BRIEF ARTICLES

Elephantiasis Nostras Verrucosa: A Case Series

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ABSTRACT

Elephantiasis nostras verrucosa (ENV) is a rare complication of chronic lymphedema that can cause significant disfiguration of the affected body part. We present a case series of two patients encompassing a spectrum of ENV severity to help medical providers become more comfortable identifying and managing ENV, with the goal of ultimately improving patient outcomes.

INTRODUCTION

Secondary lymphedema is caused by extra-lymphatic disease processes including trauma, infection, congestive heart failure, obesity, malignancy, and venous stasis.[1] An uncommon complication of longstanding secondary lymphedema is elephantiasis nostras verrucosa (ENV). Delayed identification and treatment of this disease process can lead to poor patient outcomes, (e.g. deformity/impairment of limbs, localized infections and lymphangitis, or Stewart-Treves Syndrome).[1] Here we present two patients with ENV that span a spectrum of disease severity to facilitate medical providers identification of the disease and assessment of disease progression so that they can take early intervention and improve patient outcomes and satisfaction.

CASE PRESENTATION

Patient 1 is a 74-year-old woman with a relevant past medical history of poorly-controlled diabetes on insulin, hypertension on amlodipine, and hyperlipidemia who presented to the dermatology clinic with 6 months of bilateral leg swelling and xerosis. The patient denied using any home remedies, prior treatments, and reported no pertinent travel or family history. Relevant medications included baby aspirin, furosemide, amlodipine, irbesartan, atorvastatin, and insulin. Physical examination revealed verrucous nodules and plaques scattered over bilateral lower legs down to the ankle (Figures 1A and 1B) with concomitant non-pitting edema of bilateral lower extremities and a positive Kaposi-Stemmer sign on bilateral 2nd toes. Sensation was intact, but the surrounding skin was warm and tender. Initial management included leg elevation, compression stockings, triamcinolone acetonide 0.1% ointment, and topical ammonium lactate.

Patient 2 is a 75-year-old man with a past medical history significant for hypothyroidism, hypertension on amlodipine, congestive heart failure (CHF), hepatitis C-

Figure 1. (**A**) Patient 1's left lower leg; hyperpigmented verrucous plaques, scaling fissures, and significant xerosis. (**B**) Patient 1's right lower leg demonstrating hyperpigmented and pink verrucous plaques with cobblestoning.

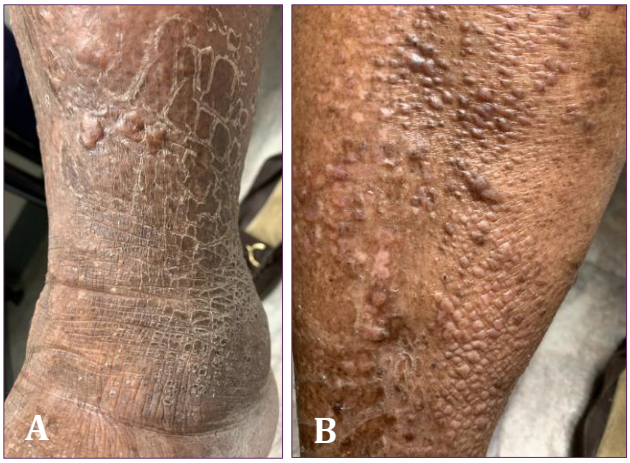
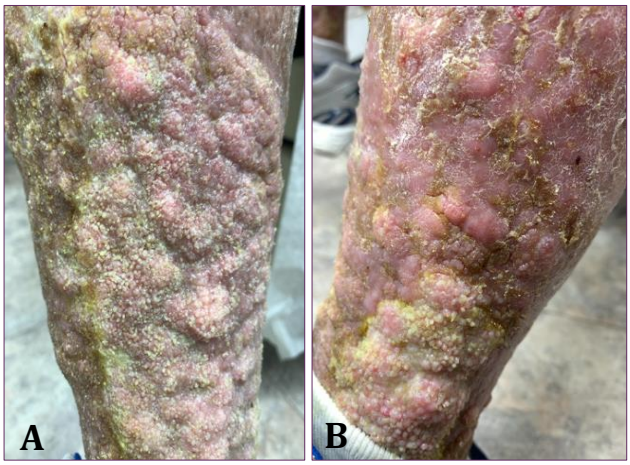


