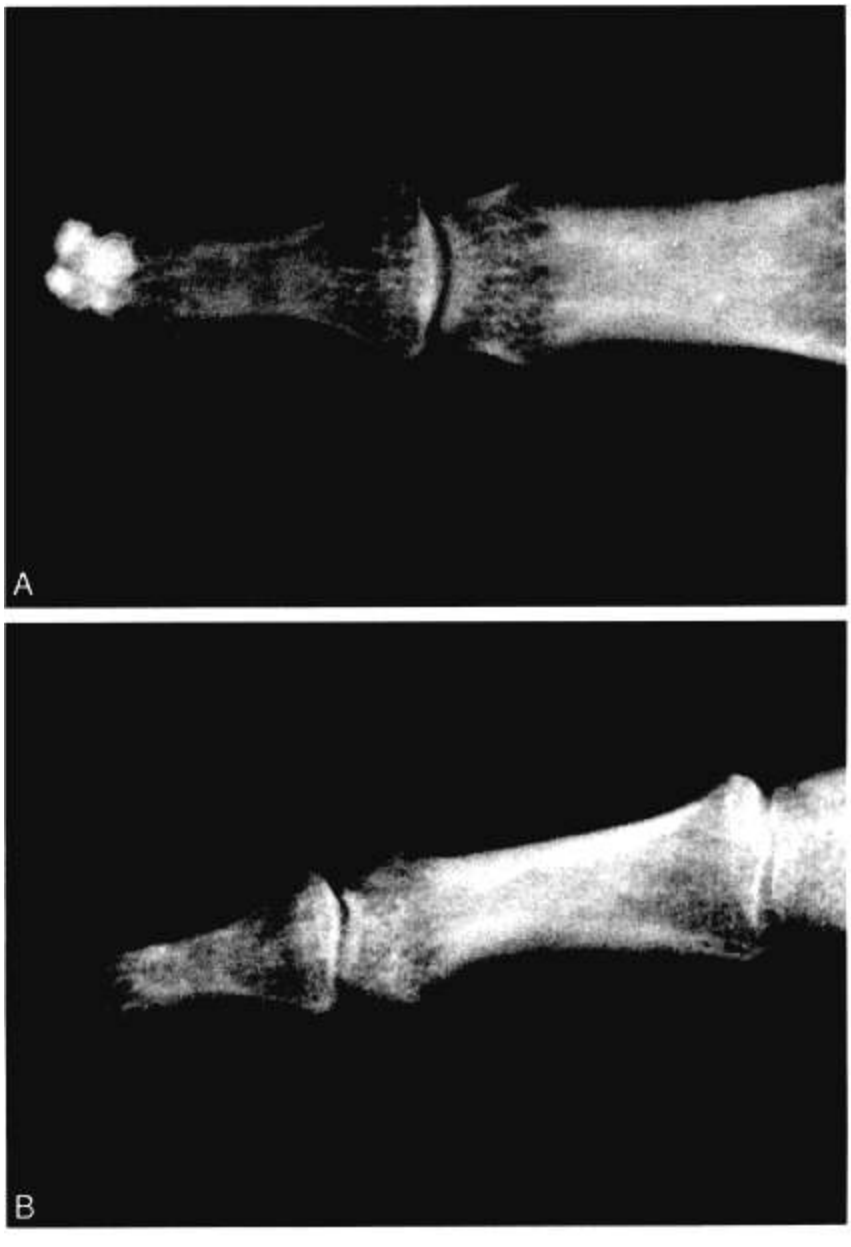
Owing to the prevalence of Raynaud's syndrome with its painful vascular compromise, digital artery sympathectomy has received increasing enthusiasm in the treatment of the scleroderma hand. As stated, if increased perfusion occurs following digital nerve blockage, a chemical sympathectomy has been achieved and the efficacy of surgical digital sympathectomy is supported. In carefully selected patients, decreased pain, increased warmth, and healing of ulcers can be anticipated.

Flatt [23], in the first report of successful sympathectomy of the common and proper digital arteries, eliminated the sympathetic vasospasm by first separating the digital nerve from the digital artery, and then by stripping the arterial adventitia for 2–4 mm. Wilgis [22], modifying this technique, extended the adventitial stripping for 2 centimeters along the common and proper digital arteries.

Jones [24] emphasized that chronic digital ischemia results not only from increased sympathetic vasocon-

Figure 3. Painful calcinosis, typical of patient with CREST syndrome, eradicated by surgical excision

(A) Typical subcutaneous calcinosis of scleroderma fingertip resulting in recurrent, painful skin ulceration with infection. (B) Careful delineation and excision of the offending calcium deposit eradicated the infected ulcer, induced rapid wound healing, and restored a highly functional finger. Radiograph obtained 3 years after surgery reveals no evidence of calcinosis recurrence.



striction but also from external compression owing to periadventitial fibrosis of the digital vessels. He attributed success of digital sympathectomy to eliminating both sympathetic vasospasm and external compression of the typical pathologic fibrosis. A thorough digital sympathectomy thus entails excision of all perivascular fibrosis, separation of the digital nerve from the artery with division of the three to four characteristic communication sympathetic fibers, and a meticulous adventitial stripping for a distance of at least two centimeters.

Despite the success of digital sympathectomy reported herein, one must be aware that an initial favorable result may later be compromised by progressive occlusive vessel disease, apt to occur in the scleroderma patient. However, one should also recognize that even temporary success may be of enormous benefit in the prevention of progressive ulceration, infection, and ultimately digital amputation.

