The pathophysiology of post-HSCT relapse is unclear, but the temporal relationship of the relapse in our patient with immune reconstitution suggests a relationship with re-emergence of autoreactive clones (Fig. 1). Studies regarding correlations between immunological parameters and relapse after HSCT for dcSSc are conflicting [7]. At baseline, our patient had a positive ANA test (1:100), anti-RNP III antibodies and anti-SSA antibodies. Anti-SSA antibodies disappeared after the first HSCT and remained absent during the relapse, although the ANA test remained weakly positive (granular staining pattern) and RNP III antibodies persist up until now.

In conclusion, our case underscores the potential benefit of a second HSCT with post-HSCT immunosuppression in SSc patients who relapse after HSCT. However, caution should be used regarding possible toxicity and long-term side-effects and a careful screening procedure remains essential, as described by us recently [8]. Ultimately, the decision to perform a second HSCT requires good multidisciplinary support as well as shared decision making with the patient.

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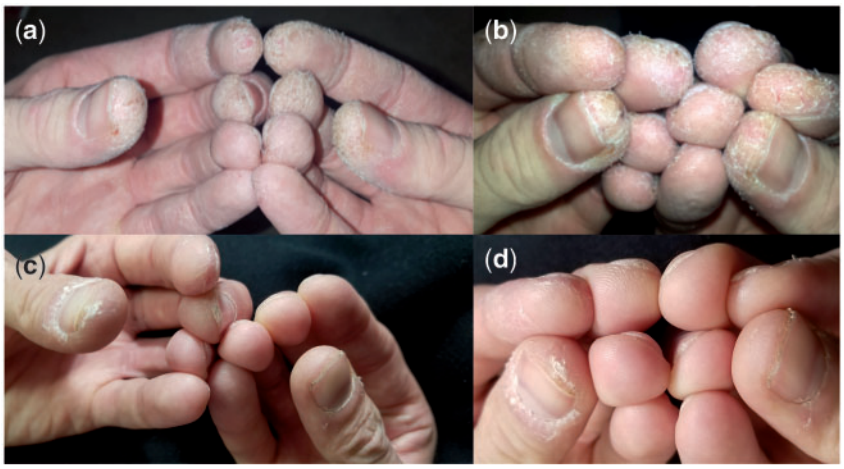
Successful treatment of refractory mechanic's hands with ustekinumab in a patient with the antisynthetase syndrome

Rheumatology key message

- Ustekinumab may be useful to treat mechanic's hands in patients with the antisynthetase syndrome.

Sir, A 29-year-old non-smoker male presented with a history of progressively worsening arthralgia, arthritis and proximal weakness. He also had hyperkeratotic scaly lesions on his fingers and hands characteristic of mechanic's hands, with no other skin manifestations. Creatine kinase levels were increased and a muscle biopsy revealed an inflammatory myopathy. The patient had no signs of lung involvement and pulmonary function tests were normal. Anti-histidyl-tRNA synthetase (i.e. anti-Jo1) autoantibodies were positive and the patient was diagnosed with the antisynthetase syndrome. Following treatment with mycophenolate and prednisone, the patient's muscle and joint involvement improved and his muscle enzyme levels normalized. However, over the next 3 years, the skin lesions continued to worsen despite treatment with multiple drugs, including corticosteroid ointments, topical tacrolimus, adalimumab and methotrexate (Fig. 1a and b). Given the refractory nature of the mechanic's hands, treatment with subcutaneous ustekinumab was initiated as recommended (45 mg initially and 4 weeks later, followed by 45 mg/12 weeks) with a dramatic improvement of the skin lesions over the following

Fig. 1 Mechanic's hands



Mechanic's hands before (**a**, **b**) and after (**c**, **d**) starting treatment with ustekinumab.

3 months (Fig. 1c and d). At the time of starting ustekinumab, the patient continued on treatment with mycophenolate and prednisone, both of which drugs were started 2 years earlier. The other manifestations of the disease remained under control after starting ustekinumab.

The antisynthetase syndrome is characterized by autoantibodies targeting one of the aminoacyl tRNA synthetases along with one or more of the following: myositis, interstitial lung disease, arthritis, Raynaud's syndrome, fever or mechanic's hands [1]. The presence of mechanic's hands is one of the most characteristic clinical features of this type of myositis, and histological analysis shows hyperkeratosis, parakeratosis and psoriasiform acanthosis with dermal and epidermic inflammation. Unlike in psoriasis [2], there is no suprapapillary thinning [3]. Little is known about the pathogenesis of mechanic's hands and, unfortunately, this cutaneous manifestation of the antisynthetase syndrome may be more refractory to treatment than other manifestations of the disease.

Ustekinumab is a human mAb directed against the p40 subunit of IL-12 and IL-23 that has been Food and Drug Administration-approved for treatment of psoriasis [4] and Crohn's disease [5]. Here, we demonstrate the successful use of this drug to treat refractory mechanic's hands in a patient with the antisynthetase syndrome. Along with the pathologic similarities between mechanic's hands and psoriatic lesions, the response of both to ustekinumab may suggest that similar inflammatory pathways are involved in each process. Additional studies are necessary to assess the efficacy and safety of using ustekinumab to treat mechanic's hands, and perhaps other disease manifestations, in patients with the antisynthetase syndrome.

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