

Elephantiasic thyroid dermopathy

A 48-year-old driver presented in 2003 with progressive bilateral lower limb swelling. He was a chronic smoker with a history of Graves' disease and mild Graves' ophthalmopathy. Radioiodine had been administered in 2000 for control of hyperthyroidism. It was nonetheless complicated by hypothyroidism, exacerbation of the Graves' ophthalmopathy, and novel development of finger clubbing and myxoedematous infiltration of the skin 4 to 6 months later. Thyroxine replacement restored him to a euthyroid state. Graves' ophthalmopathy stabilised after a course of systemic steroids and external irradiation to both eyes, with resolution of diplopia despite persistent periorbital swelling and exophthalmos. His skin condition continued to deteriorate.

Examination revealed bilateral exophthalmos, clubbing with drumstick swelling of finger and toe extremities; firm flesh-coloured nodules and plaques over his back (Fig 1a), buttocks, thighs, and upper arms; firm nodular swelling of his bacille Calmette-Guérin (BCG) vaccine scar (Fig 1b); and, most striking of all, significant enlargement of both lower limbs, with malodorous firm verrucous hyperkeratotic nodules surrounded by deep fissures and folds that affected both feet and spread up his shins (Fig 2). Skin biopsy of one of the nodules over his back revealed increased dermal mucin, compatible with the diagnosis of myxoedema. Urinary glycosaminoglycan/creatinine ratio was 5.7 $\mu\text{mol}/\text{mmol}$ (reference level, <3.6 $\mu\text{mol}/\text{mmol}$). Anti-thyroid-stimulating hormone receptor level was higher than 40.0 IU/L (reference level, <2.0 IU/L) and a thyroid scan (performed after stopping thyroxine replacement for 3 weeks and elevation of thyroid-stimulating hormone level to 188 mIU/L) showed no residual thyroid uptake.

He was prescribed decongestive physiotherapy, topical steroids, and diuretics with minimal response. Intralesional steroid injection led to softening of the nodule at his BCG scar, but could not be administered to other sites because they were so widespread. Oral prednisolone up to 60 mg daily also resulted in softening of the skin lesions and improved mobility at the metatarsophalangeal and ankle joints, sufficient to allow him to resume work. Nonetheless response to treatment was incomplete; he developed Cushing's syndrome and his condition relapsed when prednisolone was tapered to 15 mg daily. Therapeutic trials with subcutaneous injection of somatostatin and its long-acting analogue (Sandostatin LAR; Novartis Pharma AG, Basle, Switzerland) and intravenous immunoglobulin both resulted in only partial and transient improvement in lesion size that again worsened once treatment was stopped. Short trials of thalidomide, azathioprine, and colchicine were ineffective. His condition continued to persist for 5 years. He was disfigured, and his mobility was compromised by impaired ankle dorsiflexion that led to problems



Fig 1. Patient with (a) firm flesh-coloured nodules and plaques on his back and (b) firm nodular swelling of his bacille Calmette-Guérin vaccine scar

with foot hygiene and difficulty in finding socks and shoes that would fit. He also developed friction-induced local ulcerations and cellulitis. He ultimately lost his job.

Localised myxoedema, or thyroid dermopathy, is an infrequent manifestation of autoimmune thyroiditis, in particular Graves' disease. It complicates 0.5% to 4.3% of cases of Graves' disease¹ and is usually associated with



Fig 2. Patient with enlarged lower limbs showing malodorous firm verrucous hyperkeratotic nodules surrounded by deep fissures and folds

Graves' ophthalmopathy and thyroid acropachy.² There is no evidence of an association between radioiodine therapy and thyroid dermopathy, or radioiodine-induced hypothyroidism and thyroid dermopathy. Nonetheless it appears plausible in view of the association between Graves' ophthalmopathy and radioiodine therapy,³ and the similarities between the pathogenetic mechanisms underlying Graves' ophthalmopathy and thyroid dermopathy.⁴

Commonly called pretibial myxoedema because of its predilection for the pretibial area, thyroid dermopathy can also develop at sites of previous injury and scars (Fig 1b), or affect other areas including the abdomen, upper back (Fig 1a), face, pinnae, hands, neck, shoulder, forearm, and scalp.^{5,6} Lesions are classically raised, waxy, flesh-coloured or yellowish brown, at times accompanied by hyperpigmentation, hyperkeratosis, and hyperhidrosis. More extensive lesions can become indurated, with a peau d'orange appearance. Thyroid dermopathy is classified as one of four forms: non-pitting oedema accompanied by

typical skin colour changes, plaque, nodular (Fig 1) or elephantiasic (Fig 2).⁶ The elephantiasic variant, as seen in our patient, occurs in only 2.8% of patients with thyroid dermopathy,⁷ and is characterised by massive oedema, skin fibrosis, and verrucous nodule formation. It is often refractory to treatment, and can cause significant morbidity, as exemplified by this case. In the Mayo series, only 27% of patients whose thyroid dermopathy was severe enough to require treatment achieved complete remission after 17 years of follow-up, and only 23% achieved partial remission.⁷

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