

Treatment of Elephantiasis Nostras Verrucosa with CO₂ Laser

Abstract

Elephantiasis nostras verrucosa (ENV) is the most severe form of chronic non-filarial lymphedema that leads to disfiguration of body parts. Multiple topical and surgical treatments have been typically used with high relapse rates and local complications. In this context, CO₂ laser emerges as a new well-tolerated and effective treatment option. We report two cases of long-term evolution of elephantiasis nostras verrucosa recalcitrant to topical therapy and successfully treated with CO₂ laser. Both of them were septuagenarians with personal history of obesity and suffered from lower extremities edema due to heart failure in one case, and due to chronic venous insufficiency in the other. We describe the procedure, the results and the advisable maintenance for preventing recurrence in this difficult to treat and chronic condition.

Keywords: Elephantiasis, laser therapy, lymphedema

Introduction

Elephantiasis nostras verrucosa (ENV) is a deforming disorder that occurs as a result of chronic lymphedema which is defined as protein-rich fluid accumulation in the interstitial space secondary to lymphatic drainage blockage. The obstruction of the lymphatic channels can be filarial and non-filarial. Non-filarial elephantiasis includes ENV and other several conditions such as surgery, tumor, trauma, obesity, congestive heart failure, non-filarial infection (mycosis or recurrent cellulitis), and can even be associated with recalcitrant hidradenitis suppurativa.^[1,2] Clinically, ENV is defined by the development of hyperkeratosis, nodular lesions, and fibrosis in the affected area.

Case Report

We present two cases of ENV successfully treated with CO₂ laser. The first patient was a 76-year-old man with a personal history of heart failure, grade 2 obesity, prostate cancer, severe generalized osteoarthritis, and left hip and right knee total replacement. Physical examination showed swollen lower extremities, with firm coalescent skin-colored nodules. A biopsy was performed showing epidermal acanthosis and hyperkeratosis with marked dermal

edema, together with dilated lymphatic channels and fibroblasts proliferation. These findings are compatible with the diagnosis of ENV. In the second case, a 73-year-old woman with a personal history of morbid obesity, type 2 diabetes and chronic venous insufficiency, presented with lesions on her lower extremities that were clinically and histologically compatible with ENV. Positive Kaposi-Stemmer sign can be observed (inability to pinch or pick up a fold of skin at the base of the second toe because of its thickness), which is characteristic of lymphedema.

CO₂ laser treatment was carried out in both patients. Prior to the intervention, the skin was swabbed with chlorhexidine, followed by local anesthesia infiltration with 2% mepivacaine. For the treatment, we used a 10,600-nm CO₂ laser (*Smartoffice-plus; DEKA Medical Electronics Laser Associated. Firenze, Italy*). The laser was set to continuous mode at 4W or 10W output power, depending mainly on the thickness of each lesion. During the procedure, the resulting necrotic tissue was wiped away with gauze pads soaked in physiological saline solution. After treatment, the patients were prescribed oral antibiotic prophylaxis (ciprofloxacin 500 mg twice a day for two weeks) and were instructed to clean the wound with topical mupirocin and change the pressure dressing every

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day for two weeks. Significant cosmetic improvement was observed after a single CO₂ laser treatment session, and the patients were extremely satisfied with the outcome [Figure 1a, b and Figure 2a, b].

Discussion

ENV is the most severe form of chronic non-filarial lymphedema. Prolonged accumulation of protein-rich interstitial fluid induces fibroblast proliferation and weakens the local immune response, leading to dermal fibrosis and increased susceptibility to infection. The incidence of potential complications grows proportionally to the increase in the prevalence of risk factors in the general population, such as congestive heart failure or obesity. The most frequently affected areas are the lower extremities, but involvement of the upper extremities, orbital area, ears, scrotum and abdomen has also been reported.^[3,4]

