

Verrucous Cobblestonelike Papules and Nodules of the Right Lower Limb

What's the diagnosis?



An obese 58-year-old man was admitted to the cardiology department for poorly controlled congestive heart failure. He was referred to the dermatology department with progressive painless swelling of the right lower limb of a year's duration. He had chronic right lower limb insufficiency of 3 years' duration with a history of a recurrent right medial malleolus ulcer and cellulitis. There was no notable travel or

family history. Physical examination revealed woody edema of the right lower limb with verrucous cobblestonelike papules and nodules, foul-smelling odor, and thick crusts that were easily removed.

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The Diagnosis: Elephantiasis Nostras Verrucosa

Elephantiasis nostras verrucosa (ENV) is a chronic deforming disorder characterized by hyperkeratosis and papillomatosis of the epidermis with underlying woody fibrosis of the dermis and the subcutaneous tissue in the setting of chronic nonfilarial lymphedema. Mossy papules, plaques, and cobblestonelike nodules are classic clinical features of ENV (Figure). The cobblestone lesion often is foul smelling and may be colonized with multiple bacteria and fungi.¹

The differential diagnosis includes filariasis, chromoblastomycosis, and venous stasis dermatitis. Filariasis is infection with the filarial worms *Wuchereria bancrofti*, *Brugia malayi*, or *Brugia timori*. These parasites are transmitted to humans through the bite of an infected mosquito and develop into adult worms in the lymphatic vessels, causing lymphedema. Elephantiasis, the classic sign of late-stage disease, also causes swelling of the legs and genital organs that can be disfiguring. Thus travel history to Southeast Asia and Africa regions is particularly important.

Chromoblastomycosis is a chronic fungal infection of the skin and subcutaneous tissue caused by traumatic inoculation of a specific group of dematiaceous fungi, usually *Fonsecaea pedrosoi*, *Phialophora verrucosa*, *Cladosporium carrionii*, or *Fonsecaea compacta*. Chromoblastomycosis-induced lymphedema may mimic elephantiasis. Thus it is important to perform fungal culture and fungal scraping, which will show typical thick-walled, cigar-colored, sclerotic cells (known as Medlar bodies).



Verrucous cobblestonelike papules and nodules on the right lower limb.

The skin changes of venous stasis dermatitis include edema, varicosities, hyperpigmentation, atrophie blanche, diffuse red-brown discoloration representing deep dermal deposits of hemosiderin, and lipodermatosclerosis. Venous stasis dermatitis is caused by chronic venous insufficiency. It is one of the predispositions to ENV, as it can cause repeated infections and cellulitis leading to blockage of lymphatic drainage and secondary lymphedema. A study of 21 cases highlighted that chronic venous insufficiency may be an underappreciated risk factor in the pathogenesis of ENV.²

The development of verrucous cobblestonelike plaques in ENV occurs with chronic lymphedema. Chronic lymphedema can be caused by a variety of etiologies including infection, chronic venous stasis, congestive heart failure, obesity, trauma, tumor obstruction, and radiation. Our patient was obese with chronic venous stasis, congestive heart failure, and recurrent cellulitis that can lead to secondary lymphatic obstruction and edema, all contributing to the picture of ENV. Because of the unilateral presentation in our patient, we believe that recurrent infections and inflammation are the important risk factors that lead to fibrosis of the dermis and lymph channels, ultimately resulting in ENV.

The diagnosis of ENV is based on the patient's history and characteristic skin changes. However, a biopsy should be performed if the diagnosis is not clinically apparent, if the lesion looks suspicious for malignancy, or if areas of chronic ulceration exist. Histologic findings include hyperkeratosis as well as a collarette of acanthotic epidermis extending around dilated lymph vessels in the upper dermis. Sometimes a few dilated lymph vessels present in the deeper or subcutaneous tissue. Bacterial and fungal culture should be done to exclude concomitant infection. Ultrasonography can be used to evaluate the lymphatic and venous systems.

Treatment of ENV aims at improving lymphedema and preventing infection. Physical therapy to improve lymphedema includes manual lymphatic drainage, massage, compression stockings, elastic bandages, and leg elevation. In cases associated with obesity, weight loss is recommended. In cases of recurrent cellulitis or lymphangitis, long-term antibiotic therapy with agents such as penicillins or cephalosporins is sometimes used.³ Success has been reported with the use of oral and topical retinoids, such as acitretin and tazarotene, which are thought to decrease epidermal proliferation, fibrogenesis, and inflammation.^{4,5} Our patient was started on tretinoin cream. Surgery is reserved for cases of lymphedema resistant to medical therapies, including debridement,⁶ lymphatic and

lymphovenous anastomosis,⁷ lymphatic transplantation, and amputation for severe cases.

Malignancies can develop in areas of chronic lymphedema such as lymphangiosarcoma, squamous cell carcinoma, Kaposi sarcoma, B-cell lymphoma, and malignant fibrous histiocytoma. The morbidity and mortality usually are from the underlying medical problems rather than ENV itself. Our patient died 1 month after the diagnosis of ENV from his poorly controlled heart failure.

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