UVA1 for diffuse cutaneous systemic sclerosis in a Fitzpatrick skin type VI patient: outcomes in the modified Rodnan skin score

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To the Editor,

Cutaneous sclerosis is the main clinical marker of diffuse systemic sclerosis, being responsible for important limitations in daily life activities[1]. The modified Rodnan skin score is the most widely used method to evaluate the skin thickening, through clinical palpation of 17 anatomic regions[2].

Morita *et al.* showed that Ultraviolet A1 (UVA1) phototherapy depletes skin-infiltrating T cells with T-cell apoptosis. Besides, UVA1 can up-regulate the expression of collagenase-1 in dermal fibroblasts[3]. In the last decade, UVA1 has been used to treat cutaneous sclerosis, achieving long periods of remission and clinical improvement. However, most reports have focused on Caucasian population[4].

The authors present a 44-year-old Cape Verdean patient, Fitzpatrick skin type VI, diagnosed with diffuse cutaneous systemic sclerosis. He had extracutaneous involvement, presenting esophageal dysmotility and pulmonary fibrosis, which had been stable throughout the 8 years of follow-up. Besides the characteristic facies with microstomia, retraction of the lips, perioral furrows, and a beaked nose, he had significant active fingers flexion impairment, particularly of the right hand (Figure 1A). Moreover, he also had a restriction of the abduction of the arms. He had performed physiotherapy, but with no significant benefit. The patient was being treated with hydroxychloroquine, domperidone, pantoprazole and pentoxifylline during the last years.

In 2015, he started UVA1 phototherapy daily from Monday until Friday (Waldmann® 7001 UVA cabin equipped with 40 Philips TL/10R lamps – spectral irradiation between 340 and 400 nm). We performed a whole body treatment and the initial dose was 10 J/cm^2, rapidly increased up to a steady dose of 35 J/cm^2, maintained until a previously defined number of 40 sessions was achieved.

Before starting UVA1, his modified Rodnan skin score was 26. A clinically significant improvement of the skin thickening was observed after 15 sessions. At the end of 40 sessions (cumulative dose of 2575 J/cm^2), the improvement was generalized with a greater skin elasticity and strong improvement in the active mobility (Figure 1B). There was significant improvement in the score especially of the hands, arms, forearms, anterior chest and abdomen. The final modified Rodnan skin score reached 11 points. Six months after, the patient reported a worsening of the mobility of the right hand and arm, although it was not as severe as previously. He performed the same treatment this time and again one year after, reaching similar results in the modified Rodnan skin score after the three treatments. Apart from xerosis, which easily improved with a daily emollient, no other side effects were reported.

The severity of cutaneous sclerosis is predictive of disease progression and the modified Rodnan skin score may have correlation with the clinical course and prognosis of systemic sclerosis[5-7]. A higher score may reflect the persistence of cutaneous sclerosis and it may be associated with higher morbidity and mortality[5-7]. We reinforce these findings taking into account both the improvement of cutaneous sclerosis and the stable systemic disease throughout the follow-up period. Furthermore, this clinical case highlights the role of the modified Rodnan skin score as a practical and useful tool during the follow-up of patients with systemic sclerosis. Besides, it strengthens the clinical benefit of UVA1 phototherapy to improve cutaneous sclerosis, and the related mobility impairment[3] in patients with diffuse cutaneous systemic sclerosis and higher Fitzpatrick skin types[4], having a good safety profile[8]. According to

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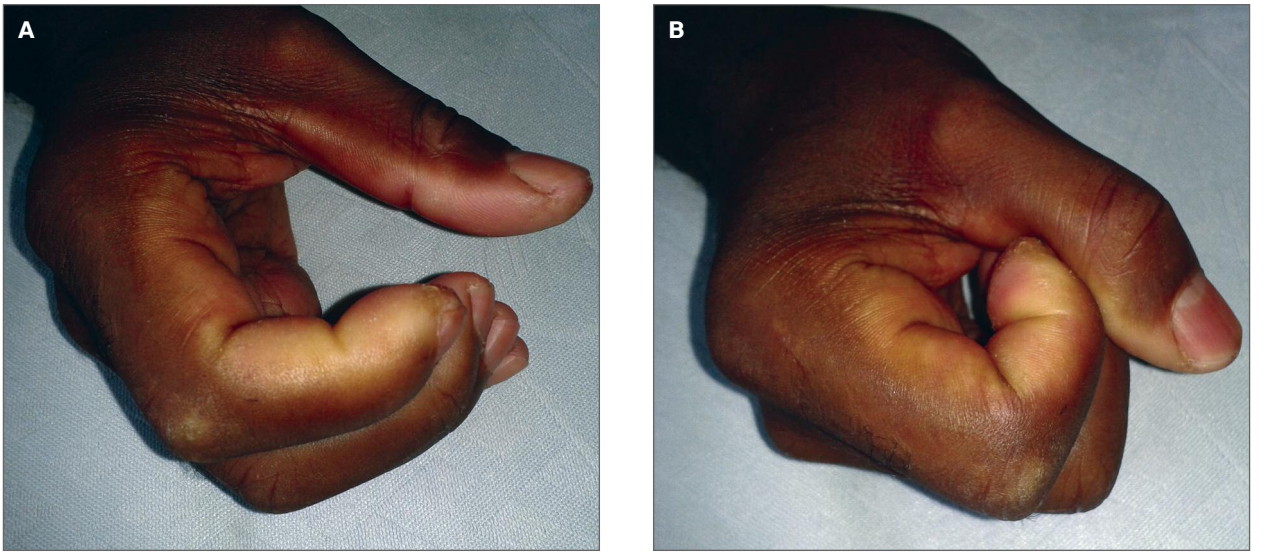

FIGURE 1A/1B. **A.** Impairment of active fingers flexion of the right hand before UVA1. **B.** Improved mobility after UVA1

the few studies published, a dose of 20-50 J/cm^2 is effective in systemic sclerosis, even in higher phototypes, as we could confirm[9]. Finally, considering the results achieved, we would suggest that a dose of 35 J/cm^2 may provide a clinically significant improvement with a lower cumulative dose.

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