The differential diagnosis of ENV is broad and includes filariasis, papillomatosis cutis carcinoides, lipodermatosclerosis, pretibial myxedema, Stewart-Treves syndrome, venous stasis dermatitis, deep vein

thrombosis, papular mucinosis, chromoblastomycosis and podoconiosis.^[1] Filarial elephantiasis is a tropical disease which is not endemic in Spain. None of our patients had travelled to endemic areas, and microfilariae were not detected in skin biopsy. Papillomatosis cutis carcinoides is a verrucous carcinoma that usually appears around the mouth, genital area and soles. Biopsy specimens of these lesions show that the epidermis is acanthotic, papillomatous, and consists of large, pale-staining keratinocytes. Little dysplasia or mitotic activity of individual cells is seen. In our cases, the biopsy is not compatible with this cutaneous neoplasm. In contrast with lipodermatosclerosis, our patients did not have the typical tapering of the legs above the ankles and the lesions were not painful. The presence of firm nodules is not a characteristic of lipodermatosclerosis. Pretibial mixedema occurs in association with autoimmune thyroid disorders; mucin deposit is the main histological finding just like in papular mucinosis. Stewart-Treves syndrome consists of a lymphangiosarcoma in the setting of chronic lymphedema and histologically displays networks of small lymphatics and proliferating vascular channels that dissect dermal collagen and may obliterate appendages. Clinically, it appears as a “spreading bruise,” or a raised purple-red papule, eventually developing tissue infiltration and ulceration.

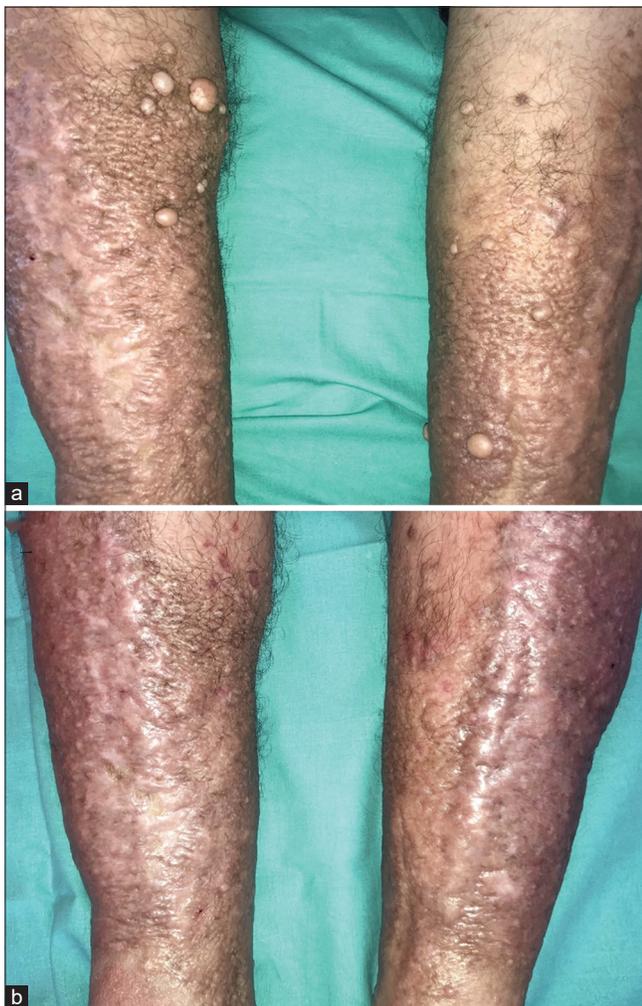


Figure 1: Patient 1. (a) ENV lesions prior to treatment. (b) Good cosmetic result 2 months after CO₂ laser treatment