Excision of subcutaneous calcinosis

Digital calcinosis is a prominent feature of scleroderma and, as the *C* of the acronym CREST implies, is most frequently encountered in patients with this syndrome. Two types of calcinosis are commonly visualized on radiographs: a relatively well-defined deposit localized to the tuft of the fingertip (Fig. 3), and a more diffuse form, often extending the length of the involved digit. In both instances the characteristic result is recurrent, painful skin ulceration with infection and serious dysfunction. Surgical treatment is aimed at excising the offending deposit, thereby decompressing the digit, eliminating the painful ulcer, and promoting prompt wound healing. For the localized form, the excision often completely eradicates the calcinosis and recurrence is a rare observation. In contrast, if the digit is afflicted with diffuse calcinosis, a debulking procedure is performed wherein only the major deposit causing the painful ulceration is excised. In the latter situation, overzealous attempts to remove all calcinosis are apt to compromise digital viability and should be avoided.

Conclusions

Hand surgery for the patient with scleroderma has proved consistently successful in alleviating pain, improving function, and enhancing aesthetics. Clearly, carefully planned and precisely performed operative treatment should be considered an integral component of the overall management of the scleroderma patient. Based on a high degree of success coupled with a high level of patient satisfaction, the traditional, but ill-founded, concept that surgery is a last resort and high-risk treatment option should be abandoned.

References

1 LeRoy EC, Black C, Fleischmajer R, Jablonska S, Krieg T, Medsger TA, Rowell N, Wollheim F: Scleroderma (systemic sclerosis): classification, subsets and pathogenesis. J Rheumatol 1988, 15:202–205.

2 Barry M, Katz L, Cooney L: An unusual articular presentation of progressive systemic sclerosis. Arthritis Rheum 1983, 26:1041–1043.

3 Cotran RS, Kumar V, Robbins SL: Diseases of Immunity. In Robbins Pathologic Basis of Disease, ed 4. Philadelphia: Saunders. 1989:204–207.

4 Herrick AL, Heaney M, Hollis S, Jayson MIV: Anticardiolipin, anticentromere and anti-Scl-70 antibodies in patients with systemic sclerosis and severe digital ischaemia. Ann Rheum Dis 1994; 53:540–542.

5 Wigley FM, Wise RA, Miller R, Needleman BW, Spence RJ: Anticentromere antibody as a predictor of digital ischemic loss in patients with systemic sclerosis. Arthritis Rheum 1992, 35:688–692.

6 Lewandowski K, Turowiecka Z, Puszczewicz J, Mackiewicz S, Zawilska K: Antiphospholipid antibodies and disturbances of phospholipid dependent coagulation tests in patients with systemic sclerosis and severe peripheral ischaemia. Thromb Res 1996, 81:693–695.

7 Velayos EE, Masi AT, Stevens MB, Shulman LE: The 'CREST' syndrome comparison with systemic sclerosis (scleroderma). Arch Intern Med 1979, 139:1240–1244.

8 Seibold JR: Critical tissue ischaemia in scleroderma: a note of caution. Ann Rheum Dis 1994, 53:289–290.

9 Zachariae H, Hackier-Sorensen L, Bjerring P, Heickendorff L: Treatment of ischaemic digital ulcers and prevention of gangrene with intravenous iloprost in systemic sclerosis. Acta Derm Venereol 1996, 76:236–238.

10 Wigley FM, Seibold JR, Wise RA, McCluskey DA, Dole WP: Intravenous iloprost treatment of Raynaud's phenomenon and ischemic ulcers secondary to systemic sclerosis. J Rheum 1992, 19:1407–1414.

11 Omote K, Kawamata M, Namiki A: Adverse effects of stellate ganglion block on Raynaud's phenomenon associated with progressive systemic sclerosis. Anesth Analg 1993, 77:1057–1060.

12 Klimiuk PS, Kay EA, Illingworth KJ, Gush RJ, Taylor LJ, Baker RD, Perkins C, Jayson MIV: A double-blind placebo-controlled trial of recombinant tissue plasminogen activator in the treatment of digital ischemia in systemic sclerosis. J Rheum 1992, 19:716–720.

13 Winkelmann RK, Kierland RR, Perry HO, Muller SA, Sams WM: Management of scleroderma. Mayo Clin Proc 1971, 46:128–133.

14 Blocka KLN, Bassett LW, Furst PE, Clements PJ, Paulus HE: The arthropathy of advanced systemic sclerosis. Arthritis Rheum 1981, 24:874–882.

15 Bassett LW, Blocka KLN, Furst DE, Clements PJ, Gold RH: Skeletal findings in progressive systemic sclerosis (scleroderma.) AJR 1981, 136:1121–1126.

16 Resnick D, Scavulli JF, Georgen TG, Genant HK, Niwayama G: Intra-articular calcification in scleroderma. Radiology 1977, 124:685–688.

17 Bjersand AJ: New bone formation and carpal synostosis in scleroderma. Am J Roentgenol 1968, 103:616–619.

18 Grassi W, Core P, Carlino G, Cervini C: Effects of peripheral cold exposure on microvascular dynamics in systemic sclerosis. Arthritis Rheum 1994, 37:384–390.

19 Ward WA, Moore AV: Management of finger ulcers in scleroderma. J Hand Surg 1995, 20A:868–872.

20 Norris RW, Brown HG: The proximal interphalangeal joint in systemic sclerosis and its surgical management. Br J Plastic Surg 1985, 38:526–531.

21 Jones NF, Imbriglia JE, Steen VD, Mesger TA: Surgery for scleroderma of the hand. J Hand Surg 1987, 12A:391–400.

22 Wilgis EFS: Digital sympathectomy for vascular insufficiency. Hand Clinics 1985, 1:361–367.

23 Flatt AE: Digital artery sympathectomy. J Hand Surg 1980, 5:550–556.

24 Jones, NF: Acute and chronic ischemia of the hand: pathophysiology, treatment, and prognosis. J Hand Surg 1991, 164:1074.