Nodular Scleroderma: Case Report and Literature Review

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ABSTRACT. ***Objective.*** To describe a unique case of scleroderma (SSc) presenting as multiple keloidal nodules and early-onset osteoarthritis (OA), and to summarize the clinical and serological data for 13 similar patients reported in the English literature since 1966.

Methods. MEDLINE review of the literature over a 35-year period (1966-2002) revealed 13 cases of nodular SSc. We describe a case of nodular SSc in a 40-year-old African-American male with localized SSc who developed progressive skin thickening and keloidal nodules on the arms, hands, chest, abdomen, and thighs with advanced osteoarthritis of the hips.

Results. In all 14 cases, diagnosis was made based on skin biopsy and evidence of keloid (nodule) formation. Ten cases occurred in women and 4 in men, with ages ranging from 9 to 66 years and a mean age of 38.9 years. The ethnicity of the patients was given in only 5 of the 13 previously reported cases. Including our patient, 4 were of African descent, and 2 were Caucasian. Most patients had symptoms of SSc consisting of arthralgias (n = 10), sclerodactyly (n = 9), Raynaud's phenomenon (n = 8), digital pitting and/or calcinosis (n = 5), shortness of breath with pulmonary fibrosis (n = 5) or pulmonary hypertension (n = 1), dysphagia or reflux (n = 3), renal disease (n =3), and elevated erythrocyte sedimentation rate (n = 3).

Conclusion. Nodular SSc is a rare variant that presents with lesions that clinically resemble keloids. OA, as documented in the present case, does not appear to be a typical feature of nodular SSc. (J Rheumatol 2003;30:2500–2)

Key Indexing Terms:
NODULAR KELOID SCLERODERMA SYSTEMIC SCLEROSIS ARTHRITIS

Nodular scleroderma (SSc) is considered a rare variant of the disease[1-4] that may occur in the setting of either systemic sclerosis[1-7] or localized scleroderma (morphea)[8-12]. We describe a 40-year-old African-American man with localized SSc presenting as multiple keloidal nodules. Our patient's disease was complicated by severe early onset osteoarthritis (OA) affecting the hips. Although scleroderma has been associated with rheumatoid arthritis[13,14], this is the first reported case of nodular SSc associated with advanced OA. The etiology of nodule formation in SSc is unknown.

MATERIALS AND METHODS

Literature review. A literature review using the MEDLINE key words scleroderma, morphea, nodular, and keloid produced 13 other cases of nodular SSc in the English literature over a 35-year period (1966 to 2002)[1-12]. All patients exhibited characteristic nodular lesions but the degree of keloid formation varied. Diagnosis of SSc and keloid formation was based on histological confirmation and/or examination by a dermatologist. Clinical and serological data from all 14 cases are summarized in Table 1.

Case report. A 40-year-old African American male with a medical history of hypertension presented after noticing the slow onset of focally thickened skin over the thorax, arms, and fingers. There was no prior history of keloids or abnormal scarring. Review of systems disclosed a 2-year history of worsening hand and knee arthralgias and lower back pain. There was no morning stiffness, oral ulcers, visual changes, dysphagia, reflux, alopecia, dyspnea, or symptoms of Raynaud's phenomenon. Medications included amlodipine 5 mg daily for hypertension. His family history was negative for connective tissue disease or keloid formation.

Physical examination revealed indurated skin on the forehead, scalp, thorax, arms, and digits. In addition, there were discrete, irregular, hypertrophic plaques resembling keloids localized to the arms and thorax (Figure 1). Salt-and-pepper pigmentary changes of the scalp and forehead were noted. His palmar skin displayed contractures that resulted in atrophy of the skin at the distal phalanges. He also had induration and keloidal nodules on the thenar and hypothenar skin. He had pain with rotation and flexion of his hips. The remaining components of his examination were unremarkable.

Laboratory evaluation revealed normal complete blood count and differential, liver function tests, chemistry profile, and erythrocyte sedimentation rate, as well as negative antinuclear antibody, extractable nuclear antigen screen, and anti-topoisomerase I (Scl-70) antibodies. Pulmonary function testing, echocardiogram, and nailfold capillary microscopy were normal. Radiographs revealed advanced osteoarthritic changes involving both hips. A biopsy of indurated skin from the inner surface of his right arm showed a minimal superficial perivascular inflammatory cell infiltrate and fibrosis within the dermis consistent with SSc. A later biopsy of skin from the ventral surface of his right arm showed the broad, brightly eosinophilic collagen bundles typical of a keloid.

DISCUSSION

The terms nodular SSc, keloidal SSC, pseudokeloidal SSc, keloid-like morphea, and nodular morphea have all been used to describe patients with features of either localized or systemic SSc who present with hypertrophic plaques that may clinically resemble keloids[1-12]. Although keloids are

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Table 1. Clinical and serological features of patients with nodular SSc.