Figure 2. (**A**): Patient 2's left lower leg demonstrating cobblestoning with pink verrucous plaques and impetiginization. (**B**): Patient 2's right lower leg demonstrating cobblestoning with pink verrucous plaques and impetiginization



induced cirrhosis now 6 years status post liver transplant, and stroke who presented to the dermatology clinic with bumps on his legs for one year. He reported that these bumps were increasing in number and spreading proximally up his lower legs. The patient had been using a topical collagenase and sodium chloride 0.9% that mildly alleviated his symptoms for a short period. Relevant medications included warfarin, levothyroxine, tacrolimus, metoprolol, amlodipine, and furosemide. Physical exam revealed bilateral lower extremity cobblestoning with erythema and diffuse honey-colored crust concerning for secondary impetiginization (Figures 2A and 2B), and concomitant non-pitting edema of bilateral feet and lower legs with a positive Kaposi-Stemmer sign on bilateral 2nd toes. The patient was prescribed doxycycline 100mg twice daily for one month in addition to topical steroids and lower extremity compression and elevation.

DISCUSSION

ENV is the result of chronic lymphedema that manifests as a nonpitting edema with a papulonodular cobblestone appearance due to excessive accumulation of proteinaceous material in the extracellular matrix. While the most common site for ENV is in the lower extremities, as they are a gravity-dependent area, ENV can occur anywhere.[2]

The diagnosis of ENV is largely clinical and includes a wide differential diagnosis (e.g. filariasis, pretibial myxedema, lipedema, chromoblastomycosis, lipodermatosclerosis, Stewart-Treves syndrome, and the more common venous stasis dermatitis). To differentiate ENV from these other diagnoses, a thorough history and physical exam must be attained.[3,4] Kaposi-Stemmer sign, as demonstrated in both of our patients, is the inability to pinch the dorsal aspect of the skin at the head of the second metatarsal and is indicative of lymphedema.[1]

Many comorbidities put pressure on the lymphatic system and increase lymph capillary permeability. Unlike primary lymphedema, which is caused by defects in the lymphatic system, secondary lymphedema is more common and is the

result of a separate primary disease process including infectious (e.g. filariasis, especially in developing countries) and noninfectious etiologies (e.g. malignancy).[5,6] Chronic uncontrolled heart failure, obesity, and hypothyroidism are also common risk factors for secondary lymphedema, with CHF (affecting > 2% of the US population) and obesity (affecting ~25% of the US population) being the most prominent.[7,8] Timely medical management of these chronic systemic conditions may help prevent the development of ENV.[3,4] As such, it is imperative patients and physicians engage in multidisciplinary care and have open communication with patients' primary care providers to adequately monitor underlying comorbidities.[9]

Successful treatment of ENV is also dependent on patient compliance. A multifaceted treatment approach involving compression, diuretics, antibiotics (if there is an infectious component), and possibly systemic retinoids is typically most effective.[10] Lifestyle changes including increased ambulation, weight loss, and leg elevation (above heart level) prove beneficial as well and are included as first-line therapy.[10,11] Surgical intervention, e.g. debridement of affected skin, may be considered in recalcitrant cases, but does not correct the underlying cause.[10,12] Early diagnosis and intervention are key as later stages are more difficult to manage and ultimately reverse. Identifying the early signs of lymphedema, such as a pitting edema, may improve patient outcomes and implementing lifestyle modifications (such as compression/elevation of the affected limbs) are essential elements of the treatment plan.[13] Furthermore, diligent follow-up (within 1 month for management of an infectious component, otherwise at least every 3-6 months) is essential for tracking progression/resolution and ensuring proper management.[10]

CONCLUSION

ENV is a rare complication of very common chronic systemic conditions. Adequate management of ENV requires multi-modal therapy, multidisciplinary care, and cooperative coordination between patient and provider.

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