WHAT IS YOUR CALL?

Cobblestone-like skin

A 70-year-old man with obesity, hypertension and congestive heart failure presented with a fever of 4-days' duration and pain in both his anterior shins. He had progressive chronic lymphedema of the lower extremities for the last 5 years. The lymphedema had started as recurrent swelling involving the dorsum of his feet, with ultimate involvement of both the front and the back of the legs; his genitals were not involved. He had not travelled outside of Italy in the 10 years before the lymphedema developed.

Physical examination revealed chronic bilateral skin changes involving the lower two-thirds of his legs. His skin was generally tender and brown, with generalized lichenification and verrucous, cobblestone-like papules and nodules that gave his legs a "woody" feel (Figure 1A, Figure 1B). Slight fissures were also present between some of the nodules.

Cultures from selected open skin areas grew *Candida albicans*, *Staphylococcus epidermidis* and β -hemolytic

Figure 1: A) Severe, woody, brown lymphedema involving both of the patient's legs and dorsum of his feet, with deformed fibrotic enlargement of the extremities. B) Verrucous, cobblestone-like skin on the lower two-thirds of his left leg.

streptococcus group A. A biopsy specimen from a representative nodular lesion showed hyperkeratosis, parakeratosis and acanthosis of the epidermis, as well as edema and dilated lymphatic spaces in the papillary and reticular dermis. No neoplastic changes were seen.

What is your diagnosis?

- Elephantiasis nostras verrucosa
- Venous stasis dermatitis
- Pretibial myxedema
- Filariasis
- Ichthyosis

See page 674 for the diagnosis.

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CMAJ invites submissions to "What is your call?" Clinical details (including images) are presented on the first page along with a multiple-choice question about the diagnosis. The answer and a brief discussion of the condition follow on the second page. We specifically invite submissions illustrating common or important radiographic and electrocardiographic diagnoses of appeal to a general audience. We allow up to 5 references and require authors to obtain consent from the patient for publication of his or her story (form available at www.cmaj.ca/authors/checklist.shtml). Submit manuscripts online at <http://mc.manuscriptcentral.com/cmaj>.

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Discussion

We diagnosed cellulitis complicating underlying elephantiasis nostras verrucosa (a) caused by his lymphedema from congestive heart failure. The patient's condition was initially treated with elevation of the legs and empiric intravenous antibiotics, followed by 2 weeks of oral antibiotics and 4 weeks of topical antifungal creams. His fever and leg pain resolved. The edema of his legs decreased, but the nodularity of his skin did not change.

Elephantiasis can occur because of a variety of obstructive diseases of the lymphatic system. Permanent obstruction of the main lymphatic channels causes progressive enlargement, coarsening, corrugation and fissuring of the skin and the adjacent subcutaneous tissue.¹

Classically, the term elephantiasis verrucosa applies to swelling of the legs caused by infestation by *Wuchereria bancrofti*. In such cases, patients acquire filarial worms in tropical areas where it is endemic. The worms block lymphatic channels, leading to severe chronic swelling in the lower legs and genitals. In 1934, the term "nostras" was added to distinguish lymphedematous disorders of temperate regions not caused by filariasis.²

Elephantiasis nostras verrucosa occurs as a result of lymphedema and recurrent streptococcal infections may also play a critical role. These cause chronic inflammation which in turn leads to fibrosis of the dermis and lymph channels. Any area with chronic lymphedema can be potentially affected.

Table 1: Selected differential diagnosis of elephantiasis nostras verrucosa

Condition	Characteristics
Elephantiasis nostras verrucosa	Severely deformed enlargement of affected limb, thickening and lichenification of the skin, cobblestone-like appearance, foul smell, bacterial and fungal colonization may occur
Venous stasis dermatitis	Medial lower leg often affected, erythema, hyperpigmentation, dilated superficial leg veins, skin induration, lipodermatosclerosis (classic inverted champagne bottle appearance)
Pretibial myxedema	Bilateral pretibial flesh-coloured nodules or plaques
Filariasis	Localized lymphedema in the lower legs and often genitals, recent travel to areas where filarial worms are endemic
Ichthyosis	Chronic, generalized and noninflammatory scaling of the skin, with possible hyperkeratotic changes

The diagnosis of elephantiasis nostras verrucosa is based on the patient's history and characteristic skin changes. Lymphangiography and histopathologic examination of biopsy specimens may provide further information. Computed tomography, magnetic resonance imaging and lymphoscintigraphy may be helpful in a limited number of patients to exclude lymphosarcoma and fibrosarcoma.

The differential diagnosis of elephantiasis nostras verrucosa includes venous stasis dermatitis, pretibial myxedema, filariasis and ichthyosis (Table 1).

Therapies include conservative measures to reduce venous stasis (e.g., elastic bandages, pneumatic stockings, mechanical massage), medical management of edema and prevention of infections (skin hygiene³) and treatment with prolonged courses of antimicrobials, diuretics, topical antibacterial and antifungal agents. However, the chronic nodular skin changes are largely irre-

versible. Retinoids have been also proposed.⁴ Rarely, surgery or amputation is performed in recalcitrant cases with solid edema that is not responsive to medical management.^{1,4}

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