

Kaposi-Stemmer sign in elephantiasis nostras verrucosa

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Description

A 70-year-old female with a history of a 20-year venous ulcer of the right lower extremity. She presented to the emergency department with a two-day history of pain in her right leg, purulent discharge, and fever. Initially, she presented with septic shock due to skin and Soft Tissue Infection (SOFA 4 points) requiring vasopressor support. Dermatology was consulted ten days later, when she was already stable and was receiving antibiotic management.

On the anterior and posterior aspects of the distal third of the right lower limb, a large ulcer with well-defined, regular borders and a base with fibrinous and seropurulent material was observed (Figure 1). On the right foot and toes, she presented a large eumchromic plaque with a cobblestone appearance, accompanied by a positive Kaposi-Stemmer sign (Figure 2).

The diagnosis of Elephantiasis nostras verrucosa secondary to a chronic venous ulcer was made. Multiple biopsies were performed to rule out squamous cell carcinoma, and they were negative for malignancy.

Elephantiasis Nostras Verrucosa (ENV) is a rare form of chronic lymphedema characterized by the presence of nonpitting edema and hyperkeratotic papulonodules with a verrucose or cobblestone appearance [1]. Differential diagnosis includes venous stasis dermatitis, pretibial myxedema, and lipedema. The cutaneous lymphatic system forms a vascular

network that drains lymph from the dermis and hypodermis. Obstruction generates lymphostasis and extravasation of lymph between fibroblasts, adipocytes, and keratinocytes. Excess protein activates fibroblasts, which causes fibrosis of the dermis, lymphatic system, and subcutaneous tissue [2]. Causes of ENV include filariasis, chronic ulcers, recurrent erysipelas, neoplastic infiltration, and others. In 1976, Stemmer described the inability to pinch the skin of the proximal phalanx of the second or third toe in patients with lymphedema. Thus, if the examiner is unable to grab the dorsal skin between their thumb and index finger, it is considered positive. This test has a sensitivity of 92% and a specificity of 57% [3].

References

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Figure 1: Chronic venous ulcer.



Figure 2: Positive Kaposi-Stemmer sign.