Unilateral generalised morphea successfully treated with rituximab and mycophenolate mofetil

Sirs,

Localised scleroderma (LS) encompasses a heterogeneous group of dermatoses characterised by epidermal atrophy and dermal thickening. Rarely, LS can extend up to the deep fascia and the underlying muscle.

A 46-year-old Asiatic man presented to us due to a 7-year history of a skin rash affecting the left side of his body.

Concomitant disease includes a necrotising myositis, diagnosed about 10 years earlier, and for which he was on treatment with prednisone 1mg/kg/day and intravenous immunoglobulins at a 3-week interval. The disease led to a progressive weakness of the proximal muscles of the upper and lower limbs, that becomes gradually more severe on the left side, leading to an obvious claudication. Physical examination revealed multiple indurated, atrophic, red to brown plaques, that involved the left side of his trunk and back and the proximal upper and lower left limbs and distributed along the lines of Blaschko. No lesions were present elsewhere (Fig. 1). Laboratory tests showed elevation of serum creatine phosphokinase and aldolase; anti-U1 Ribonucleoprotein (anti-U1 RNP) IgG antibody (Abs) titre (6.9 UI/mL; normal range <0.2 UI/mL) was also elevated; anti-nuclear Abs, anti-double-stranded DNA Abs, anti-extractable nuclear antigen Abs and myositis-specific Abs were within normal range. Electromyography revealed signs of persistent activity of the associated myositis, especially on the left side. A lesional skin biopsy showed epidermal atrophy, hyperpigmentation of the basal layer, and thickened and closely packed collagen bundles in the deep dermis; atrophy of adnexa and a mild lymphocytic perivascular infiltrate were also present (Fig. 2). Both clinical and histopathological findings were consistent with a diagnosis of morphea; considering the unilateral distribution of the lesions, a diagnosis of unilateral-generalised morphea showing an atypical distribution along the lines of Blaschko was made.

Mycophenolate mofetil (MMF) at a dose of 360 mg twice daily and a cycle of rituximab (500 mg once a week for 4 weeks) were added to his treatment schedule. Rituximab 500mg was subsequently applied at a 6-month interval for 2 more years. After the 12-month follow-up, the patient achieved a good clinical response, with softening of the skin lesions; muscle enzymes were also within the normal range.

The term generalised morphea refers to patients with four or more lesions larger than 3 cm in diameter, affecting 2 or more of seven anatomical sites (head-neck, each extremity, the anterior and the posterior trunk) (1, 2).

