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ELEPHANTIASIS NOSTRAS VERRUCOSA MIMICKING DEEP FUNGAL INFECTION – A CASE REPORT.

Medical Science

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ABSTRACT

Elephantiasis nostras verrucosa is a rare condition characterized by severe irreversible lymphedema of skin with no premorbid abnormality of lymphatics of the dependant parts especially lower limb or other and immobile body parts & it is associated with supervening recurrent cellulitis or secondary infection. ENV of left foot mimicking deep fungal is reported in a 40 yr old male from border city of Punjab.

KEYWORDS

Elephantiasis nostras verrucosa (ENV), lymphedema, Edema.

INTRODUCTION

Elephantiasis nostras verrucosa synonymous with lymphangitis recurrens elephantogenica is a rare, disfiguring skin condition in which the skin has woody indurated appearance with verrucoid pebbled plaques. There is progressive disfiguring enlargement of the body part. With recurrent soft tissue infection lymphatics become increasingly fibrotic and affected body part becomes more edematous & enlarged even after subsidence of acute infection, subsequently skin undergoes epidermal hyperkeratosis with fibrosis of dermis & subcutaneous tissue with resultant ENV which is an enlarged body part with woody edema.

ENV is a rare clinical entity that occurs due to chronic obstructive lymphedema. Characteristically the edema is non pitting with grotesque enlargement of body part and overlying skin becomes hyperkeratotic and lichenified. ENV occurs due to mycotic, syphilitic, tuberculoid, neoplastic or traumatic causes of lymphatic obstruction (Castellani1969)[1]. Factors which precipitate ENV are recurrent cellulitis, chronic venous stasis & prolonged standing. Lymphedema may also arise from dysfunctional lymphatics when patient sits on chair continuously 'armchair legs'[2]. There is minimal lymph drainage leading to pathological changes in lymphatics and later irreversible lymphedema[3-5].

CASE REPORT

40 yr old moderately built male patient presented with asymptomatic nodular lesions along with swelling of left foot since 2.5yrs. Patient started developing papulonodular lesions on the left foot insidiously **(Fig.1&2)** with history of recurrent cellulitis. He had fracture left femur 15 years back due to trauma after falling from height . Patient was operated upon and had difficulty in walking. Slowly the overlying skin became hypertrophic with gradual swelling of left foot and lower half of left leg. On examination multiple verrucous spherical nodules of variable size 1cm -3cm diameter over dorsum of foot moreso over the dorsal aspect of ankle joint & metatarsophalygeal joints showed hyperkeratotic surface, hyperpigmentation with loss of hair and thickening of skin. Skin over the left lower leg was dark, puckered with peau d orange appearance. Oedema was non-pitting in nature. Patient was normotensive, various routine investigations were normal including ESR, PBF & ECG. Fungal element was not found on skin scrapings and culture. H/P examination showed mild to moderate papillomatous hyperplasia in the epidermis **(Fig.3)**. Dermis shows moderate infilteration by mononucleear cells, focal small collection of neutrophils. Color Doppler of the left leg was conducted which showed normal arterial flow pattern and velocities with no evidence of intimal calcification. There is evidence of s/c edema with venous stasis. Patient was advised to apply emmolients & keratolytics then reffered to surgery for surgical intervention.

DISSCUSSION

ENV is a form of chronic secondary lymphedema which is dysfunction of lymphatic system caused by another primary disease process in which the overlying skin becomes verrucous cobblestone like with woody edema. Primary infections include filarial, staphylococcal and streptococcal infections. Non infectious etiologies include disruption of lymphatic system after trauma or surgery, obstruction by malignancy, CHF, obesity and lymphatic fibrosis by malignancy, radiation, venous stasis, portal scleroderma and surgery. The most common cause of secondary lymphedema in developing nations is filariasis and among industrialized nations is malignancy. In 1934 word nostras was used by Castellani to distinguish it from filariasis[6]. Sites involved are mainly extremities, gravity dependant , less commonly pendulous abdomen[7], periorbital region, buttocks, scrotum, lips, ears. Complications are mainly due to swelling and infection. Swelling leads to discomfort, limb heaviness, reduced motility and impaired function. Thickening of skin causes pseudoscleroderma and impaired small joint motility. Difficulty in finding proper shoes and clothes creates soc due to streptococcus[8] are precipitated by strenuous exercise or journey as the microorganisms are always present and becomes reactivated like herpes simplex. Local immune deficiency is a contributing factor in the development of these changes. Repeated infection may stimulate proliferation of keratiocytes which may contribute to hyperkertosis and fibroblast proliferation. Hence lymphatics become fibrotic with fibrosis of dermal and subcutaneous tissue also extravascularly leucocytes from inflamed vasculature migrate out with release of TGF@ which further stimulate production of collagen by fibroblasts[9].

