

## Elephantiasis Nostras Verrucosa of Lower Limb: a Case Report

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**Abstract-** *Elephantiasis nostras verrucosa* (ENV) is a rare condition in which hyperkeratosis, fibrosis and disfiguration of dermis occurs. It is caused mostly by non infectious diseases such as surgery, trauma, tumors, and venous obstructions. To our knowledge there is not any case report of ENV in Middle East region. In this patient, ENV caused by trauma and Patient presented with enlargement of right lower limb.

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### Introduction

*Elephantiasis nostras verrucosa* (ENV) is a rare chronic pathologic condition caused by non-filarial lymphatic obstruction. Infection, surgery or trauma, tumors, chronic venous stasis, congestive heart failure, radiation and obesity are the main etiologies. Lymphatic obstruction leads to non pitting edema, hyperkeratosis, and dermal fibrosis gradually (Figure 1). Cellulitis, limb dysfunction and cosmetic problems are the complications of ENV (Figure 2).

In this report authors describe a patient with trauma induced ENV, pathophysiology of ENV and its differential diagnoses.

### Case Report

A 40-year-old man came to our clinic with the chief complaint of progressive right leg and foot enlargement and several crusted lesions. His problem has started since 10 years ago after a car accident and trauma to his right leg. The patient denied any surgery, radiation, and travel to a tropical region or medical illness. He did not take any medication.

On physical exam there were mossy, cobble stone and verrucous papules, nodules, plaques on enlarged right leg and dorsum of the right foot (Figure 1). His lesions were malodorous with crust and oozing. *Staphylococcus aureus* grew in lesion's culture.

Laboratory tests including complete blood count, C-reactive protein, thyroid function test were normal. Skin biopsy showed hyperkeratosis, dermal fibrosis and

dilated lymphatic channels. KOH preparation and acid-Schiff for fungal infection were negative. Venous Doppler sonography showed normal flow and was negative for venous thrombosis. According to these characteristics, ENV was diagnosed for him.



**Figure 1.** A: papulonodules and plaques on right leg and dorsum of right foot. B: large nodule on heel. plaques and lesions with oozing. C: hard and cobble stone appearance, papule and plaques

Cephalexin was prescribed to treat secondary bacterial infection and Acitretin to improve hyperkeratosis. He also was referred to orthopedist for debridement. Unfortunately, the patient did not return for followup.

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## Discussion

*Elephantiasis nostras* was first used by Castellani in 1934 to distinguish between elephantiasis caused by *Wuchereria* worms called *elephantiasis tropica* from elephantiasis caused by other etiologies including trauma, tumor, radiation, congestive heart failure and obesity (3). In this condition due to lymphatic obstruction, protein rich fluid accumulates at dermis, below site of obstruction, and stimulates fibroblasts, keratinocytes and adipocytes proliferate, therefore causing dermal fibrosis. Because lymphatic vessels have an important impact on immune cell trafficking, their obstruction leads to immune system dysfunction. Moreover, lymphatic pressure at dermis can cause skin

break and therefore predisposes the tissue to cellulitis, lymphangitis and other infections (1,2).

At first lymphatic obstruction induces soft tissue swelling and pitting edema. But by time and chronicity of obstruction the dermis and lymphatic channels become fibrotic and lead to skin disfigurement such as cobblestone like appearance, woody edema and tense nodules (1,3). Due to fibrosis of lymphatic channels the process becomes irreversible.

Diagnosis of ENV is clinically by history and physical examination. ENV differential diagnoses include venous stasis dermatitis, lipodermatosclerosis, lipedema, pretibial myxedema, chromoblastomycosis, and verrucous carcinoma which have its specific feature and signs (Table 1).

**Table1. Common differential diagnoses of ENV**

Diagnosis	Characteristics
Primary lymphedema	Congenital anomaly and dysfunction of the lymphatic system. The most common form is bilateral and below the knee. It can present at any age. Family history may be positive. Its clinical feature is the same as ENV (common)
lipedema	Its onset is at early age of life, especially in females. It is usually present with symmetric legs' enlargement. It is also called "riding breech thighs" or "stove pipe legs" <sup>5</sup> .
Lipodermatosclerosis	It just involves above the ankle in the patient with chronic venous disease. Venous insufficiency leads in subcutaneous fibrosis. It presents like "inverted wine bottle" <sup>1,4</sup> .
Pretibial myxedema	Mucin deposition at the dermis in patients with hyperthyroidism (such as graves' disease) causes non-pitting edema and nodular plaque on the anterior of legs and dorsum of foot <sup>1,3</sup> .
Venous stasis dermatitis	It presents as a pitting edema, desquamative and pruritic plaques with dilated superficial veins <sup>3</sup> .
Verrucous carcinoma	It can be ruled out by pathologic finding of atypical keratinocyte. It is mostly seen in a person who chews tobacco <sup>1</sup> .
Poplar mucinosis	It can be diagnosed by deposition of glycosaminoglycan at papillary dermis. It presents as waxy papules <sup>3</sup> .
Chromoblastomycosis	It is a fungal infection caused by soil fungus penetrated thorough minor trauma of skin <sup>4</sup> .
Deep vein thrombosis	It presents as tender, warm and swelled calf in patients who have risk factor of venous thrombosis <sup>5</sup> .

ENV: Elephantiasis nostras verrucosa

The main treatment of ENV is eradicating the underlying cause. Management of lymphostasis includes elevation of the affected limb, compressive stocking, and pneumatic compression device. If there is any sign of active infection, antibiotic with good coverage of staphylococcus and streptococcus should be used. Oral and topical retinoids can be used to reduce skin hyperkeratosis and fibrosis. If there is not any improvement by medical treatment, surgical debridement or lymphovenous anastomosis should be considered (3,4).

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