Elephantiasis nostras verrucosa secondary to congestive cardiac failure

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DESCRIPTION

A 70-year-old woman with ischaemic cardiomyopathy was admitted and treated for decompensated congestive cardiac failure (CCF). She had previously received treatment for recurrent episodes of lower limb cellulitis and reported progressively worsening swelling interfering with her daily activities. She had no history of filariasis or Milroy’s disease.

Cutaneous examination showed grossly oedematous lower limbs with a woody, indurated feel on palpation (figure 1). There were multiple, tiny verrucous papules interspersed with postinflammatory hyperpigmentation, macerated web spaces and a positive Kaposi-Stemmer sign (figure 2). The patient was treated for CCF and lower limb cellulitis.

First described as elephantiasis nostras verrucosa (ENV) by Castellani in 1934, ENV is characterised by hyperkeratosis, papillomatosis and verrucous changes leading to grotesque, disfiguring cutaneous hypertrophy of the involved body part, most likely lower limbs. Aetiologies for ENV include CCF, radiation, pretibial myxoedema, portal hypertension, obesity, tumour or surgical damage to the lymphatic system. Although the exact pathogenesis of ENV is unclear, a major contributing factor is a vicious cycle of chronic lymphoedema and repeated infections. An important differential diagnosis of ENV is papillomatosis cutis carcinoides.

Initial treatment is by simple measures such as elevation of limbs and pneumatic compression devices, with a few cases of success using topical and oral retinoids, and surgical interventions in advanced cases. Progressive ENV and recurrent bouts of osteomyelitis can cause limb amputations and fatalities from sepsis.

The lack of well-studied, easily accessible, established treatment strategies for ENV highlights the importance of primary prevention.

Learning points

▸ The rise in incidence of elephantiasis nostras verrucosa (ENV) is associated with an increasing prevalence of cardiac failure and obesity.
▸ Prompt identification and treatment in early stages is crucial to prevent the morbidity and mortality associated with ENV due to the lack of definitive treatment strategies in established cases.

Contributors SM was involved in the literature search, preparing the figures and writing the manuscript. FZA was the consultant looking after the patient and involved in patient consent and final review.

Competing interests None declared.

Patient consent Obtained.
REFERENCES