ENV can be diagnosed with history and physical examination alone. On physical examination Kaposi- Stemmer sign i.e . inability to pinch the skin on the dorsal toes because of thickened skin is suggestive of lymphedema[10]. Biopsy may be helpful to find out other causes but not diagnostic of ENV. On histopathology ENV shows hyperkeratosis , pseudoepithelial hyperplasia, loss of dermal papillae, fibrosis of dermis and subcutaneous tissue and widened lymphatics. Radiological investigations for malignancies were normal.

Differential diagnosis include filariasis, chromoblastomycosis, congenital lymphedema, lipodermatosclerosis, pretibial myxedema, Stewart-Treves syndrome, mucinosis, podoconiosis, papillomatosis, cutis carcinoides, verrucous carcinoma.

Treatment is by correction of underlying cause. Care of skin and prevention of infection with prolonged course of antibiotics. Regular application of emmolients hydrate the hardened skin. Hyperkertosis can be improved with regular application of 5% salicylic acid ointment. Compression with bandage or stockings, massage, pneumatic compression devices. Topical retinoids decrease epidermal proliferation, fibrinogenesis and inflammation[11]. Exercise and movement are crucial to lymph drainage. Surgical option of treatment is of value in patients with interference of mobility even after conservative treatment[12]. Prognosis is based on severity of ENV and the primary condition. Our case was having history of recurrent attacks of cellulitis with increase in swelling with subsequent attacks and therefore development of nodules. Patient's history of fracture femur may have damaged draining highlight a rare and interesting presentation.

It is important to be aware of this entity. Precise evaluation is required to recoginize the condition in early stages to prevent debility or deformity,

control of infection and edema and maximize patient's functional capacity .As the disease is progressive it's progression should be curtailed by early intervention. Management in advanced stages results in unsatisfactory outcome.

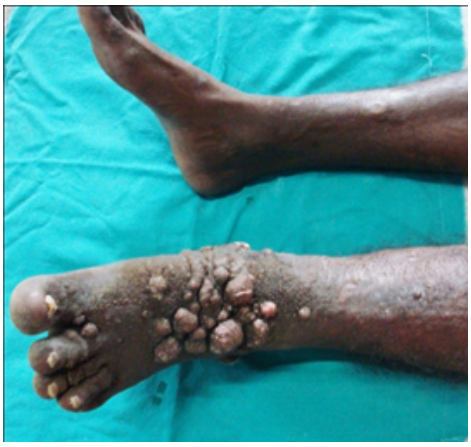

FIG.1 Left foot of the patient having Papulonodular verrucous growth.

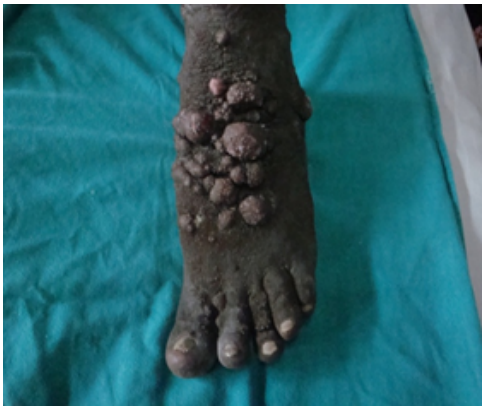

FIG.2 superior view of left foot

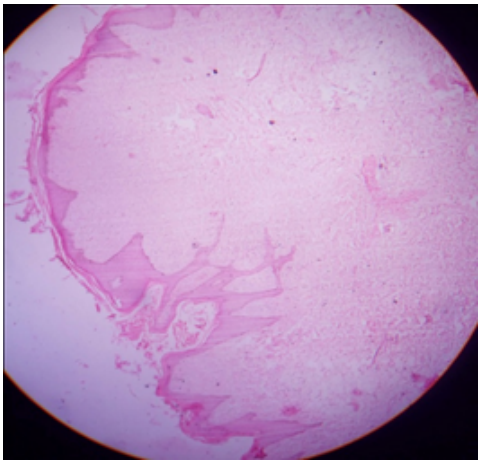

FIG.3 5x view of the biopsy of the lesion shows hyperkeratosis, psedoepithelial hyperplasia, loss of dermal pappilae and fibrosis of dermis.

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