

COVERAGE OF CUTANEOUS ULCERS IN WERNER'S SYNDROME

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Abstract. We describe 6 years of experience with a 51-year-old female with Werner's syndrome in order to encourage skin grafting for recurrent cutaneous ulcerations of this disease. The validity of prompt skin grafting should be no longer in question, for the "granulation bed" in the cutaneous ulcers, when properly prepared, will support skin grafts. The result in addition to achieving skin coverage is to provide some protection over the weight-bearing areas so the patient will again tolerate some ambulatory trauma, and thereby often maintain near-normal activity.

The cutaneous ulcers in Werner's syndrome present a difficult but not impossible problem in skin grafting. Although pressure and trauma are important causes of the ulcers, it is the unusual atrophy of skin in the syndrome that suggests a difficult coverage problem (7). Premature ageing of all tissues characterizes Werner's syndrome. Not only are the epidermis and dermis atrophic, but also the subcutaneous tissue is extremely thin or absent in some areas. There may be very little "padding" over bony prominences, and therefore cutaneous ulcers develop over malleoli, heels, toes, and plantar surfaces of the feet with tiresome regularity (3).

Even the underlying bone in these areas and on the sole of the foot shows radiographic changes consistent with severe osteoporosis and hypertrophic arthritis. Areas of calcification are seen in the skin and subcutaneous tissue, but it is uncommon for frank osteomyelitis to occur in the areas of ulceration (1).

The progeria of Werner's syndrome may include severe arteriosclerosis, but the skin changes usually antedate the vascular lesions (4). Collected experience with this disease shows that treatment will be more successful than might be assumed. Granulation tissue of fairly good quality can be

established regularly even in distal areas such as the foot. Split-thickness skin grafts will "take" on these areas so long as all the usual precautions are exercised. Prolonged support with elastic bandages is required when skin grafts are placed over the malleoli and calcaneal regions, but under these circumstances the patient will tolerate ambulatory trauma fairly well (2, 5).

We observed that donor sites in the thigh region healed with no delay and with only minimal scarring when the skin grafts were a 10/1000 to 15/1000 inch thickness (Fig. 1).

Prior reports of successful skin grafting of the cutaneous ulcers in Werner's syndrome mention isolated cases with brief follow-up of the patients. In Werner's syndrome, prolonged treatment of recurrent ulcerations is more usual (6).

Our case was a female patient followed over six years, who was able to remain a productive member of society throughout, partly because of the previously grafted intact skin over the pressure points of her feet.

CASE HISTORY

This 46-year-old white female was first seen at the University of Missouri Medical Center in 1963 because of cutaneous ulceration over her Achilles tendon that had appeared three years previously (Fig. 2). She had had premature greying of her hair since age 16. By age 25, she had loss of muscle mass and thinning of her skin particularly on her legs. While still young, she developed visual defects and arthritis of old age.

She had had a left mastectomy at age 30 for a "benign" tumor mass. She had successfully terminated a single pregnancy at age 37 years, which is unusual in this syndrome, but she subsequently underwent early menopause at age 41 years. Her family history was not remarkable.

The features of Werner's syndrome were as follows:

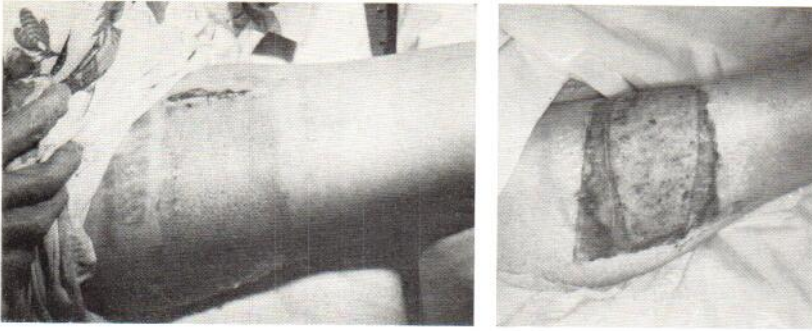


Fig. 1. Donor site on thigh (early and late).

