Scleroderma Skin Ulcers: Ovine Extracellular Matrix With Hyaluronic Acid Xenograft, A Promising Approach To An Orphan Disease

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INTRODUCTION

Systemic sclerosis (SSc), commonly referred to as scleroderma, is a chronic autoimmune rheumatic disease affecting multiple systems. Excessive fibrosis, blood vessel abnormalities, and immune system disruptions mark this rare condition. When SSc impacts only the skin, it is categorized as "localized" scleroderma.[1] Medical professionals must thoroughly assess each patient to identify specific symptoms, monitor disease activity, and evaluate the risk of organ-specific complications, which guides appropriate screening and treatment. Individuals with SSc may experience ischemic digital ulcers, leading to noticeable epithelial loss, contractures, and intense pain, with an increased risk of infections and potential amputation.[2] Furthermore, losing a finger can significantly diminish a person's quality of life and should be avoided whenever possible.

METHODS

The setting of this case report was a hospital-based outpatient diabetic wound care clinic at Vaiola Hospital, Nuku'alofa, the main hospital in the Kingdom of Tonga. The patient was provided standard wound care for a right fourth finger wound with a combination of topical therapies and selective debridement. In Tonga, there is limited availability of chemotherapy or immunotherapy for treating systemic sclerosis. After remaining refractory for three months to standard wound care, a cellular, acellular, matrix-like product (CAMP) was added to the patient's wound care treatment regimen. The standard-of-care techniques were continued while incorporating a xenograft composed of a layer of glycosaminoglycans (hyaluronic acid) between sheets of an ovine forestomach matrix (OFM) made from decellularized extracellular matrix (ECM) (Symphony*, Aroa Biosurgery, Auckland, New Zealand).

RESULTS

The patient is a 54-year-old female with the diagnosis of type 2 diabetes mellitus and scleroderma. She lives with her husband and seven children. Denies alcohol and tobacco use. In 2017, she underwent a left below-the-knee (BKA) amputation, followed by a right BKA in 2022. Secondary to her bilateral BKAs, the patient has been mobilizing via a wheelchair since 2014. On 03/01/2024, that patient presented to a satellite non-communicable diseases (NCD) clinic with an open wound on her right fourth finger that occurred due to crawling on the ground. She was referred to the diabetic wound clinic at Vaiola Hospital, where a NCD nurse prescribed oral Bactrim and topical silver sulfadiazine cream to be applied to the open ulcer daily.

* Symphony grafts donated by **Aroa Biosurgery** to the non-profit diabetic foundation '**Amanaki Fo'ou** for use at Vaiola Hospital in the Kingdom of Tonga.

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Results

The lesion initially responded to treatment and temporarily closed but recurred 25 days later. From 03/26/2024 through 07/19/2024, the lesion remained refractory to a number of available standard topical therapies, such as iodine gauze, normal saline (NS) moistened alginate dressings, and selective debridement.

The right fourth digit, after 12 weeks, remained non-responsive to conservative standard-of-care techniques, such as previously mentioned, NS moistened alginate dressings, along with selective debridement. On 07/19/2024, more aggressive sharp surgical debridement was performed along with the application of an OFM graft moistened with a stabilized hypochlorous acid solution.

On 07/24/2024, five days after the first OFM xenograft graft with hyaluronic acid was applied, a visible improvement in the appearance of granulation tissue was observed in the base of the ulcer.

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Results

On 08/20/2024, 32 days after adding a CAMP to the treatment regimen, the hard-to-heal right fourth finger ulcer had resolved. After recurrence, this wound persisted for 115 days before initiating the OFM xenograft.

A total of six applications of the OFM graft containing a layer of hyaluronic acid were applied until the closure of the scleroderma dermal ulcer was achieved. On average, the xenografts were placed every 5.3 days.

DISCUSSION

This case report highlights the successful integration of an ovine forestomach matrix (OFM) containing hyaluronic acid xenograft into the wound care regimen for a complex scleroderma-related dermal ulcer. In a patient with type 2 diabetes and systemic sclerosis, whose right fourth digit wound had remained refractory to standard-of-care therapies, the addition of this novel CAMP led to marked improvement. Notably, visible enhancement in granulation tissue was observed just five days after application, and complete wound closure was achieved within 32 days following six applications. These results suggest that even in resource-limited settings like the Kingdom of Tonga, such an advanced approach with an OFM xenograft can be a promising adjunct for managing hard-to-heal ulcers associated with orphan diseases. Further studies are warranted to confirm these findings and explore this treatment strategy's broader potential for similar challenging cases.

References
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