All Is Not Swell





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70-year-old man is referred to dermatology after trying "everything else" for problems he has had for at least 15 years. In that time, he has been hospitalized repeatedly for swelling and pain in his legs, with odoriferous drainage.

Despite extensive treatment attempts multiple antibiotics, oral and topical steroids, and OTC creams-the condition is worsening. In hospital-based chronic wound care clinics, the patient received other treatments, including debridement, whirlpool therapy, and soft compression casts. The latter helped a bit but were too uncomfortable to leave on, as was the compression hose prescribed for him. Twoweek hospital stays with oral antibiotics and strict bed rest helped considerablybut only for a short time. Most recently, his

primary care provider prescribed a diuretic, which helped for one month.

The patient denies having cancer or deep vein thrombosis (both of which he has been thoroughly checked for), as well as congestive heart failure. He states that almost every morning, upon rising, the swelling in his legs is considerably lessened.

Both legs are swollen, red, and edematous from just below the knees down. Advanced, pebbly, hyperkeratotic plaques cover the lower two-thirds of both legs, favoring the anterior over the posterior portions. Pitting edema is elicited with minimal digital pressure but does not cause any pain.

The patient is in no acute distress but is clearly uncomfortable. He has been confined to a wheelchair for years due to back problems; he can barely stand when asked to do so. He is extremely obese.

The most likely diagnosis is

- a) Cellulitis
- b) Venous insufficiency
- c) Lymphedema
- d) Elephantiasis nostras verrucosa

ANSWER

The correct answer is elephantiasis nostras verrucosa (ENV; choice "d"). Cellulitis (choice "a"), venous insufficiency (choice "b"), and lymphedema (choice "c") are all factors in the broader diagnosis of ENV.

DISCUSSION

ENV is an unusual condition that represents hypertrophic fibrosis secondary to repeated episodes of lymphangitis. This begins with venous insufficiency, which is made worse by increasing obesity (which impedes venous return) and repeated bouts of cellulitis. With ENV, fibroblasts are increased due to extravasation of high-molecular-weight protein (lymphorrhea), which leads to a buildup of keratinocytes, ultimately expressing as extreme hyperkeratosis.

In this patient's case, his sedentary lifestyle and constant seated position contribute to the problem. Many of his past treatments were reasonable, but—as in many ENV cases—his condition is beyond the point of treatment.

Typically, in-home treatment includes compression and elevation of the legs. Topical application of urea creams is often used to soften the rough skin, but in this patient's case, the cream burned so badly that it was of no use. Alas, the very things he needs to do are those he cannot: walk, burn calories, and avoid long periods of inactivity.