The Journal of Foot & Ankle Surgery 54 (2015) 747–750

Contents lists available at ScienceDirect

The Journal of Foot & Ankle Surgery

journal homepage: www.jfas.org

Excision of Elephantiasis Nostras Verrucosa Lesions in a Patient With Hereditary Lymphedema: Case Report and Review of the Literature

Austin A. Pitcher, PhD [1], Carlos A. Pagan, MD [2], Kevin Small, MD [2], David M. Otterburn, MD [2]

[1] *Columbia University College of Physicians and Surgeons, New York, NY*
[2] *Weill Cornell Medical College, New York, NY*

ARTICLE INFO

Level of Clinical Evidence: 4

Keywords:
biopsy
elephantiasis nostras verrucosa
lymphedema
surgical excision

ABSTRACT

Elephantiasis nostras verrucosa (ENV) is a rare cutaneous sequela of chronic lymphedema. Treatment of ENV remains poorly elucidated but has historically involved conservative management aimed at relieving the underlying lymphedema, with a few cases managed by surgical intervention. We report a case of a 27-year-old male with primary lymphedema complicated by large painful ENV lesions on his left foot that we excised surgically with good functional and cosmetic results as validated by the patient. To our knowledge, this is the first report of a case of ENV with a pedunculated morphology and the presence of a deep invasive stalk.

Elephantiasis nostras verrucosa (ENV) is a rare cutaneous sequela of chronic lymphedema characterized by hyperkeratosis and papillomatosis of the epidermis with dense fibrosis of the dermis and underlying tissues. Management of this condition is difficult, and no current therapeutic options are completely satisfying (1–3). Recurrence has been the rule. Previously described treatment modalities have included conservative therapy aimed at managing the underlying lymphedema, oral or topical retinoids, and superficial shaving of the lesions. We present the case of a young male with chronic lymphedema who presented to our practice with a number of painful ENV lesions involving the left foot that we treated by deep surgical excision.

Case Report

A 27-year-old male with hereditary lymphedema recently presented to our office complaining of a mass on his left foot that had been increasing in size for the previous 10 years. He described the mass as having been bothersome for the past decade; however, he stated that it had become painful to the point at which he could no longer wear shoes and experienced extreme discomfort with ambulation, significantly impairing his day-to-day activities. He had previously been scheduled to undergo resection of the lesions by his podiatrist; however, the surgical procedure was canceled. He was subsequently referred to our practice because of concern about the high risk of soft tissue infection associated with surgery. The examination revealed an overweight male (body mass index 27 kg/m^2) with a massively dilated left leg and a large fungating, mildly tender mass on the lateral aspect of his left foot. Several smaller masses were also present over his middle toe and heel that the patient described as being new (Fig. 1).

Magnetic resonance imaging of the left lower extremity showed massive swelling of the soft tissues of the dorsum of the foot (Fig. 2). Arising from the lateral aspect of the left fifth toe was an enhancing,

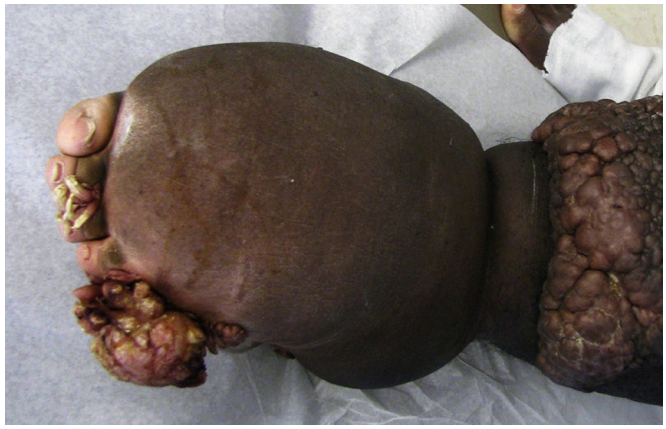

Fig. 1. Preoperative photograph of the patient's left foot showing massive lymphedema with a large mass resembling fingernails arising from the lateral aspect of the left foot.

Financial Disclosure: None reported.
Conflict of Interest: None reported.
Address correspondence to: David M. Otterburn, MD, Weill Cornell Medical College, 425 East 61st Street, 10th Floor, New York, NY 10065.
E-mail address: dmo9004@med.cornell.edu (D.M. Otterburn).

1067-2516/$ - see front matter © 2015 by the American College of Foot and Ankle Surgeons. All rights reserved.
http://dx.doi.org/10.1053/j.jfas.2014.08.010

