

Elephantiasis Nostras Verrucosa Cutis

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Elephantiasis nostras verrucosa cutis (ENV) is a rare presentation of chronic lymphedema which affects gravity dependant body parts - especially lower limbs. It is clinically characterised by non-pitting edema and progressive cutaneous hypertrophy leading to hyperkeratotic papulonodules which causes a verrucose or cobblestone like appearance [1]. Differential diagnoses include lymphatic filariasis, chromoblastomycosis as well as non-infectious conditions like lipodermatosclerosis, pretibial myxedema, lipidema and venous stasis dermatitis [1,2,3]. Although the exact pathogenesis is still not known, it is believed that as the lymphatic channels are damaged and blocked, excessive protein rich fluid accumulates in the dermis and subcutaneous tissues. This leads to decreased oxygen tension leading to decreased local skin immunity. As a result, there is increased susceptibility to infections. All these ultimately leads to swelling, fibrosis and disfiguration of the area and a vicious cycle ensues [1,4].

Our case is of a 42 years normotensive, non-diabetic male diagnosed as lymphatic filariasis 10 years ago. He presented with progressive swelling of both lower limbs, left more than right with multiple clustered small nodules each of few millimetres in size affecting left lower limb. He had no other comorbidities. On physical examination, his lower extremities showed bilateral non-pitting edema, lichenification and hyperpigmentation with induration of skin (Figure 1, 2).

There were non-tender multiple nodules which were firm in consistency in left lower limb extending below the knee. Both the lower limbs were non-erythematous and temperature was not raised. His blood tests which included complete blood count, liver function tests, lipid profile, renal function tests did not reveal any abnormality. C-reactive peptide was also not raised. Excision biopsy of few nodules with histopathological examination showed hyperkeratosis, papillomatosis, pseudoepitheliomatous hyperplasia and extensive fibrous tissue hyperplasia (Figure 3). Gram stain and fungal stains did not reveal any microorganism. Circulating filarial antigen was negative and microfilaria could not be detected from peripheral blood. Doppler ultrasonography of lower limbs revealed dilated lymphatic channels. Arterial flow was normal but venous flow was sluggish. No thrombi were detected. The patient was given loop diuretic- torsemide and referred to plastic surgery department.

Regarding the management, it is vital to treat the underlying conditions. Conservative non pharmacological measures include compression stockings, medical elastic bandages. Diuretics also have some role in reducing edema. Topical keratolytics also have some benefit for the hyperkeratotic plaques. Surgical intervention, is however the treatment of choice in advanced cases [1]. As outcomes are not satisfying in advanced cases with late presentation

and owing to cumbersomeness of plastic surgical interventions, early diagnosis on part of all physicians and primary health care providers is warranted so as to prevent progression and reduce morbidity.



Figure (1)



Figure (2)

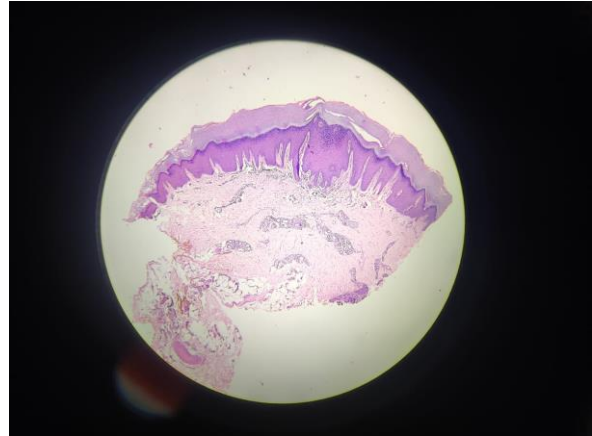


Figure (3): Histopathological slide showing hyperkeratosis, hyper granulosis, acanthosis, papillomatosis, mixed inflammatory infiltrate in upper dermis and perivascular areas.

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Authors' Declaration: The authors declare that the image has not been published before.

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