Author Reference, year	Sex	Age*	Areas of Skin Involvement	Type of Scleroderma	Symptoms	Positive Laboratory Tests
Cantell[3], 1980	M	66	Trunk, neck, arms	Systemic	Sclerodactyly, RP, arthralgias, renal involvement	BUN 27 mg/dl
James[1], 1984	M	24	Arms, thighs	Systemic	Sclerodactyly, RP, arthralgias, pulmonary involvement, esophageal dysfunction	ANA, nucleolar
	F	17	Chest, back	Systemic	Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement	ANA, speckled
Sasaki[7], 1992	F	66	Abdomen	Systemic	Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement, esophageal dysfunction	ANA, speckled, RF
Wilson[4], 1992	F	42	Chest, abdomen, arms	Systemic	Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement	ANA, speckled
Yamamoto[6], 1994	F	36	Chest	Systemic	Sclerodactyly, RP, renal and pulmonary involvement	ANA, speckled, anti-Scl-70 ESR, antiRNP, CrCL, 27ml/min
Krell[2], 1995	F	40	Trunk, neck, arms	Systemic	Sclerodactyly, RP, arthralgias	ESR, BUN 41mg/dl, ANA, speckled
Mizutani[5], 1995	F	42	Chest	Systemic	Sclerodactyly, RP, arthralgias, calcinosis, pulmonary involvement	ANA, speckled, anti-Scl70
Akintewe[8], 1985	F	17	Trunk, neck, arms	Localized	Sclerodactyly, arthralgias	None
Micalizzi[10], 1994	F	64	Arms, breast, abdomen, back	Localized	Esophageal dysfunction	ANA, speckled
Stephanato[9], 1992	M	20	Suprascapular, supraclavicular	Localized	None	ESR
Kubo[11], 1997	F	61	Neck, chest, back, face	Localized	None	None
Hsu[12], 1999	F	9	Right arm, right axilla, right chest	Localized	Arthralgias	Not reported
Cannick, 2003	M	40	Arms, chest, abdomen, thighs	Localized	Arthralgias	None

* Age of presentation with keloidal nodules. RP: Raynaud's phenomenon; ANA: antinuclear antibody; RF: rheumatoid factor; BUN: blood urea nitrogen; CrCL: creatinine clearance; ESR: erthrocyte sedimentation rate; RNP: ribonucleoprotein; Scl70: scleroderma 70.

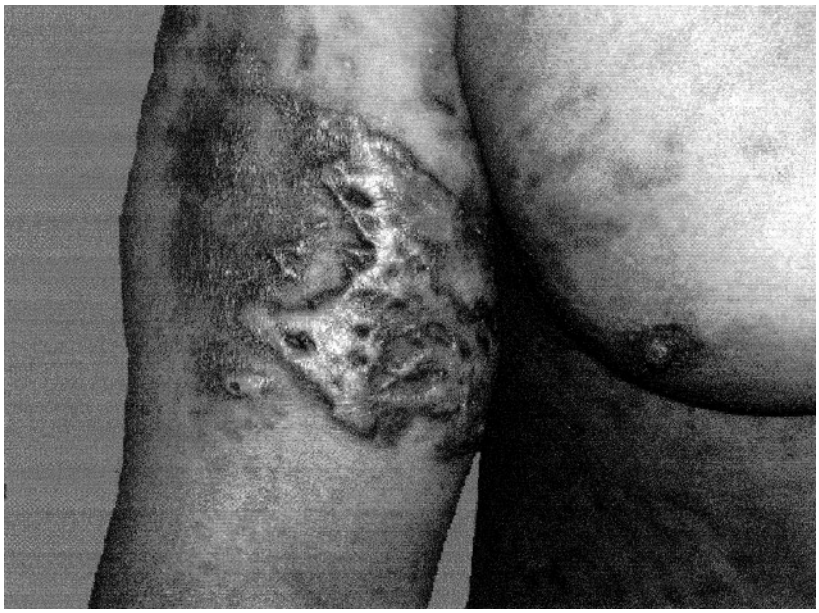

Figure 1. Photograph shows a large keloid on the ventral arm. In addition, there are numerous flat indurated, hyperpigmented plaques on the lateral chest and medial surface of the arm. These lateral lesions are typical of localized scleroderma (morphea).

well-known phenomena primarily in individuals of African descent, their formation in SSc is rare. In patients with nodular SSc, keloids may represent an aberrant response to the early inflammatory phase of the disease[2].

Our case report is similar to other reported cases of patients with SSc who later developed keloids or nodules. The distribution of the lesions primarily on the arms and upper torso in our patient is also similar to other case reports. However, none of the clinical photographs in the literature so far show the dramatic keloid formation observed in our patient.

To date, our patient with nodular SSc is the only one described with symptoms and radiographic evidence of advanced OA. It is our opinion that whatever autoimmune process is driving his SSc is also driving his OA. The lack of symptoms of esophageal dysmotility, overt Raynaud's phenomenon, sclerodactyly, and telangiectasias suggests that our patient has localized rather than systemic SSc. His condition progressed rapidly despite treatment with hydroxychloroquine and methotrexate.

Nodular SSc is a rare disorder affecting a minority of patients with SSc. Comparisons between cases are confounded by the variability in terminology, presentation, symptoms, and appearance of the lesions. The pathogenesis is still undetermined, but may involve a combination of environmental and genetic factors.

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