Lipodermatosclerosis: A clinicopathological study of 25 cases

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Background: Lipodermatosclerosis is a chronic fibrosing process that involves both lower legs and classically affects Caucasian women in their 60s.

Objective: The objective is to define the histologic spectrum seen this condition.

Methods: A total of 25 cases were collected prospectively and all were stained with hematoxylin-eosin, von Kossa, and Verhoeff-van Gieson.

Results: Patient age ranged from 33 to 84 years (mean age 62.6 years). The female to male ratio was approximately 12 to 1. All lesions were present on the lower extremities, between the knee and ankle. Lesion duration ranged from 2 months to 2 years (mean 9.5 months). Clinically, the lesions were erythematous, tender, indurated plaques or nodules. The characteristic histologic findings were seen almost exclusively in the subcutaneous tissue, involving primarily the lobules but also the septa. Adipose changes included micropseudocyst and macropseudocyst formation, necrotic adipocytes, lipomembranous change, and lipogranulomas with xanthomatous macrophages. The lesions were largely devoid of dermal, septal, or lobular neutrophilic or lymphocytic inflammation. Medium vessel calcification was seen in 13 cases. The accumulation of basophilic elastic fibers located deep in the septa was present in all the cases. These fibers had a moth-eaten appearance and resembled pseudoxanthoma elasticum. In 21 of 25 cases, these fibers were positive with both the von Kossa and Verhoeff-van Gieson stains.

Limitations: The cases in the series were not compared with age-, sex-, and location-matched biopsy specimens or biopsy specimens of other fibrosing conditions.

Conclusion: This constellation of histologic changes is diagnostic of lipodermatosclerosis. The presence of pseudoxanthoma elasticum–like changes is a helpful, but not unequivocal, clue in this condition. (J Am Acad Dermatol 2010;62:1005-12.)

Key words: hypodermitis sclerodermiformis; lipodermatosclerosis; pseudoxanthoma elasticum; sclerosing panniculitis; venous insufficiency.

Lipodermatosclerosis, originally named "hypodermitis sclerodermiformis" and subsequently termed "sclerosing panniculitis," among others, is a chronic fibrosing condition that is most commonly seen in obese Caucasian women in their 60s. Affected individuals are often smokers and typically have underlying vascular disease, ranging from venous insufficiency to arterial ischemia. Although the cause may be multifactorial, the end result is constant and is characterized by painful indurated hyperpigmented plaques on the lower extremities. Despite the consistent and well-documented clinical findings, the microscopic changes have not been extensively detailed in the literature.[1]

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METHODS

The cases were collected prospectively from our dermatopathology practice between September 1998 and December 2005. Multiple cuts of standard

hematoxylin-eosin–stained sections were examined. Von Kossa and Verhoeff-van Gieson stains were performed on all cases.

Patient demographics, lesion characteristics, and differential diagnoses were gathered from the submitted specimen reports and treating physicians.

RESULTS

The patients ranged in age from 33 to 84 years, with a mean age of 62.6 years. Of the 25 cases, there were 23 women and two men (female: male ratio of approximately 12:1).

All lesions were present bilaterally on the lower extremities with site specificity as follows: lower leg (15), ankle (6), calf (3), and shin (1). The known duration of the lesions ranged from 2 months to 2 years, with a mean of 9.5 months. In 14 cases, lesion duration was not available.

Clinically, the lesions were described as warm and tender erythematous indurated, often hyperpigmented, hard woody plaques and nodules (Fig 1).

The clinical differential diagnosis was somewhat limited, with lipodermatosclerosis being the most common (9). Other diagnostic considerations, in order of frequency, were erythema nodosum (8), panniculitis (6), stasis dermatitis (6), morphea (3), vasculitis (3), and one each of necrobiosis lipoidica, atrophie blanche, diabetic dermopathy, lymphedema, and squamous cell carcinoma. In 5 cases, there were no clinical differential diagnoses provided (Table I).

All specimens included the subcutaneous adipose tissue, with the type of procedure varying from punch, incisional, or wedge biopsies to elliptical excisions. On gross pathological examination, the specimens ranged in size from $0.3 \times 0.3 \times 0.4$ cm to $1.8 \times 0.6 \times 0.9$ cm; the average dimensions were $0.6 \times 0.4 \times 0.7$ cm.

The histopathologic findings are summarized in Table II. Microscopically, there was partial replacement of the subcutaneous adipose tissue by fibrosis with widening of the septa. The adipose lobules appeared both expanded and atrophic (Fig 2, *A*). The impression of expansion resulted from variably sized cystlike cavities, both large and small, within the adipose tissue (Fig 2, *B*). Many of the pseudocysts had an eosinophilic scalloped feathery lining (Fig 2, *C*). Active formation of this crenulated membrane was demonstrated by the presence of multinucleated giant cells acting on the necrotic adipocyte. The atrophic appearance was a result of fat necrosis and adipocyte dropout. Collections of foamy macrophages were present, some forming lipogranulomas (Fig 2, *D*). There was minimal, if any, chronic inflammation within the lobules, consisting mainly of lymphocytes, plasma cells, and few eosinophils. Vasculitis was not seen. Several cases showed some degree of stasis within the superficial capillary plexus of the papillary dermis; otherwise, the dermis was relatively uninvolved.

CAPSULE SUMMARY

- Consistent histologic findings of lipodermatosclerosis include lobular and septal involvement of the subcutaneous tissue, fat necrosis, macropseudocyst and micropseudocyst formation, and calcification of adipocytes.
- Lipomembranes are classic but not definitive for the diagnosis.
- Pseudoxanthoma elasticum-like septal elastosis with calcification is a consistent and helpful microscopic clue.

Verhoeff-van Gieson stains highlighted the presence of abundant short frayed elastic tissue within the septa, which could be seen as basophilic fibers on hematoxylin-eosin stain (Fig 3). Calcification of the abnormal elastic fibers was confirmed by positive staining with von Kossa, in a pattern resembling pseudoxanthoma elasticum (PXE) (Fig 4, *A* and *B*). In addition to calcification of septal elastosis, there was also variable calcium deposition within the intima and media of medium blood vessels, in the interstitium, in the walls of pseudocysts, and surrounding adipocytes (Fig 4, *C* and *D*). Soft tissue ossification was noted in two cases (8%).

DISCUSSION

Lipodermatosclerosis is a disease process that clinically appears as erythematous indurated hyperpigmented woody plaques occurring on the bilateral lower extremities. Numerous other names have been applied and proposed for this condition. "Hypodermitis sclerodermiformis" was the first to appear in the literature in 1955 as reported by Huriez et al,[2] who described a scleroderma-like hardening of the legs thought to result from infection of venous ulcers. "Liposclerosis" was the name given by Browse et al[3] in 1977 to the condition in which patients with severe varicose veins developed thickening, pigmentation, induration, and ultimately eczema and ulcers in the skin of the lower medial third of the legs. In 1980, Burnand et al[4] used the term "venous lipodermatosclerosis," which they defined as "liposclerosis; the changes in the skin and subcutaneous changes commonly known as 'postphlebitic leg.'" "Sclerosing panniculitis" was proposed by Jorizzo et al[5] in 1991 in an effort to unify the designations of "hypodermitis