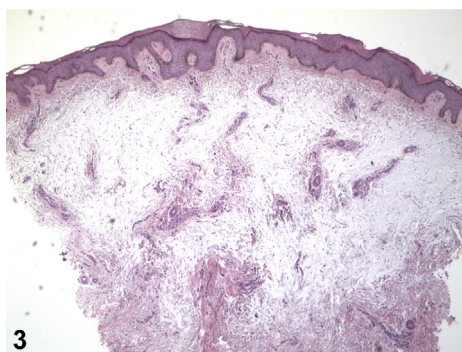


Edematous lower extremities with overlying verrucous plaques



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A 45-year-old African American man with a past medical history of Graves disease (GD) presented to the dermatology clinic for evaluation of a 10-year history of painful lesions on his bilateral lower legs. The patient reported symptoms of lower extremity edema, which worsened by prolonged standing. Treatments included

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compression stockings without improvement. A physical exam demonstrated firm, non-pitting edema and hyperpigmentation of his bilateral lower legs with many verrucous papules and plaques (Fig 1). There was no thyroid enlargement; however, prominent digital clubbing and exophthalmos were noted (Fig 2). A punch biopsy of his right shin was performed (Fig 3).

Question 1: Based on the clinical presentation, history, and histology, what is your diagnosis?

- A. Elephantiasis nostras verrucosa
- B. Elephantiasic pretibial myxedema
- C. Lymphatic filariasis
- D. Lymphedema, not otherwise specified
- E. Papillomatosis cutis carcinoides

Answers:

A. Elephantiasis nostras verrucosa – Incorrect. While this patient’s clinical presentation closely resembles elephantiasis nostras verrucosa with enlarged, disfigured extremities with a fibrotic, cobblestone-like appearance, the presence of digital clubbing and exophthalmos hints toward a different diagnosis.

B. Elephantiasic pretibial myxedema – Correct. Elephantiasic pretibial myxedema presents with verrucous papules and plaques, most commonly on the anterior aspects of the lower legs, on a background of non-pitting edema. The presence of digital clubbing and exophthalmos further supports a diagnosis of underlying GD. Histological examination demonstrates abundant mucin accumulation in the dermis, collagen attenuation, and stellate fibroblasts.¹

C. Lymphatic filariasis – Incorrect. Filariasis is a parasitic infection, common in tropical countries, and caused by the filarial parasites *Wuchereria bancrofti* or *Brugia malayi/timori*, presenting as elephantiasis. While clinically, the condition may be similar to the one presented here, the patient’s history does not support a diagnosis of filariasis.

D. Papillomatosis cutis carcinoides – Incorrect. Papillomatosis cutis carcinoides describes a form of verrucous carcinoma, secondary to the human papilloma virus, which presents as verrucous papules and plaques involving the lower extremities. While the clinical appearance may be similar to the condition presented here, the disease is often unilateral and asymmetric, and the histologic findings in this case do not support the diagnosis of papillomatosis cutis carcinoides.

E. Lymphedema not otherwise specified – Incorrect. Lymphedema not otherwise specified can

present with unilateral and bilateral lower extremity swelling. Without proper treatment, it may take on a similar presentation. However, the histopathologic findings do not support this diagnosis.

Question 2: What is the best initial treatment for the management of elephantiasic pretibial myxedema?

- A. Pentoxifylline
- B. Rituximab
- C. Iodine-131
- D. Complete decongestive physiotherapy
- E. Topical or intralesional corticosteroids.

Answers:

A. Pentoxifylline – Incorrect. Pentoxifylline inhibits the proliferation of fibroblasts and production of glycosaminoglycans and may be used in the treatment of severe pretibial myxedema²; however, it is not the best initial treatment.

B. Rituximab – Incorrect. In combination with plasmapheresis, rituximab has demonstrated success in the treatment of pretibial myxedema. However, this treatment option is aggressive and is not the initial step in management.

C. Iodine-131 – Incorrect. A case report from 2014 noted improvement in elephantiasic pretibial myxedema following treatment of underlying GD with iodine-131.³ However, this is not the best initial treatment option.

D. Complete decongestive physiotherapy – Incorrect. While complete decongestive physiotherapy has demonstrated success in the management of lymphedema and filariasis, it is not a first-line treatment for pretibial myxedema.

E. Topical or intralesional corticosteroids – Correct. The management of pretibial myxedema is difficult. However, the rare elephantiasic variant poses a greater therapeutic challenge. The initial treatment is medium-to-high potency topical or intralesional steroids, which may be used with or without an occlusive dressing.⁴ Unfortunately, the disease is often refractory to topical steroids and patients often report little-to-no improvement.

Question 3: Which of the following is true regarding elephantiasic pretibial myxedema?

- A.** The disease is most often found in association with hypothyroidism
- B.** While there are different presentations of pretibial myxedema, the most uncommon is the elephantiasic variant
- C.** The disease is often responsive to topical corticosteroids and supportive treatment
- D.** Elephantiasic pretibial myxedema resolves after control of underlying thyroid disease
- E.** The main cause of elephantiasic pretibial myxedema is chronic lymphedema

Answers:

- A.** The disease is most often found in association with hypothyroidism — Incorrect. Elephantiasic pretibial myxedema is typically a result of longstanding GD with associated hyperthyroidism. However, in rare cases, it may be associated with non-thyrototoxic GD or Hashimoto thyroiditis.
- B.** While there are different presentations of pretibial myxedema, the most uncommon is the elephantiasic variant — Correct. There are four distinct variants of pretibial myxedema: non-pitting, plaque-like, nodular, and elephantiasic.⁵ The elephantiasic variant is the most uncommon and it occurs in less than 1% of patients with pretibial myxedema.⁵
- C.** The disease is often responsive to topical corticosteroids and supportive treatment — Incorrect. The initial management of elephantiasic pretibial myxedema involves topical and intralesional corticosteroids. However, the disease is often treatment refractory.

D. Elephantiasic pretibial myxedema resolves after control of underlying thyroid disease — Incorrect. While mild cases of pretibial myxedema may improve with control of underlying thyroid disease, the elephantiasic variant is typically treatment refractory. Further, there is no demonstrable relationship between the management of thyrotoxicosis and the improvement of elephantiasic pretibial myxedema.⁴

E. The main cause of elephantiasic pretibial myxedema is chronic lymphedema — Incorrect. Elephantiasis refers to the enlargement and hardening of an anatomic region, which may have various pathogenic causes. While lymphedema contributes to many forms of elephantiasis, it does not contribute significantly to elephantiasic pretibial myxedema.

Abbreviation used:

GD: Graves disease

Conflicts of interest

None disclosed.

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