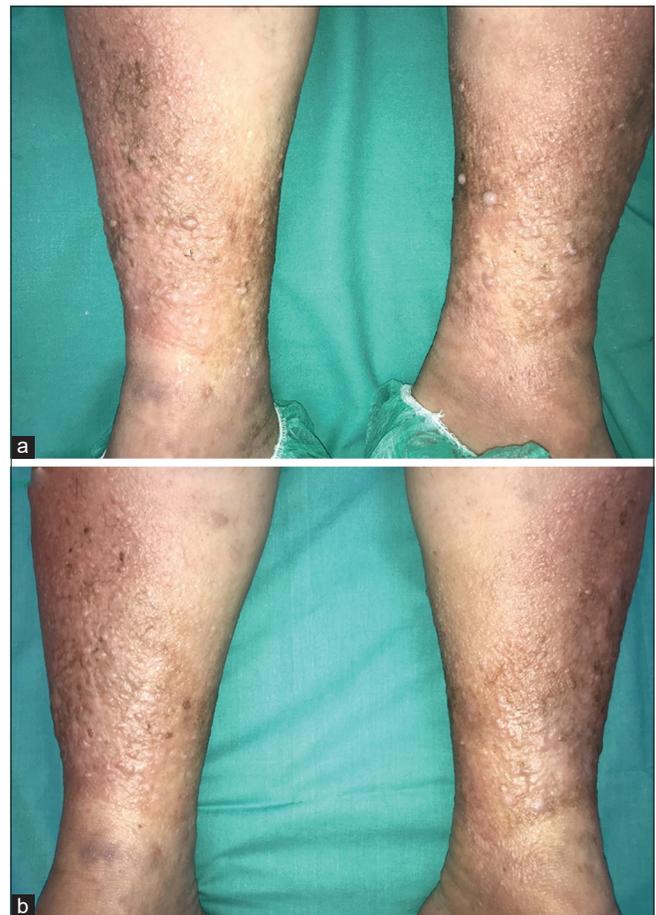


Figure 2: Patient 2. (a) ENV lesions prior to treatment. (b) Good cosmetic result 2 months after CO₂ laser treatment

Chromoblastomycosis is a chronic fungal infection described worldwide but with a higher prevalence in rural populations in countries with a tropical or subtropical climate, which is not the context of our patients. Furthermore, although it can clinically resemble ENV, the histopathologic examination of tissues confirms the diagnosis by demonstrating fungal forms. Podoconiosis also presents with lymphedematous legs but it is a geochemical disease occurring in individuals exposed to red clay soil of volcanic origin.

In both cases, based on the physical examination, the results of the skin biopsy and the personal history, we made a diagnosis of elephantiasis nostras verrucosa (ENV).

However, the treatment of ENV is a challenge. First of all, it is essential to treat the underlying disease if possible. Use of topical corticosteroids, retinoids,^[5] and topical keratolytic agents has been described, together with a wide range of surgical options,^[6-8] which are accompanied by a high rate of complications, and do not rule out the possibility of recurrence. Ablative carbon dioxide laser is able to treat localized ENV due to its dermis-altering capabilities. It causes vaporization and coagulation of the acquired epidermal acanthosis and alters underlying collagen. The thermal damage induced denatures collagen fibers, causing them to shrink, resulting in dermal tightening. The damage also stimulates the remodeling of abnormal collagen by matrix metalloproteinases and dermal neocollagenesis by fibroblasts. The combination of skin tightening and remodeling may lead to the shrinkage of gross ENV lesions. Also, by destroying the superficial reticular dermal lymphatic vessels seen in ENV histologically, laser therapy may prevent future lymphatic leakage, thereby stopping the fibrotic cycle.^[9] Lesions treated with CO₂ laser didn't relapse in our patients after 6 months of follow-up. As primary condition continues to exist, in the future skin manifestations of the disease will probably reappear. For preventing this, we propose a conservative management through proper local hygiene, compression with elastic bandages, weight loss, walking, leg elevation and regular use of a moisturizing cream containing keratolytic agents. Manual and mechanical massage have also been described to be beneficial. Therapy with low-dose acitretin has proven to be helpful in alleviating the symptoms and exudation of the ENV, and it can be maintained for long periods of time without worsening of serum lipids and liver function. Thus, it has been suggested as a complementary option to prevent recurrence.^[10] In case of relapse, there is no objection to use CO₂ laser once again.

As far as we are aware, after carrying out a detailed review of the literature, only two case reports describing the use of CO₂ laser in the treatment of ENV have hitherto been published.^[9,11] Although we cannot be sure that the patients

will not develop new ENV lesions in the future, we consider the CO₂ laser a therapeutic option to be considered in these patients, given its documented efficacy and good tolerance.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that his name and initials will not be published and due efforts will be made to conceal his identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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