



ACASE OF ELEPHANTIASIS NOSTRAS VERRUCOSA

Medicine

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KEYWORDS

INTRODUCTION

Elephantiasis nostras verrucosa (ENV) is dermatologic condition, its singular array and rare condition, that can complicate chronic lymphedema (1). It can lead to intense disfiguration of body parts with gravity-dependent blood flow, especially the lower extremities (2). (ENV) characterized by profound hyperkeratosis, plaques, verrucous papules, nodules with underlying woody fibrosis of the dermis and subcutaneous tissue, and lichenification (1,3). (ENV) Caused by assortment of etiologies including, Recurrent cellulitis, infection, tumors, trauma, obesity, congestive heart failure, radiation, and chronic venous stasis (1,4).

Epidemiology data regarding this disorder are bounded to information collected from reports of isolated case typically involving 3 patients or fewer (1).

we present a case of ENV in a 49-year-old female, who had type II diabetes mellitus.

CASE

A 49-year-old female presented to the outpatient department complaining of painful swelling in the right leg for the past several years. She has a history of lymphedema in the right leg for more than 25 years. The reason of Edema beginning is unknown.

She has never suffered from filariasis and has no family history of familial lymphedema. Her medical history revealed type II diabetes mellitus. Her surgical history revealed a hernia repair and a cholecystectomy.

physical examination; showed that her right lower extremity showed diffuse pitting edema admixed with lichenified indurated, cobblestone-like papulonodules; and plaques (Figure 1). lesions extending from the malleolus to several inches below the knee. The surrounding area was erythematous, warm to the touch, and tender.) Kaposi-Stemmer sign (ie, the inability to pinch the dorsal aspect of the skin at the base of the second toe) was present (Figure 2).



Figure 1); diffuse pitting edema admixed with lichenified indurated, cobblestone-like papulonodules; and plaques.



(Figure 2); Kaposi-Stemmer sign

At Laboratory evaluation; Vascular Ultrasound for the lower limb was done to rule out Deep Vein Thrombosis (DVT) and reflux and showed no sonographic evidence of DVT or reflux. Bilateral lower limb lymphoscintigraphy was done and revealed Right lower limb lymphedema (congenital aplasia). This is most likely lymphedema tarda.

DISCUSSION

The common mechanism is an underlying lymphatic obstruction. A variety of etiologies can lead to lymphatic obstruction and edema, and the resultant prolonged accumulation of protein-rich interstitial fluid in the affected area induces fibroblast proliferation and increases susceptibility to infection and inflammation, which in turn leads to further fibrosis of the dermis and lymph channels. The overlying epidermis slowly develops a cobblestoned, verrucous appearance.[1-4] The precise role of some pathogenic organism, either bacterial or filarial, in "idiopathic" lymphatic obstruction is uncertain. Streptococcal and, less often, staphylococcal lymphangitis is considered to be the most likely causative agent.[1,2] Inoculation can result from insignificant trauma, poor hygiene, and dry fissured skin.[3]

ENV is a form of chronic secondary lymphedema; primary lymphedema is caused by congenital defects in the lymphatic system. [5] Secondary lymphedema is the dysfunction of the lymphatic system caused by another primary disease process. Primary infectious etiologies include filarial, staphylococcal, and streptococcal infections, which cause direct inflammation and fibrosis of the lymphatic vessels. Noninfectious etiologies include disruption of the lymphatic system after trauma or surgery; obstruction by malignancy, congestive heart failure CHF, and obesity; and lymphatic fibrosis by malignancy, radiation, venous stasis, congestive heart failure CHF, obesity, portal hypertension, and scleroderma.[1,5] The most common cause of secondary lymphedema in developing nations is filariasis; among industrialized nations it is malignancy.[5] Two other causes of secondary lymphedema congestive heart failure CHF and obesity are on the rise in the United States.[6] It is currently unclear which patients

with chronic lymphedema progress to ENV.[3]

ENV typically appears in gravity-dependent regions, most commonly the lower extremities, but has been reported in the upper extremities, abdominal pannus, buttocks, orbital area, lips, ears, and scrotum.[1,4,7]

Chronic nodular skin changes are almost always progressive, and management of advanced stages usually results in unsatisfactory outcomes. Early diagnosis is paramount. Without appropriate early intervention, ENV continues to worsen and may result in gross deformity requiring amputation.[1,8]

By history and physical examination alone we can diagnose ENV, but there are other tools that can assist the physician. In physical examination we can find stemmer sign which is inability to pinch dorsal skin of the toes because of thickening of it due to lymphedema. We can evaluate lymphedema and rule out other cause by some diagnostic studies like the following : 1- biopsy can rule out other causes of secondary lymphedema such as malignancy ; histologically there is hyperkeratosis , loss of dermal papillae , fibrosis of dermis and SC tissue and widened lymphatic vessels. 2- lymphoscintigraphy for assessment of lymphedema. 3- U/S shows filaria in lymphatics. 4- CT and MRI are used to rule out malignancy.[1,9]

The differential diagnosis for ENV is broad and is listed in Table 1.

