Worsening indurated pink translucent nodules and severe hyperkeratosis of the lower extremities: A Case of Elephantiasic Pretibial Myxedema.

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LEARNING OBJECTIVES

- Recognizing elephantiasic pretibial myxedema (PTM)
- Understanding pathophysiology of elephantiasis PTM.
- Creating treatment plan for patient suffering from elephantiasis PTM.
- Dx for elephantiasis PTM.

CASE PRESENTATION

- HPI: A 61 year old white woman presented with bilateral lower extremity dermatitis, swelling and skin thickening that began 5 years ago; shortly before she was diagnosed with Graves disease (Figures 1-3).
- Patient’s symptoms progressively worsened post thyroidectomy and achievement of euthyroid state with levothyroxine. Previous diagnoses included cellulitis and lymphedema treated with multiple failed attempts of oral antibiotics. No family history of related conditions. No previous biopsy was obtained.
- PHYSICAL EXAM: Indurated, 1-2cm thick violaceous plaques with interspersed pink translucent nodules; associated deep fissures with active serous drainage and overlying yellow-white crust on bilateral pretibial areas, ankles and dorsal feet. (Figures 1-3) Planar surface was covered by thick scale.
- Other physical exam findings included proptosis, exophthalmos and surgical scar on the anterior neck.
- DX: After obtaining informed consent 2 biopsy specimens were obtained for hematoxylin-eosin and other special stains (Figures 3-6).
- Elephantiasis pre tibial myxedema was diagnosed based on these clinical and histological findings.

DISCUSSION

- EPIDEMIOLOGY: elephantiasic pretibial myxedema (PTM) is the most severe variant of non-fibular myxedema occurring in only 1% of patients with Grave’s disease.
- PATHOPHYSIOLOGY: It is theorized that T-cells stimulate shared antigens between the thyroid and pretibial tissue and release TGF-B and IL1-alpha that stimulate fibroblasts to produce and deposit mucin-like glycosaminoglycans in tissue. The pretibial fibroblasts may be more sensitive to this stimulation.
- The Pretibial area is favored secondary to hydrostatic forces, decreased lymphatic system clearance and dependent position.
- CLINICAL: Grossly enlarged and disfigured appendage, usually with functional restriction and cosmetic concerns for the patient. Cutaneous changes include non-pitting edema of lower extremities that does not resolve with elevation. The initial cobblestone appearance later becomes mossy and verrucous. Because hair follicles are prominent, it produces the characteristic d’orange appearance.
- Ulceration and bacterial seeding with recurrent cellulitis or fungal infections are common, with patients complaining of pain or pruritus.

- PATHOLOGY: Large amounts of mucinous deposition are seen in the reticular dermis. There is a lack of angioplasia and hemisiderin. Sparse lymphocytic deposition in perivascular spaces and moderately increased mast cell deposition are seen.
- The number of collagen fibers is reduced with increased edema, and occasional acanthosis, hyperkeratosis, and papilomatosis.
- TREATMENT: Cosmesis and restoration of function are the primary aims in ENV treatment.
- Therapeutic modalities like the complete decompresive phyITHERAPY, topical corticosteroids with occlusive dressing, psoriatane, ointme and weight reduction have proven beneficial.
- Tobacco cessation is imperative as it has been linked to autoimmune manifestations of Grave’s disease.

FIGURES

1. Figures 1-3: Clinical images of indurated 1-2cm thick violaceous plaques with interspersed pink translucent nodules; associated deep fissuring and non pitting edema. Figure 3: Active serous drainage with overlying yellow-white crust on pretibial area.

2. Figures 4-5: H&E of a 6mm punch biopsy on the right shin and right dorsal foot. Hyperkeratosis, papillomatosis, and acanthosis of the epidermis. Large quantities of mucin are deposited within the reticular dermis, causing collagen bundles to separate and the dermis to thicken. A grenz zone of normal collagen is also observed. Figure 6: Colloidal iron stain demonstrating an abundance of mucin in the throughout the dermis.

REFERENCES