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# Elephantiasis nostras verrucosa: an atypical presentation following intrapelvic lymphoma

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## Abstract

Elephantiasis nostras verrucosa is a progressively debilitating and disfiguring disease commonly presenting with verrucous, cobblestone-like papules, nodules, or plaques with nonpitting edema in the lower extremities. Histopathology is marked by hyperkeratosis and dermal or subcutaneous fibrosis as a result of chronic lymphedema. Risk factors include obesity, recurrent cellulitis, chronic venous insufficiency, congestive heart failure, scleroderma, radiation, trauma, and tumors. We report a 72-year-old man who presented to the dermatology clinic for an 11-year history of edematous legs, occasionally associated with ulcerations. The findings developed within a year of intrapelvic non-Hodgkin lymphoma and progressed gradually over 10 years after lymphoma remission. Physical examination revealed atypical features including compressible cysts and pitting edema extending from the lower legs to the thighs bilaterally. The patient was noncompliant for the recommended compressive devices and the condition progressively worsened over the course of 7 months of follow-up. Early interdisciplinary management using compressive devices and a lymphatic pump are recommended. Underlying causative factors should be assessed with regular follow-up to optimize treatment outcomes.

*Keywords: elephantiasis nostras verrucosa, elephantiasis, lymphedema, lymphoma*

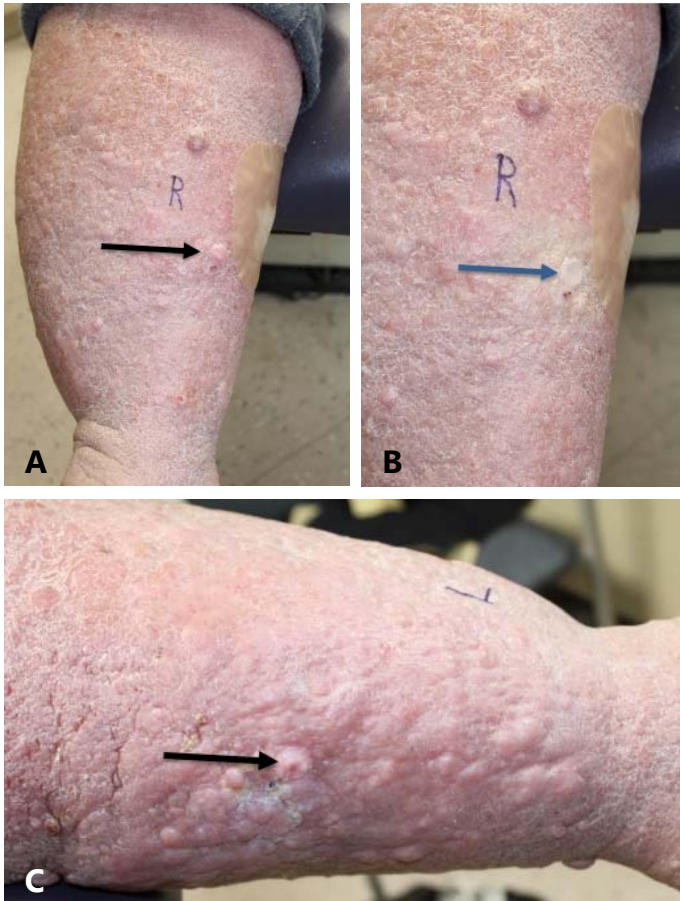
## Introduction

Elephantiasis nostras verrucosa (ENV) is a debilitating and cosmetically disfiguring complication of chronic

lymphedema [1, 2]. ENV commonly presents with diffusely scattered, verrucous, cobblestone-like papules, nodules, or plaques with nonpitting edema in the lower extremities; histopathology is marked by hyperkeratosis and dermal or subcutaneous fibrosis [1, 2]. Other locations that have also been reported include the upper extremities [3], ears [4], abdomen [5], back [6], sacrum [7], buttocks [7], and scrotum [8]. Reported risk factors include obesity, recurrent cellulitis or other soft-tissue infection, chronic venous insufficiency, surgery or other trauma, tumors, radiation, congestive heart failure, and scleroderma [1, 3]. We report an atypical case of ENV presenting with compressible cysts in the bilateral legs and thighs, with pitting edema extending to the thighs, that unusually persisted for 10 years after lymphoma remission.

## Case Synopsis

A 72-year-old man presented to the dermatology clinic for an 11-year history of edematous legs, occasionally associated with ulcerations, with an insidious onset within a year of developing intrapelvic lymphoma. The patient reported feeling “very depressed” and physically disabled owing to his condition. He denied any pain, pruritus, fever, chills, weight loss, headache, nausea, vomiting, or international travel. Past medical history was significant for intrapelvic non-Hodgkin follicular B cell lymphoma status post-remission with chemotherapy, hypogammaglobulinemia, and morbid obesity. The patient did not have a history of lower extremity cellulitis.



**Figure 1.** (A-C). Clinical examination. Right **A**) and left **B**) lower extremities exhibited verrucous, flesh-colored to erythematous nodules and cysts (black arrows). Cysts were compressible upon manually applied pressure (blue arrows). **C**) Follow-up at four months of non-compliance to compressive devices or lymphedema pump revealed progression of cutaneous lesions.

