Elephantiasis nostras verrucosa secondary to recurrent erysipelas

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DESCRIPTION
A 79-year-old man presented with worsening lower leg oedema, nausea, prostration, fever and chills. He had history of several episodes of erysipelas during 10 years, chronic venous insufficiency and type 2 diabetes mellitus. Physical examination revealed lesions of a mossy and verrucose appearance, with exophytic and agglomerated lesions on the left lower limb (figure 1). The dermatological findings of epidermal thickening, lymphoedema and fibrosis of the dermis and subcutaneous tissue were consistent with the final stage of erysipelas in the rare form of elephantiasis nostras verrucosa (ENV). The patient received intravenous furosemide and heparin to control oedema and to prevent deep vein thrombosis and antibiotic therapy.

ENV is a serious complication of chronic lymphoedema that causes progressive cutaneous hypertrophy.1 Lymphoedema, if left untreated, causes fibrosis and sclerosis, which when is exuberant makes the skin verrucose and with a mossy appearance (figure 1). ENV is often observed in gravity-dependent blood flow parts of the body, especially in the lower extremities.2

History and physical examination are sufficient to diagnose ENV but laboratory tests and imaging studies may be used to exclude malignancy and to differentiate ENV from other diseases. Treatment should focus on reducing lymphostasis and preventing recurrent infection.3

The underlying causes of lymphatic obstruction must be investigated and treated and the complications associated with lymphoedema prevented. It is important that physicians recognise this unusual pathology in its initial stage to prevent debilitating deformities and impairment of the involved limb.

The chronic lymphoedema due to erysipelas may lead to deformities evidencing the manifestation of elephantiasis, which in its verrucose form is characterised by hyperkeratosis and papillomatosis of the epidermis, associated with dermis and subcutaneous fibrosis.

Learning points

► The exact pathogenesis of elephantiasis nostras verrucosa (ENV) is still unclear, but a vicious cycle of chronic lymphoedema and repeated infections is a very important contributing factor. So, the underlying cause of lymphatic obstruction should be investigated and treated to the maximal extent possible.

► Considering the lack of definitive treatment strategies in established cases, prompt diagnosis and early treatment are needed to prevent debilitating deformities associated with ENV.

► This clinical case aims to alert to the existence of this rare pathology and that it is poorly reported in the literature, and to create an index of suspicion necessary for its diagnosis.

Contributors All authors know and agree with the case described. AF collected the information and wrote the article. JFMR reviewed the work.

Competing interests None declared.

Patient consent Detail has been removed from this case description to ensure anonymity. The editors and reviewers have seen the detailed information available and are satisfied that the information backs up the case the authors are making.

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REFERENCES

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