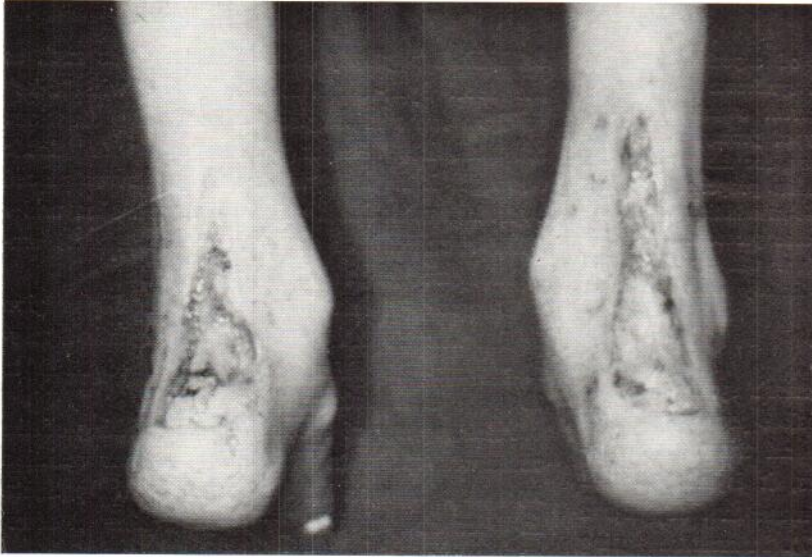


Fig. 2. Heels, skin grafted successfully six years ago.

she was small in stature with a weight of 68 lbs., and a height of five feet. She had a disproportionate thinness of her extremities with atrophic skin that was ulcerated over the Achilles tendon, toes, plantar surface and medial malleoli. Her toes were also atrophic and partially

absent. The toenails were brittle and scant. She had a "bird" face with grey hair, a pointed nose, and a relative exophthalmus of her eyes with bilateral anterior and posterior subcapsular cataracts. Her voice was of a weak quality and high pitched. Peripheral pulses were present



Fig. 3. Left Achilles region showing ulceration, early and late skin grafting.

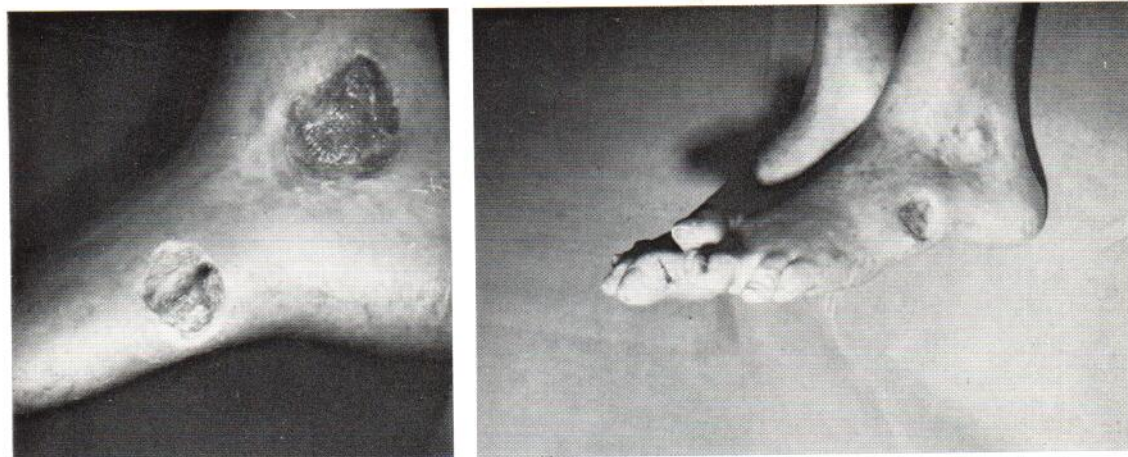


Fig. 4. Malleolus and lateral surface of foot showing ulceration and coverage with skin grafts.

but dorsalis pedis was diminished on the left and posterior tibial was diminished on the right.

The laboratory examination was essentially normal. Her fasting blood sugar and two hour postprandial blood sugar were normal. No sugar nor acetone was ever noted in her urine. A urinalysis, CBC, BUN, creatinine, calcium, phosphorus, alkaline phosphatase, total proteins, 17 Keto and 17 Hydroxy steroids were entirely normal. A uric acid was not obtained but a serum creatine was 0.96 mg%.

The X-rays showed marked osteoporosis of all visible bones, premature degenerative changes in the hands and soft tissue calcification in the knees and elbows. EKG was abnormal with ST segment elevation suggesting epicardial injury. A skin biopsy on January 10, 1964, showed generalized atrophy of the skin with thin epidermis showing normal maturation but no rete-pegs. There was generalized absence of adnexal skin glands (hair follicles, sebaceous glands, etc.) with decreased collagen and fragmented elastic fibrils. No subcutaneous fat was seen in microscopic sections.

On two hospitalizations separated by 5½ years, she was successfully skin grafted. One Achilles tendon graft remained partially intact during the interval while the other almost completely broke down. Additional skin grafts did "take" in both of these areas (Fig. 3). The malleoli required repeat skin grafting which was successful (Fig. 4), but grafting of the plantar surface of the heel proved unsuccessful on the last hospitalization.

At present the patient lives as a housewife, but has limited activity outside the home. Her son, who is now 15 years of age, and her husband are attentive and help relieve some of the household responsibilities. The patient's attitude remains positive and realistic and although additional arthritic changes have further deformed her feet, she continues to walk well.

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