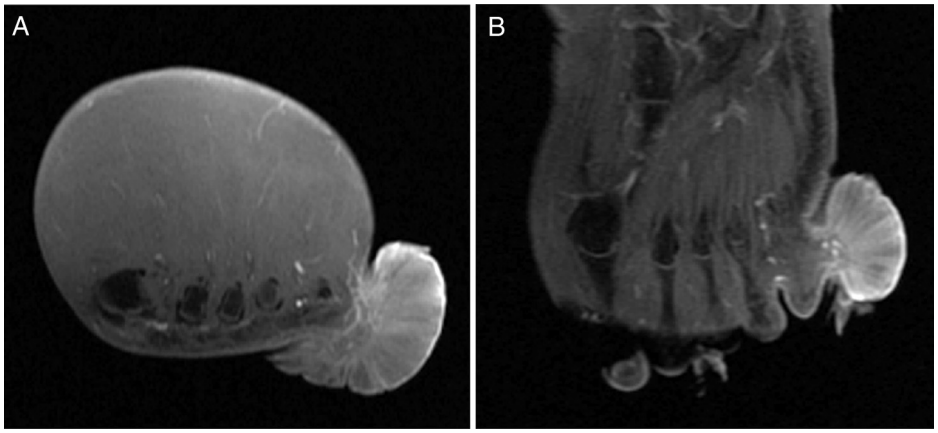

Fig. 2. (*A*) Axial and (*B*) coronal views of T_1-weighted contrast-enhanced magnetic resonance imaging scan of the left foot showing an enhancing, exophytic, fungating, multilobulated mass measuring 3.1 cm in depth and 4.7 × 5.9 cm across. The mass was connected to the toe by a thick stalk but was distinct from the normal-appearing bones of the underlying fifth toe.

exophytic, fungating, multilobulated mass resembling fingernails and measuring 3.1 cm in depth and 4.7 × 5.9 cm in diameter, with a thick stalk connecting it to the toe. A similarly appearing mass was present that extended from the dorsal web between the second and third toes. A smaller mass on the heel was incompletely imaged. Both identified masses were separate and distinct from the normal-appearing bones of the foot (Fig. 2).

After counseling the patient about the high risk of the procedure, he elected to undergo surgical resection of the masses. We excised a 6.5 × 6.0 × 5.7-cm lesion from his left lateral foot and a second 2.0- × 1.3- × 0.3-cm lesion from his left second web space, leaving a 2 × 2-cm and 1 × 1-cm wound, respectively. These were packed with Xeroform® (Invacare Supply Group, Holliston, MA) and allowed to granulate. Histopathologic examination of both masses was consistent with a diagnosis of elephantiasis nostras verrucosa (Fig. 3). The wounds healed without evidence of lymphatic leak or infection, and the patient was happy with the result (Fig. 4). The surgical site was well healed at 8 months postoperatively, with the follow-up examination at 12 months showing no regrowth of the excised lesions. The patient underwent evaluation by a dermatologist at our institution for consideration of cryotherapy for some of these smaller lesions.

Discussion

Lymphedema is an extremely debilitating condition affecting an estimated 140 million people worldwide. It is associated with abnormal function of the lymphatic vessels, leading to an accumulation of fluid in the interstitial space and often to severe dysfunction and recurrent infection in the affected extremities (1,2). Primary lymphedema refers to an uncommon class of conditions with an incompletely understood etiology and is separated into categories according to patient age at the onset of symptoms (3). However, these categories are problematic owing to poorly defined criteria, inconsistent usage within published studies, and imprecise language; also, they have no biologic basis (4). Patients with symptoms present at birth have typically been considered to have congenital hereditary lymphedema (Milroy disease). Although a complete understanding of the pathogenesis of hereditary lymphedema is still lacking, recent studies have implicated defects in the vascular endothelial growth factor receptor subtype 3 as playing a central role (5).

Lymphedema has historically been a difficult condition to manage, often refractory to multiple treatment modalities. Mild cases can be managed with conservative therapy aimed at the reduction of edema, usually with the use of compression from inelastic lymphedema bandaging or serial use of compression garments (2,3). Surgical treatment has been reserved for patients in whom a trial of conservative management has failed and can be divided into 3 broad approaches: resective interventions, liposuction, and microsurgical procedures. Resective approaches have been exemplified by the Charles procedure, first reported in 1912, described as the radical excision of skin and subcutaneous tissue with delayed skin grafting (6). A number of modifications to the Charles procedure and alternative resection techniques have since been published, but the underlying principles remain similar. All resection procedures result in significant morbidity and have had high complication rates (7–9). Suction-assisted lipectomy offers a much less invasive alternative to

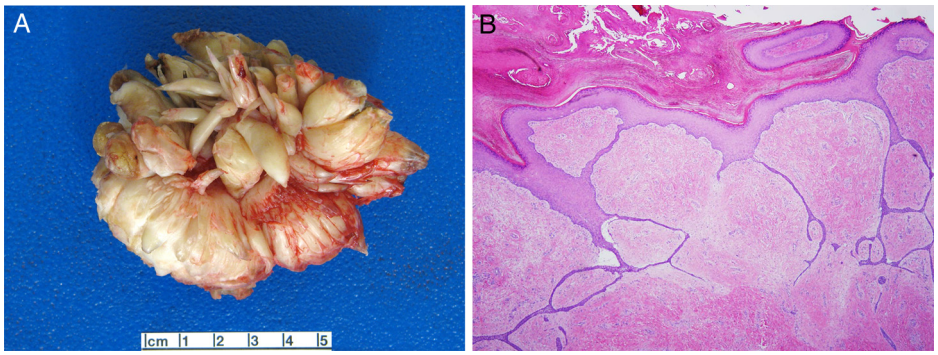

Fig. 3. (*A*) Gross appearance of the larger of the 2 excised masses. (*B*) Representative sections of lesions excised from the left lateral foot. The epidermis shows hyperkeratosis, papillomatosis, and pseudoepitheliomatous hyperplasia and marked fibroblast proliferation and lymphatic dilation.