LIPODERMATOSCLEROSIS PRESENTATION OF 3 CASES

Mohammed Mohy El Din Selim, MD
Amna Mohamed Mohamed Sharif, MRCP, DDs
Fahad Abdullah Ibrahim, MD
Adel Kamal Abdul Rahman, MD
Department of Dermatology
Hamad Medical Corporation

INTRODUCTION:

The clinical entity described as hypodermatitis sclerodermaformis(1), sclerosing panniculitis(2) and lipodermatosclerosis(3) perhaps refer to one condition appearing in different stages(4).

The chronic stage of all these entities is characterized by skin induration and scleroderma like hardening of legs usually affecting middle aged patients mostly obese females who suffer from venous insufficiency. The indurated plaques have irregular sharp border and vary in size and location and feel woody hard on palpation and can be differentiated from localized scleroderma and scleromyxedema.

Other accompanying skin changes may include erythema, telangiectasia, varicosities, oedema and mottled hyperpigmentation, scars from healed ulcers, stasis ulcers and stasis dermatitis.

The acute phase of lipodermatosclerosis may be unnoticed by the patient and may occur without obvious signs of venous disease(5). In the acute phase there is erythema, pain and discomfort at the site affected with no preceding illness or trauma. The commonest site affected is the medial leg and rarely the lateral leg. The acute stage is either undiagnosed or misdiagnosed as persistant cellulitis, stasis dermatitis, phlebitis, erythema induratum, erythema nodosum or nodular vasculitis. Biopsy is usually unnecessary. Kirrner et al(6) do not recommend doing biopsy if clinically lipodermatosclerosis is obvious because of the possibility of development of chronic ulcer in 50% at biopsy sites. If biopsy is necessary it should be performed at the edge of the erythematous indurated area and the wound should be closed primarily after a thin longitudinal incision.

First Case

MRH a male patient 50 years old. He presented with a one year duration of red hard indurated swelling affecting both legs but mainly the lateral part of the left leg (Fig 1&2). The condition began with malaise, pain, tenderness and redness of both legs. His condition was diagnosed as cellulitis and was treated with systemic courses of specific antibiotic (Velosef + Keflex for one month) with no response.

The patient is obese and had no systemic disorder or varicose veins. He had hyperkeratotic palmoplantar psoriasis and plaque psoriasis in front of both knees. Both legs were swollen showing patches of dark red hard indurated skin on lateral sides. The induration became stationary and the erythema is persistent for nearly one year.

Correspondence:
Dr. M. M. Selim
Dept. of Dermatology and Venereology
Hamad Medical Corporation
P. O. Box 3050 - Doha - Qatar

Fig 1: Red indurated swelling on the lateral part of the left leg
Fig 2: Indurated plaques on the right leg
All investigations were normal and included CBC, ESR, blood biochemistry, kidney function, liver function, serum amylase, serum lipase, serum fibrinogen and fibrinogen degradation products, and no findings suggestive of stasis dermatitis.

**Second Case**

A 57 year old Qatari man, mentally handicapped, presented with two months history of pain, discomfort and skin changes on both lower limbs.

Examination revealed erythematous, well demarcated, hyperpigmented induration on the medial side of both legs. Skin felt hard. No ulceration but minor superficial varicose veins were present (Fig 3 & 4). Clinical diagnosis of lipodermatosclerosis was made.

**Third Case**

H.A. is a 29 years old male Qatari patient, presented with red sclerotic nodules on inner side of the lower third of both legs. The condition began one year back as a well defined erythematous tender patch on medial and back of the lower third of both legs with more affection on left than right - condition has been slowly progressing. Patient did not suffer from varicose veins or any systemic disease. All investigations done were within normal and included CBC, blood biochemistry, RPR, ASOT, ANA, fibrinogen and fibrinogen degradation products. Skin biopsy showed panniculitis. Patient was put on stanazolol 5 mg twice daily.

**Discussion:**

We describe 3 cases which showed clinical manifestations which characterize the diagnosis of lipodermatosclerosis mainly irregular erythema, induration and hardness of long duration affecting both legs in three males.