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| Primary (congenital) lymphedema | Congenital abnormality in formation or function of lymphatic vessels and has a morphology similar to that of ENV. Onset is usually before the age of 25 and there is often a family history of lymphedema. |
| Lipedema | Early age of onset; positive family history; typically affects overweight women; abnormal accumulation of subcutaneous fat in lower limbs and buttocks; always symmetric and bilateral; feet not affected 1,4,5 |
| Lipodermatosclerosis | Related to venous stasis, hyperpigmentation, and nonpitting edema with subcutaneous fibrosis. Characteristic "inverted wine bottle" appearance: swelling of proximal parts and fibrosis of the distal parts of the lower limbs 1,4 |
| Venous stasis dermatitis | Pitting edema, erythematous to brownish pruritic patches with dilated superficial veins over the medial lower leg 1-3 |
| Pretibial myxedema | Caused by hyperthyroidism. Mucin accumulation of the dermis resulting in nonpitting edematous papulonodules or plaques over anterior surface of the legs and dorsal aspect of the feet 1,4 |
| Filariasis | Infection by the parasite <i>Wuchereria bancrofti</i> . History of travel to an endemic tropical area. Obstruction of lymphatic ducts causes secondary bacterial infection. Localized lymphedema in the lower legs and genitals 1,4 |
| Chromoblastomycosis | Chronic fungal infection; fungus is commonly found in soil. Infection through minor trauma of skin. Verrucose papulonodules and plaques. Cultures are positive for fungus 4 |
| Papillomatosis cutis carcinoides | Morphology similar to that of ENV, typically presents with ulcers, and is considered a low-grade squamous cell carcinoma. Distinguished from ENV by biopsy. |
| Stewart-Treves syndrome | Angiosarcoma that develops in chronic lymphedema and presents as an erythematous mass. Seen in patients after radical mastectomy and in congenital lymphedema. |
| Deep venous thrombosis | Acute onset of pitting edema, typically in lower extremities, with no epidermal changes. |

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| Papular mucinosis | Idiopathic disease distinguished by acid glycosaminoglycan infiltration of the papillary dermis. Presents clinically as waxy papules that can coalesce into plaques. |
| Podoconiosis | Lymphatic obstruction caused by chronic, barefoot walking on volcanic soils with high silica dust content. |

The differential diagnosis for ENV source .[6]

In our study patient can't recall how the edema started. Also She has never suffered from filariasis and has no family history of familial lymphedema. Also not suffer from heart problems or obese. She reported that she didn't travel out of the country before. But she had a hernia repair and a cholecystectomy surgery before. Hence the precedent surgery may played a key role in the occurrence of disease.

Regarding the treatment, currently there's no established standard of care for the treatment of ENV. Therapies are just attempt to correct the underlying cause, increase the ability of patient to use the affected limb, and prevent additional complications.[9]

Initial conservative treatments such as; rising the affected limb, compression using bandages or stockings, pneumatic compression devices, and massage.[8]

In case of acute lymphangitis, should use suitable antibiotics to treat the infection. But When there is no active infection, prophylactic use of antibiotics may be considered. There are many reports of success with the use of topical and oral retinoids, such as tretinoin and tazarotene, respectively, that's decrease epidermal proliferation, fibrogenesis, and inflammation. For cases that have not responded well to conservative or medical treatment, surgical intervention should be attempted. surgical intervention include lymphatic and lymphovenous anastomosis, debridement, lymphatic transplantation, while for in which the outlook of limb preservation is bleak, amputation.[10]

Because of grotesque features and embarrassing Patient's anxiety ,depression, and social isolation can be developed. Patients may suffer from disability due to the loss of use of the affected limb. With further progression, fungal and bacterial super-infections of the skin and underlying bone can develop. Ultimately patient may die because of sepsis associated with ENV or from the advancement of their primary condition [1, 11].

CONCLUSION

Our patient don't remember how the edema started, has never suffered from filariasis and has no family history of familial lymphedema. But she had a hernia repair and a cholecystectomy surgery before. Therefore we believe that the cause ENV could be the surgery. Regrettably, ENV is a disease without well-studied, standardized, or effective treatment, and its prognosis is poor. Also different conditions can cause ENV. But history taking and physical examination are sufficient to diagnose ENV. Laboratory tests and imaging studies will provide more information and assist physicians in differentiating ENV from other diseases.

Recommendation

- recognize this rare condition in its initial stages and control infection and edema as soon as possible to prevent debilitating deformities.
- the underlying cause of lymphatic obstruction should be investigated and treated to the maximal extent possible.
- Stay away from sources of filarial as a precaution especially at travel to tropical regions .
- Sterilize wounds directly to prevent the entry of bacteria.

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