Physical examination revealed a verrucous, flesh-colored-to-erythematous eruption consisting of compressible, weeping cysts and nodules of the bilateral legs and thighs associated with 2+ pitting edema extending to the thighs (**Figure 1**). No ulcers were observed at the time of examination. Leg circumference measured 30cm in bilateral ankles, 47cm and 51cm at 15cm above the left and right ankles, respectively, and 49cm and 52.5cm in left and right calves, respectively. Capillary refill was 3-4 seconds. Body mass index (BMI) was 37.45.

Thyroid stimulating hormone was within normal limits. Serum IgG levels were significantly decreased at <200mg/dL. Complete metabolic panel was unremarkable for chronic kidney or liver disease. Echocardiogram was negative for heart failure. Venous duplex reflux studies displayed superficial

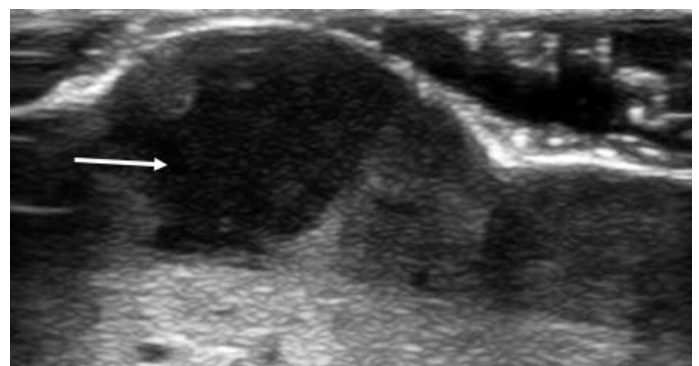
cystic lesions consistent with lymphatic ectasia (**Figure 2**) bilaterally and evidence of superficial venous reflux with duration of 2.6-4.3 seconds involving the greater saphenous vein in the right lower leg only.

A diagnosis of ENV was made. Differential diagnoses included venous stasis dermatitis, filariasis, pretibial myxedema, and lymphedema. The patient was advised to use triamcinolone ointment, 0.1%, customized Circaid compression device, and a lymphatic pump daily, although there is currently no standard guideline on frequency of lymphatic pump therapy.

However, he was noncompliant for the recommended management as he reported difficulty with using the compressive device and lymphatic pump independently at home and considered the triamcinolone ointment to be too "greasy." The lesions progressively worsened upon follow-up at one month, four months, and seven months (**Figure 1C**). Home health care visits were arranged to provide help with the compressive device and lymphatic pump at home in order to optimize management.

## Case Discussion

According to a previously published retrospective analysis of 21 cases from 2006 to 2008, the most significant predisposing risk factors for ENV were obesity (100%) and cellulitis or other soft-tissue infection of the lower extremity (86%), [1]. The most likely precipitating factor for our patient's initial onset was intrapelvic lymphoma obstructing the lymphatic channels.



**Figure 2.** Ultrasound findings. Hypoechoic superficial cystic lesion (arrows) exhibited absence of flow using color Doppler.

Interestingly, despite our patient's remitted lymphoma, the patient continued to be affected by ENV bilaterally in the lower extremities. The persistence of the condition after remission of the tumor is highly unusual. Contributing factors to the persistent bilateral presentation may include increased hydrostatic pressure related to lymphatic obstruction by fibrosed intrapelvic lymph nodes following chemotherapy and biopsy, as well as decreased osmotic pressure related to hypogammaglobulinemia, promoting osmotic flow from the vasculature to the interstitium. The superficial venous reflux observed in the right lower leg may also indicate venous insufficiency as one contributory factor. Unlike most reported cases of ENV, the patient exhibited pitting edema and compressible cysts and nodules bilaterally. This unusual presentation could be a long-term consequence of chronic lymphatic ectasia extending to the superficial lymphatic channels concurrently with significantly altered osmotic pressures that worsen lymphatic flow mechanics, particularly in a patient noncompliant to recommended management with multiple risk factors (obesity, history of tumor, hypogammaglobulinemia, venous insufficiency).

Application of a compressive device and lymphatic pump as initial therapy, along with early interdisciplinary management of underlying risk factors, is advised [1, 2] in order to prevent long-term sequelae including the severe, persistent edema and

nodulocystic changes seen in our patient. Successful treatment with surgical excision [8, 9], lymphaticovenular anastomosis [10], systemic retinoids [11, 12], and ablative carbon dioxide laser [13] have also been reported in the literature. Limitations in patient compliance to recommended compression have hampered resolution in this case.

## Conclusion

Elephantiasis nostras verrucosa is a progressively disfiguring disease commonly marked by extensive nonpitting edema and fibrotic papules, nodules, or plaques. Rarely, a patient may present with pitting edema and superficial, compressible cysts, as seen in our case. Furthermore, it is unusual for ENV associated with a tumor to persist after the tumor has been treated.

Patients with physical or psychosocial disabilities may benefit from having a caregiver or regular home care visits arranged to increase compliance with the recommended treatment and prevent disease progression. Early interdisciplinary management using compressive devices and a lymphatic pump may be helpful. Addressing underlying causative factors, is strongly advised with regular follow-up to optimize treatment outcomes.

## Potential conflicts of interest

The authors declare no conflicts of interests.

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