The first patient was an obese 50 year old who gave history of acute phase, provisionally diagnosed as cellulitis that did not respond to antibiotic and the condition persisted for over one year. He had no evidence of venous insufficiency or any preceeding illness or trauma. The biopsy taken was not deep enough to include the subcutaneous fat.

The second patient was 57 years old who presented with the clinical pattern diagnostic of lipodermatosclerosis and was accompanied by mild telangiectasia and superficial varicosity of both legs. Biopsy was not done.

The third patient was 29 years old who was diagnosed to have lipodermato sclerosis clinically with no varicose veins or systemic illness. Histopathologically the biopsy showed panniculitis. All investigations done to the 3 patients were normal including fibrinogen estimation and fibrinogen degradation products.

Kirsner et al.\(^5,6\) believe that there is an acute inflammatory stage of lipodermatosclerosis that is followed in several months or perhaps even years by a chronic stage characterized by fibrosis and sclerosis associated with venous insufficiency. Pericapillary fibrin is deposited in lipodermato-sclerotic skin with venous disease without ulceration.\(^3,4\) This pericapillary fibrin deposition is not only due to venous hypertension but also due to faulty and reduced fibrinolytic system.\(^6\) Patients with venous disease have a prolonged euglobulin lysis time and increased fibrinogen levels.\(^5\) The tissue fibrin deposition is linked pathogenetically to the faulty fibrinolysis in patients with venous disease and lipodermatosclerosis.\(^3\)

The pathogenesis of venous leg ulcers is based on the increase in size and permeability of dermal skin capillaries resulting from prolonged venous hypertension thus allowing fibrinogen to leak leading to pericapillary fibrin cuff which by impeding
exchange of oxygen and other nutrients between blood and dermis causes tissue anoxia and ultimately ulceration. In addition to increased venous pressure other mechanisms take place such as distension of endothelial pores, decreased fibrinolytic activity, increased platelet adhesion to damaged endothelium, fibroblast proliferation, new collagen deposition and angiogenesis. The capillaries also get plugged with white cells. The fibrin around the capillaries is undegraded. Tumor necrosis factor and elastase activities were detected in monocytes and polymorphonuclear cells respectively in the granulation tissue of the ulcer. The tumor necrosis factor alpha induce the formation of pericapillary fibrin cuff and the toxic metabolites released by polymorphonuclear cells may explain the absence of wound repair. Patients who have liposclerosis show histologic changes of lipomembranous panniculitis as a result of inflammatory reaction related to different diseases including venous insufficiency with connective tissue disease or previous leg ischaemia or both. Unusual acid fast microbes were seen in tissue section from two cases of hypodermitis sclerodermiformis.

Treatment:

1. Ultrasound was successfully used to treat 6 cases of hypodermitis sclerodermiformis (HS).

The mechanism of action of ultrasound in HS remains undetermined. Ultrasound therapy has been effective in healing cutaneous infection as sporotrichoses possibly because of its heating effect. Ultrasound was used also to treat psoriasis and cutaneous leishmaniasis.

2. The use of graded compression stockings is essential.

3. Stanozolol which is an anabolic steroid that enhances fibrinolysis is used effectively to treat lipodermatosclerosis on the basis of the presence of a reduced fibrinolytic activity in blood and vein walls and pericapillary fibrin deposition in such patients. The dose is 5mg twice daily for 3-6 months. The drug is also used to treat conditions associated with excess fibrinogen such as Raynaud's phenomenon and cryofibrinogenemia. Side effect of stanozolol include sodium retention, hirsutism, acne, liver function abnormality, lipid abnormality and dysmenorrhea.

4. Antibiotics, nonsteroidal anti-inflammatory drugs and anti metabolites were also used.

Our three patients could benefit from a combination of the use of ultrasound heating, compression stocking and nonsteroidal anti-inflammatory drugs. No specific cause could be detected in the 3 patients.

REFERENCES: