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Successful treatment of microstomia with UVA1 phototherapy in systemic sclerosis

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None declared.

To the Editor,

The introduction of phototherapy has substantially changed the therapeutic response of localized scleroderma (LS). Systemic agents have been proposed for the treatment of LS, some with potential side effects and varying degrees of success. There is sufficient evidence in the literature to demonstrate that low dose (20 J/cm^2) and medium dose (50 J/cm^2) UVA1 phototherapy is beneficial in LS. The development of a metal halide lamp emitting high levels of UVA1 radiation (340–400 nm) was first described in 1981 (1), but the therapeutic potential for UVA1 phototherapy did not emerge until 1992 with the first successful report of treating patients for acute exacerbations of atopic dermatitis. It has now been shown to soften thickened plaques, increase skin elasticity and reduce lesional skin thickness in scleroderma (2–4). Recently (4), low-dose UVA1, medium-dose UVA1 and narrowband UVB phototherapy were compared demonstrating comparable efficacy of narrowband UVB and low-dose UVA1 but medium-dose UVA1 being more effective. In systemic sclerosis (SSc), studies of UVA1 are limited. Improvement has been documented in acrosclerosis in those patients with SSc (5). There is no documentation of treatment of other features of SSc by phototherapy in the literature. We describe a case of successful treatment of microstomia with UVA1 phototherapy in a patient with SSc.

Case report

A 45-year-old woman with SCL70+ve systemic sclerosis presented with symptomatic sclerodactyly and microstomia, which had been progressively worsening over the last 5 years. Previous treatments included hydroxychlorquine and she had declined systemic corticosteroids and other systemic immunosuppressant therapy. There was no relevant past medical history or family history and she worked as a solicitor.

On examination, there was evidence of significantly tight, thick skin affecting the hands, face, arms and upper chest and microstomia. She found it difficult to open her mouth and articulate her words and there was a slight pinching of her nose. SCL 70 antibodies were present in peripheral blood and other blood tests including inflammatory markers, FBC, urea/creatinine, LFTs and complement were normal. She was commenced on 50 J/cm^2 UVA1 phototherapy to the whole skin surface 2–3 times weekly. After 10 treatments all the sclerotic areas of skin including the perioral area were less indurated on examination. After 21 treatments she could articulate words normally and there was reduced furrowing around the mouth. After 40 treatments and a total of 2225 J/cm^2 she could open her mouth more normally: the anterior–posterior diameter from the lower margin of the front teeth to the upper margin of the lower teeth had increased by 1 cm (25%) from 4 to 5 cm (Figs 1 and 2).

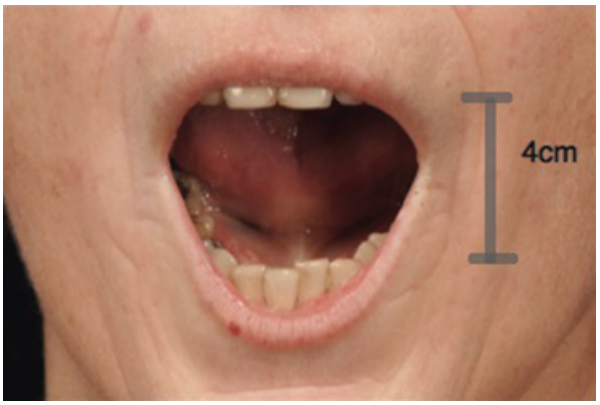

Fig. 1. March 2009.

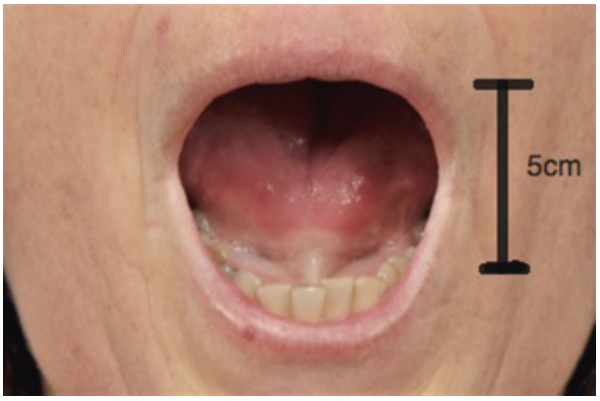

Fig. 2. April 2010.

Currently she says that brushing her teeth, eating and talking have all become significantly easier following the treatments.

Discussion

Equipment to deliver UVA1 (340–400 nm) waveband has been available from 1981 but it is only in the last 20 years that increasing beneficial use of UVA1 has been documented (4). UVA1 is accepted effective treatment for morphoea(2–4), systemic scleroderma (3) and granulomatous chronic GVHD (3). It is also used and may be effective in urticaria pigmentosa (3, 4), scleredema adultorum Bushke (3, 4), granuloma annulare (4), nodular prurigo (4), mycosis fungoides (3), atopic eczema (4), polymorphic light eruption (4) and SLE (3).

Systemic sclerosis is difficult to treat. Despite advances in disease-specific treatment of other rheumatologic diseases, disease-targeted treatment in systemic sclerosis continues to be elusive suggesting treatment involves a complex interaction of specific targets. There are no published studies of treatment of microstomia in systemic sclerosis. UVA1 phototherapy exerts its therapeutic effects through modulation of three predominant pathogenic mechanisms in sclerosis: immune dysregulation, imbalance of collagen deposition and endothelial dysfunction (5). In our case, UVA1 appears to have been effective in softening sclerotic perioral skin and improving symptoms caused by the microstomia. This case highlights how UVA1 phototherapy should be considered early if patients with systemic sclerosis and significant disability with skin involvement such as microstomia are unable to tolerate systemic therapy, but also importantly as an early adjunct to systemic therapy.

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