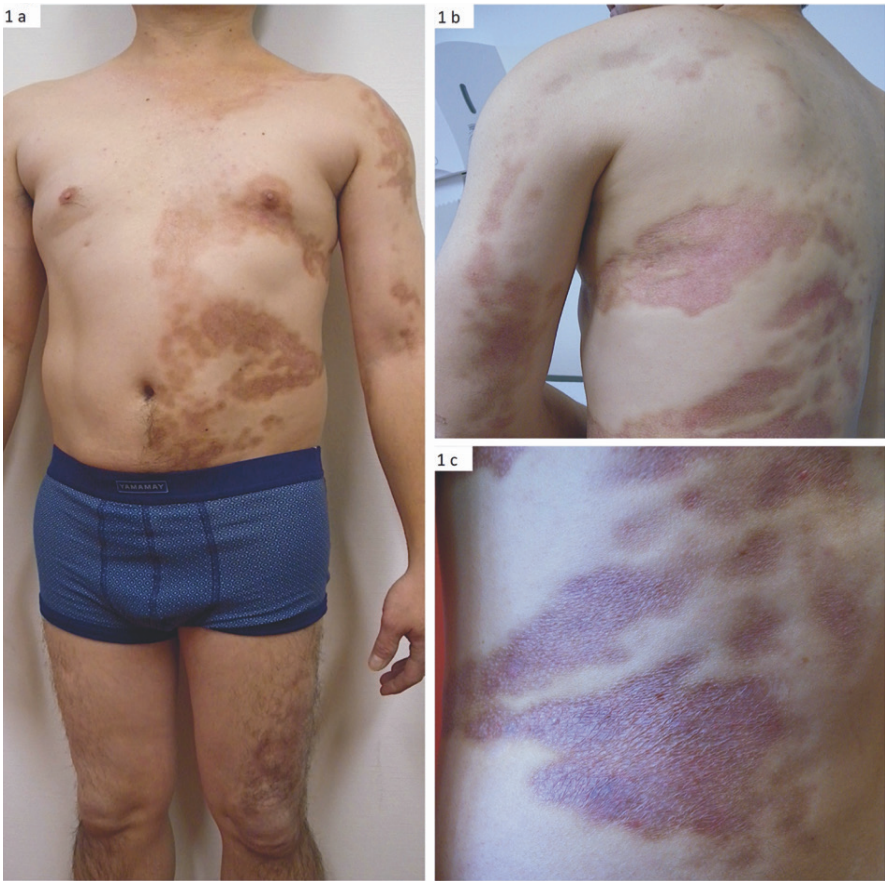

Fig. 1. (**a**-**b**) Unilateral depressed plaques with atrophic skin distributed over the left side of the body following the Blaschko lines. (**c**) Detail of the skin lesions at the patient's back.

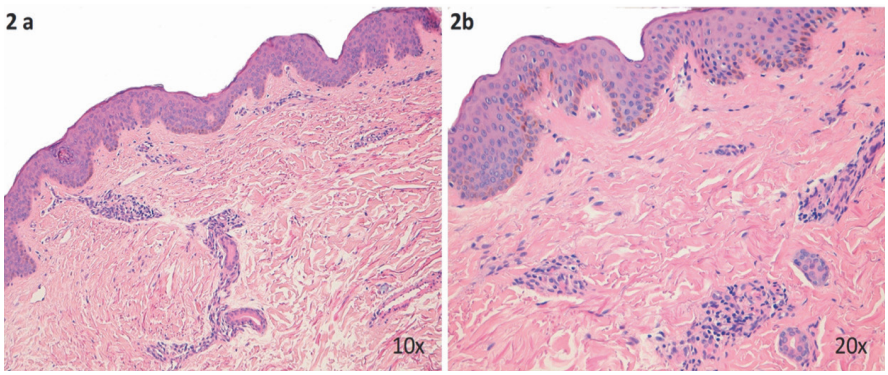

Fig. 2. (**a**) Histopathology showed epidermal atrophy, increased dermal thickness with sclerotic collagen bundles and atrophy of the adnexa (haematoxylin and eosin, magnification 10x);
(**b**) Thickening of the walls of small blood vessels, hyperpigmentation of the basal layer and a mild lymphocytic perivascular infiltrate (haematoxylin and eosin, magnification 20x).

When inflammation and sclerosis are limited to one hemisoma, the term Unilateral-Generalised Morphea (UGM) has been proposed (3). Here, we showed a case of UGM with skin lesions characteristically distributed along the lines of Blaschko. Differential diagnosis in our case included linear idiopathic atrophoderma of Pasini and Pierini (APP) and linear atrophoderma of Moulin (LAM). APP is characterised by atrophic hyperpigmented patches. Unlike morphea, induration is characteristically absent (4). LAM is characterised by unilateral, hyperpigmented and atrophic patches that follows the Blaschko lines. Unlike morphea, both the epidermis and dermis have a normal thickness; inflammation and sclerosis are absent (5).

Another interesting finding of this case is that the myositis was shown to be more evident by electromyography examination at the left hemisoma, presumably related to the presence of the skin lesions in these anatomical sites. Our patient significantly improved upon treatment with MMF and rituximab. MMF selectively blocks the

inosine monophosphate dehydrogenase, an enzyme that is pivotal for lymphocyte proliferation (6). Rituximab works by inducing B-cell depletion (7). Our case strengthens previous observations regarding the successful use of these drugs in patients with generalised morphea resistant to conventional treatments (8).

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Competing interests: none declared.

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