

# Elephantiasis Nostras Verrucosa : A Rare Thyroid Dermopathy in Graves' Disease

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## Abstract

Elephantiasis Nostras Verrucosa (ENV) is a rare form of pretibial myxedema, which is nearly always associated with Graves' disease. A case is presented here of Graves' disease who had elephantiasis variety of pre-tibial myxedema (PTM). ©

## INTRODUCTION

Elephantiasis Nostras Verrucosa, a form of pre-tibial myxedema or more appropriately thyroid dermopathy is a term used to describe localised lesions of the skin resulting from the deposition of hyaluronic acid and chondroitin sulfate usually as a component of thyroid disease. PTM is nearly always associated with Graves' disease.<sup>1</sup> The mechanism that causes myxedema is unclear, although animal model studies suggest that thyroid hormones affect the synthesis and catabolism of mucopolysaccharides and collagen by dermal fibroblasts. The fibroblasts in the orbital and pre-tibial dermis share antigenic sites that underlie the autoimmune process that causes Graves' disease.<sup>2</sup>

ENV, which is a persistent swelling of one or both lower legs, may also occur due to recurrent erysipelas, cellulitis, as sequelae to chronic venous or lymphatic stasis of diverse origin. The skin over the toes, dorsum of feet, ankles, less often the lower legs becomes greatly thickened. The verrucous lesions are made up of crowded wart like growths which may occur in irregular plaques of different size and shape or may involve a rather large area diffusely. The solitary papillomas often look like fleshy, flattened, smooth growths attached by a slender or broad pedicle but may assume any form, varying in size from 0.5-2cm. There may be associated dermatophytosis and various bacterial species within the lesions.<sup>3</sup>

## CASE REPORT

A 29 years male diagnosed as Graves' disease in 1998 on carbimazole and propranolol presented with firm, multiple nodules, plaques and cerebriform hypertrophy

of both lower legs and feet. He initially (in 1998) noticed stiffness and tightness in the pre-tibial region of both legs which increased slowly and involved the dorsa of feet bilaterally. He also noticed protrusion of eyeballs which occurred prior to the leg swelling. He had all other classical features of Graves' disease like palpitations, weakness, weight loss, increased frequency of defaecation, increased appetite, with h/o occasional fever.

General examination revealed Grade II clubbing, tachycardia, tremor, mild pallor, BP of 130/86 mmHg. Local examination revealed – non-pitting edema, flesh coloured to erythematous, firm, confluent, polypoid nodules and fissured plaques extending from the shins to the dorsa of both feet. Examination of eyes revealed – proptosis, Von Graefe's, Joffroy's & Moebius's sign positive. Thyroid gland was enlarged with no nodularity. The systemic examination findings were within normal limits. The Laboratory investigations revealed – Hb-10gm/dl, TC-11,200/cu mm, DLC-N<sub>74</sub>L<sub>15</sub>M<sub>8</sub>E<sub>3</sub>, ESR-2 mm, platelet count – normal. The Thyroid profile was - T<sub>3</sub> – 3 ng/ml (.7-2.2 ng/ml), T<sub>4</sub> – 150 ng/ml (55-135 ng/ml), TSH-0.2 mIU/ml (0.5-4.25 mIU/ml). The Ultrasonography of thyroid gland showed diffusely enlarged gland with no nodules. The Thyroid peroxidase and thyroglobulin antibody was absent. The Liver function test were within normal limit. The CT scan orbital region increased retro-orbital fat and muscle bulk. The ASO titre, CRP and Mantoux test were Negative. The Wedge biopsy of skin thinned out stratified squamous epithelium with areas of ulceration and increased mucin deposition. Subepithelium showed loose and edematous fibro-collagenous fatty tissue. The Filarial antigen test was Negative. Thus a diagnosis of elephantiasis nostras verrucosa variant of pre-tibial myxedema with ophthalmopathy with thyroid acropachy with Graves' disease was made.

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Fig. 1 : Showing eye signs and thyroid enlargement

## DISCUSSION

The above presented case was of Graves' disease which presented as elephantiasic pretibial myxedema in the presence of ophthalmopathy and thyroid acropachy. The association of ENV with Graves' disease though reported in literature (International Journal of Dermatology Vol 4. 2001) is rare.

PTM or more appropriately thyroid dermopathy, present in 4.3% of patients with Graves' disease and in 15% of patients with Graves' ophthalmopathy is an uncommon late manifestation of Graves' disease and is always preceded by significant ophthalmopathy, in addition to thyroid gland enlargement and thyroid acropachy.<sup>4</sup> In a previously reported series of 150 patients with PTM, 58% had nonpitting edema (brawny), 20% nodular variant, 21% plaque type and less than 1% either polypoid or elephantiasic morphologies. There were no distinguishing histologic features for the various morphologic types.<sup>5</sup> Features of the elephantiasic form resemble lymphedema. Myxedema is not always confined to the pre-tibial area, however can involve the hands, arms, shoulders, ankles, ears, face and sites of trauma or scars.

The pathogenesis of localised myxedema is unclear and treatment remains problematic. Systemic or



Fig. 2 : Showing elephantiasis nostras verrucosa (Pre-tibial myxedema) with clubbing (thyroid acropachy)

intralesional glucocorticoids, topical glucocorticoids under occlusion i.e. complete decongestive physiotherapy or high dose IV Ig have been reported to offer relief.<sup>6</sup>

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