Lazarus ME, Larsen TB, Napolitano JD. Elephantiasis Nostras Verrucosa. *Brown Hospital Medicine*. 2025;4(2):63-65. doi:10.56305/001c.129041

Images in Hospital Medicine

Elephantiasis Nostras Verrucosa

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Journal of Brown Hospital Medicine
Vol. 4, Issue 2, 2025

Article Information

Keywords: Papillomatosis, elephantiasis, verrucosa, woody induration, wart like, lichenification

https://doi.org/10.56305/001c.129041

Submitted: January 07, 2025 EDT

Accepted: January 20, 2025 EDT

Published: April 01, 2025 EDT

Abstract

76-year-old male skilled nursing facility resident with a history of bilateral, deep venous thromboses of his lower extremities, chronic venous stasis ulceration of his legs and recurrent episodes of lower extremity bacterial cellulitis was admitted with chronic, bilateral lower extremity woody induration and papillomatosis resembling elephantiasis. He had no exposures to Filarial infection, normal thyroid function, no history of diabetes, congestive heart failure or malignancy. A clinical diagnosis of elephantiasis nostras verrucosa was made. This rare chronic condition is cause by chronic lymphatic inflammation and fibrosis. He was treated with topical retinoids, pneumatic compression and bandages.

A 76-year-old man was admitted with lethargy and failure to thrive from a skilled nursing facility. His prior medical history included atrial fibrillation and bilateral, provoked deep venous thromboses of his lower extremities on anti-coagulation. He also had chronic venous stasis ulceration of his legs with the right side worse than the left as well as a history of recurrent episodes of lower extremity bacterial cellulitis. His surgical history included a sigmoid volvulus requiring sigmoidectomy and partial colectomy with ileostomy and subsequent take-down. Two years prior he had bilateral knee replacements for severe osteoarthritis. He denied fever, chills or night sweats and had not experienced recent weight loss. He had been ambulating with a walker but had recently been sedentary. Medications included metoprolol, digoxin and apixaban. He denied alcohol but had a remote history of tobacco use. He had no history of Filarial infection, nor had he ever travelled to endemic areas. His vital signs were significant for a low systolic blood pressure of 90 mmHg which normalized with intravenous fluids. Cardiopulmonary exam was otherwise unremarkable. His abdominal exam revealed a healed midline abdominal scar, no palpable masses, organomegaly or adenopathy. His lower extremities and feet were notable for papillomatous and lichenified, circumferential lesions with scattered chronic erythematous areas particularly on the thighs and around his left ankle. His right lower leg was more swollen than his left. (**Figure 1**) On palpation, his skin lesions were coarse, indurated and scaly hard with a wood like consistency. Skin thickening around the ankles had a verrucous appearance, greater on the right than left side (**Figure 2**). Bilateral midline scars on both knees, consistent with prior knee arthroplasties, were noted. Bilateral lower extremity venous doppler ultrasound was negative for acute deep venous thrombosis. His thyroid function testing was unremarkable, excluding a diagnosis of pretibial myxedema. Based on the history, chronicity and appearance of his legs a diagnosis of Elephantiasis Nostras Verrucosa (ENV) was made.

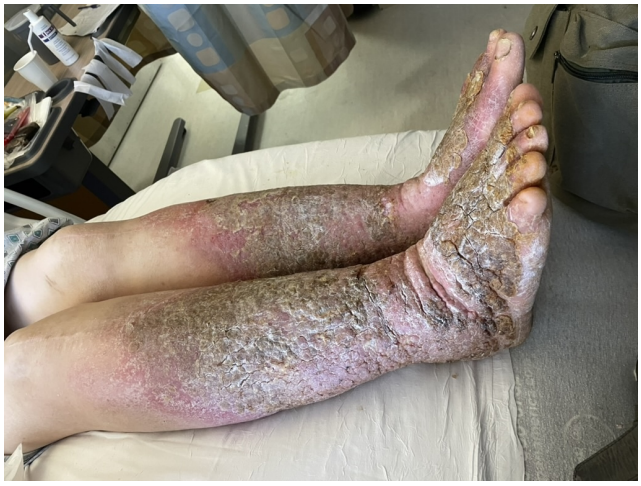

Figure 1. Bilateral papillomatous and lichenified, circumferential lesions with scattered chronic erythematous areas particularly on the thighs and around his left ankle

While the cause of ENV is unknown, the pathogenesis of our patient's leg swelling likely began with recurrent lower extremity staphylococcal cellulitis and streptococcal erysipelas leading to chronic lymphangitis.[1] This eventually progressed to fibrosis of the dermis and lym-

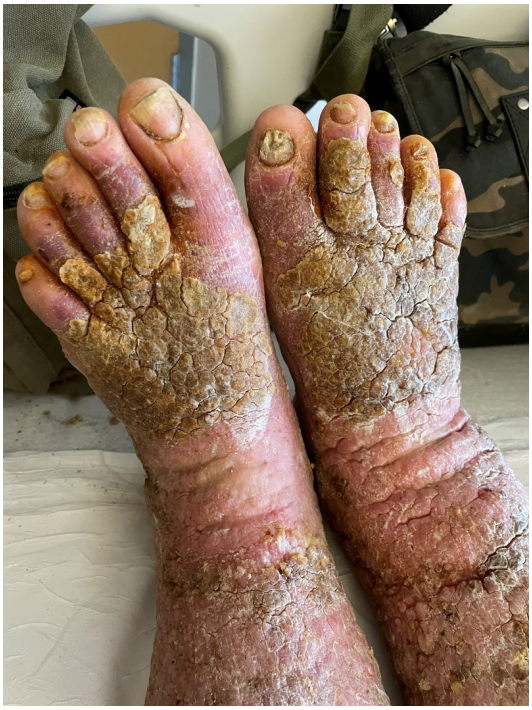

Figure 2. Skin thickening around the ankles had a verrucous (wart-like) appearance, greater on the right than left side

phatic system of his lower legs causing progressive lymphedema. Impaired lymphatic drainage accompanied by chronic venous stasis and reduced mobility perpetuated the cycle.[2] ENV can be caused by malignancies and obstructing masses that block lymphatic drainage including lymphoma and are ruled out by abdominopelvic computed tomographic scans. Other notable causes of secondary lymphedema resulting in ENV include surgical procedures, radiation and scleroderma. In rare cases, secondary malignancies such as squamous cell cancer and/or angiosarcoma should be ruled out with skin biopsies. Obesity and congestive heart failure are promotors of secondary lymphedema which have increasing prevalence in the US and often coexist in ENV patients.[3]

After obstructive lesions were ruled out, treatment began by reducing the lymphedema with bandages, compression stockings, and pneumatic compression applied to the lower extremities daily. Topical retinoids to reduce lichenification and papillomatosis were started.[4] Subsequently, urea and 10% salicylic acid treatments can be considered to reduce keratinization and, if needed, surgical debridement of verrucous areas can be attempted.[5] Dermabrasion has been used to thin dermal thickening in some patients. Unfortunately, a cure for this chronic condition is unlikely.

Author Contributions

All authors have reviewed the final manuscript prior to submission. All the authors have contributed significantly to the manuscript, per the International Committee of Medical Journal Editors criteria of authorship.

- Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
- Drafting the work or revising it critically for important intellectual content; AND
- Final approval of the version to be published; AND
- Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

Disclosures/Conflicts of Interest

The authors declare they have no conflicts of interest

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