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Case Report

Unmasking the unusual: a comprehensive case study of elephantiasis nostras verrucosa on an amputation stump

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ABSTRACT

Elephantiasis nostras verrucosa (ENV) is a rare form of chronic lymphedema that causes progressive cutaneous hypertrophy. It can lead to severe disfiguration of body parts with gravity-dependent blood flow, especially the lower extremities. Various factors can cause obstruction of the lymphatic system and result in ENV. Clinically, ENV is characterized by nonpitting edema and superimposed hyperkeratotic papulonodules with a verrucose or cobblestone-like appearance. Elephantiasis nostras verrucosa can be very difficult to treat, and comprehensive, long-term studies on the management of elephantiasis are unfortunately lacking. A case of 24 years old obese male presented with verrucous growth at a distal part of stump of the left leg since 6 months associated with itching and oozing. Clinical examination revealed pink coloured indurated cobble-stone like papulonodules with overlying crusting. Histopathological examination revealed papillomatosis in epidermis and increased in thick walled capillaries and lymphatics in upper reticular dermis consistant with lymphedema with verrucosa nostra. Treatment with topical keratolytics along with compression stocking is advised. ENV occurring on an amputation stump is scarcely reported, leaving clinicians without clear diagnostic or therapeutic guidance. Timely diagnosis through clinical evaluation and histopathological correlation is essential to distinguish ENV from other verrucous dermatoses and neoplastic conditions. Due to the progressive and disfiguring nature of ENV, early recognition and intervention are crucial in preventing further morbidity. This report adds to the limited literature on atypical presentations of ENV and underscores the importance of individualized patient care.

Keywords: Elephantiasis nostras verrucosa, Amputation stump, Papillomatous skin lesions, Chronic lympedema

INTRODUCTION

Elephantiasis is thought to develop as the result of chronic lymphedema. Lymphedema is categorized into primary and secondary etiologies. Primary lymphedema can be caused by congenital agenesis, hypoplasia, or obstruction of lymphatic vessels, while secondary lymphedema results from a disruption or obstruction of previously normal lymphatic vessels.[1] Worldwide, secondary lymphedema is most often caused by filariasis. Elephantiasis nostras verrucosa (ENV) is uncommon, arising in the setting of chronic nonfilarial lymphedema caused by bacterial or noninfectious lymphatic obstruction.

Elephantiasis nostras verrucosa presents as a grossly enlarged and disfigured appendage, most commonly of the lower extremities and feet, with a cobblestone or mossy appearance. The skin feels "woody," the edema is nonpitting, and does not resolve upon elevation of the extremity. With time, ulceration and crusting may complicate and the lesion eventually becomes chronically colonized by bacteria or fungi and emits a fetid odor.

The diagnosis of ENV is mainly based on patient history, physical examination, and typical cutaneous lesions. To identify causes of secondary lymphedema, skin biopsy and imaging techniques can be necessary.

In the management of ENV, it is crucial to treat the underlying causes. Lymphostasis can be managed conservatively using medical bandages, compression stockings, and mechanical massages. Hyperkeratotic plaques can be treated with topical keratolytics or systemic retinoids.

CASE REPORT

A 24 years old male presented with verrucous growth over the distal part of the stump of left leg. The lesion started developing 6 months before the visit and had progressively increased in size with crusting and oozing since 3 months. The patient had ectrodactyly, a genetic condition that had caused a median cleft of the right hand and hypoplasia of the left leg. The latter led to transfemoral amputation of the left leg when he was 11 years old and had been using Prosthesis since then. His medical history was relevant for morbid obesity (Body mass index -38 kg m^{-2}). The personal and family history was not contributory.

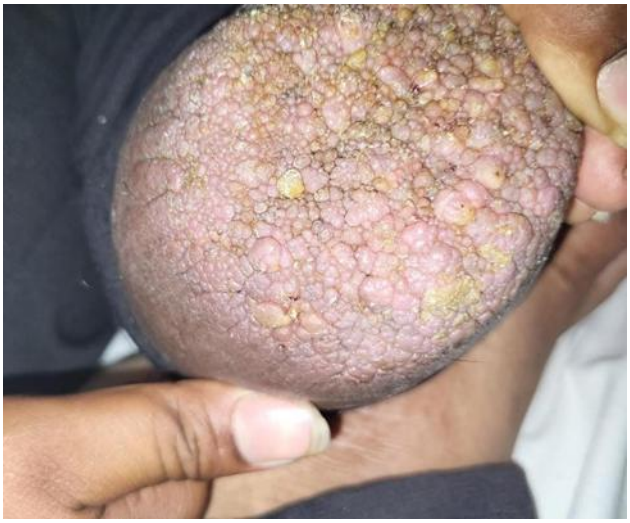

Figure 1: Elephantiasis verrucosa nostra.

On examination, nonpitting edema with pink coloured, cobblestone-like papulonodules over the distal part of the stump of left leg with overlying crusting.

Laboratory investigations, including a complete blood count, liver and renal function test, thyroid function test revealed results within normal limits. Dermoscopy revealed yellow-orange globules, cobblestone appearance, comedo-like openings, yellowish crusting, pinkish background.

Histopathological examination of lesional skin showed marked thickening of papillary dermis with an increased number of thin collagen bundles separated by pale bluish ground substance. The collagen bundles are thickened and arranged in concentric pattern around an increased number of thick walled capillaries and lymphatics in the upper reticular dermis. Also, can be seen in a thick papillary dermis is scant lymphocytic infiltrate. The surface shows papillomatosis and epidermal hyperplasia with orthohyperkeratosis consistent with a diagnosis of lymphedema with verrucous nostra.

