Complex decongestive physiotherapy for pretibial myxoedema

Lymphoedema is a chronic, debilitating disorder resulting from congenital abnormality of the lymphatic system (primary lymphoedema) or acquired causes (secondary lymphoedema). Complex decongestive physiotherapy is the 'mainstay' intervention for lymphatic insufficiency comprising manual lymph drainage, in selected cases intermittent pneumatic compression, multilayered compression bandaging with appropriate padding, exercise, and meticulous skin care (Consensus Document of the International Society for Lymphology 2003). Manual lymph drainage encourages fluid mobilisation, protein reuptake from interstitial space, and softening of fibrotic tissue. Pneumatic compression aids manual drainage. Short-stretch bandages have low resting and high working pressure, which means that contracting muscles will be compressed and aid lymph flow. Exercise while using multilayered compression also aids manual lymph drainage. Once active therapy is completed, patients continue maintenance therapy with custom-fitted stockings. As symptoms become more severe, the active phase has to be repeated.

A 40-year-old-woman with history of Graves' disease for more than 21 years presented with a progressive enlargement of both legs that significantly impaired her mobility. At the time of presentation for Graves' disease, she had excessive exophthalmos. Two years after the onset of thyroid disease she underwent partial strumectomy and 6 years ago received two sessions of radioactive iodine with a total dose of 763 MBq and intravenous corticosteroid therapy in repeated cycles. Subsequently, she gained around 18 kilograms in weight. In 1992, a biopsy of the right leg showed epidermal hyperplasia with largely increased dermal mucin.

Figure 1. The dorsal aspect of the legs a) before and b) after complex decongestive physiotherapy.

On admission, the patient demonstrated significant, non-pitting, swelling in both legs accompanied by verrucous hyperplasia and multiple, firm nodules giving the skin an accentuated, 'peau d'orange' appearance (Figure 1a). Pretibial myxoedema is a known manifestation of Graves' disease that occurs in association with diffuse thyroid gland enlargement, exophthalmos, and thyroid acropachy (Kriss 1987). Mild forms of pretibial myxoedema may regress spontaneously, but the rare, severe elephantiasic variant is often resistant to different therapeutic approaches including local and systemic corticosteroids, compression, plasmapheresis, and immunosuppressive agents (Felton et al 2003, Srebrnik et al 1992, Schwartz et al 2002). Structural and functional abnormalities of the lower limb lymphatic system have been found in pretibial myxoedema (Bull et al 1993). Therefore pretibial myxoedema is secondary lymphoedema and this warranted the application of decongestive physiotherapy for this patient. Decongestive physiotherapy comprising 45 min of manual lymph drainage plus another 45 min of intermittent pneumatic compression (30 Hgmm pressure) using Lympha Press Plus^{TM} and multilayered compression bandaging was performed once daily for 5 days in an in-patient setting by a specialised physiotherapist (Szolnoky et al 2008). Two 5-day courses were included over 8 weeks. Volumetry was performed with Kuhnke's disc model (Földi and Kubik 2000) to determine any reduction in swelling. Knee flexion range of motion, mobility and patient's perception of swelling were also measured (Szolnoky et al 2007).

After 8 weeks, the patient had lost over 3.5 kg; her right leg volume was reduced by 9% and the left by 11%. The most significant change in appearance occurred around the heels (Figure 1a and b). There was less nodularity and a smoother surface to the legs. Knee flexion increased from 90 deg to 105 deg. Walking speed over 10 m increased from 0.47 m/s (SD 0.03) to 0.60 m/s (SD 0.02). This was accompanied by an increase in step length from 31 cm (SD 1) to 36 cm (SD 1), a decrease in step width from 10 cm (SD 1) to 8 cm (SD 1), and an increase in cadence from 72 to 73 steps/min. Her perception was that the swelling had decreased from 8.3 to 4.5 on a 10-point visual analogue scale. She now regularly takes part in a belly dancing course.

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