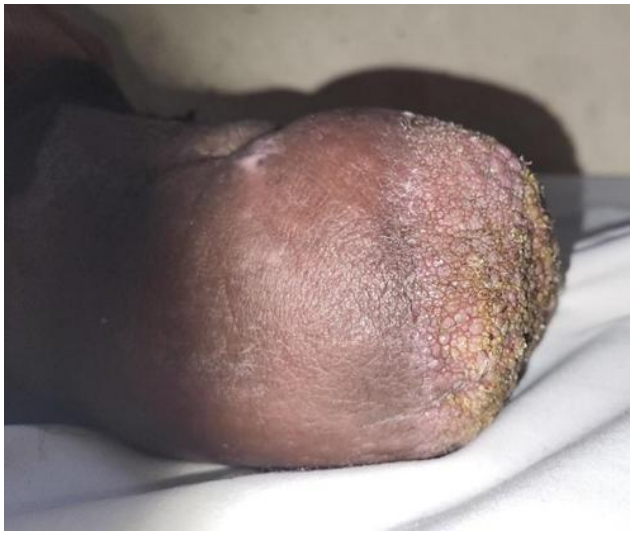

Figure 2: Amputated limb.

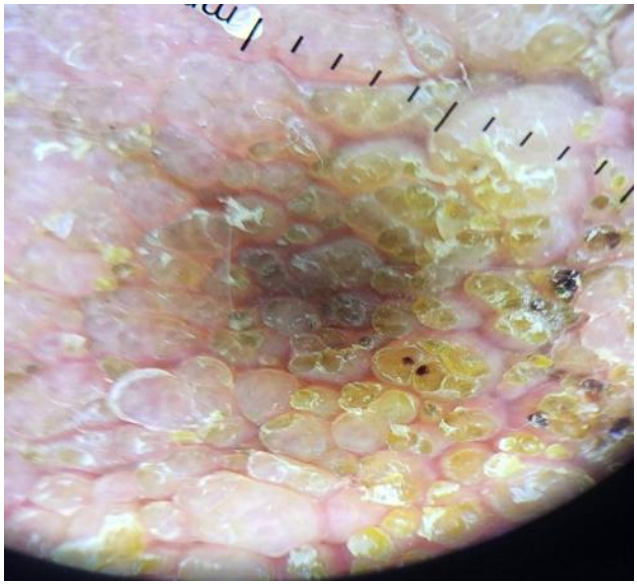

Figure 3: Dermoscopy of ENV.

The patient was managed conservatively with advice for weight reduction, use of compression stockings, systemic antibiotics (Cap. Amoxycillin 500 mg + Potassium Clavulanate 125 mg TDS for 7 days, after food) along with gastric protection (Tab. Pantoprazole 40 mg OD, before food), and twice daily application of 10% urea cream, with follow-up scheduled after one week; however, the patient was lost to follow-up.

DISCUSSION

ENV represents a severe manifestation of secondary, non-filarial lymphedema. A variety of factors that obstruct lymphatic flow—such as malignancy, trauma, radiation therapy, congestive heart failure, obesity, hypothyroidism, chronic venous insufficiency, and filarial infection—can lead to the development of lymphedema.[2,4] The underlying mechanism of ENV involves long-standing cutaneous lymphatic obstruction, resulting in the accumulation of protein-rich lymphatic fluid that promotes fibroblast proliferation and subsequent dermal fibrosis. Persistent lymphatic impairment also compromises local immunity, predisposing the affected area to recurrent infections such as lymphangitis.[5]

ENV typically occurs in gravity-dependent regions of the body, most often affecting the lower limbs. However, it may also involve other sites, including the face, scrotum, abdomen, buttocks, and upper extremities.[6] The disease usually begins with mild, chronic, pitting edema that gradually progresses to thickened, indurated skin exhibiting hypertrophic, verrucous, and cobblestone-like changes.

Diagnosis is primarily clinical, relying on a detailed patient history and characteristic cutaneous findings. To identify the etiology of secondary lymphedema, additional investigations such as skin biopsy, computed tomography (CT), magnetic resonance imaging (MRI), lymphangiography, or lymphoscintigraphy may be warranted.

The differential diagnosis of ENV includes filariasis, chromoblastomycosis, lipodermatosclerosis, verrucous carcinoma, papular mucinosis, and obesity-related lymphoedematous mucinosis.[7,8]

Management of ENV remains challenging, and long-term, comprehensive studies are limited. Addressing the underlying cause of lymphedema is the essential first step in therapy. Once vascular integrity and infection control are ensured, the initial approach should emphasize reduction of lymphedema through manual lymphatic drainage, multilayer inelastic bandaging, and compression garments.[9] Adjunctive pharmacologic measures—such as topical keratolytics and oral or topical retinoids—may further enhance outcomes when combined with physical therapy.[10] In refractory cases, surgical debridement may be considered when conservative or medical interventions fail to achieve adequate results.

CONCLUSION

This case of ENV in a young male with congenital limb malformation and prior amputation to highlight the rare occurrence of ENV developing over a prosthetic stump. The presence of underlying risk factors such as morbid obesity, chronic mechanical trauma from prosthesis use, and preexisting lymphatic compromise made this a unique and diagnostically challenging presentation.

This report emphasizes the importance of recognizing ENV early in atypical clinical settings. Dermoscopy and histopathology were instrumental in confirming the diagnosis, helping differentiate it from infectious or neoplastic verrucous lesions. Timely conservative management with compression therapy, antibiotics, and keratolytics prevented progression and improved local skin condition.

The study aim to raise clinical awareness about ENV, especially in patients with limb deformities, chronic lymphedema, or prosthetic limb use. Documenting such rare presentations will contribute to better diagnostic vigilance and encourage prompt, multidisciplinary management to prevent complications.

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