Successful therapy of refractory dermatomyositis skin rash with anifrolumab

Sirs,

Dermatomyositis (DM) is a chronic inflammatory disorder of striated muscles and skin. We present a case of highly resistant DM that responded dramatically to anifrolumab.

A 34-year-old Caucasian lady presented to us in 2016 with a heliotrope (dark pink) skin rash, Gottron (purple) papules, proximal muscle weakness and dysphagia. Creatine kinase (CK) was 445 U/L (normal <171), anti-TIF1gamma was positive, while skin biopsy showed interfacies dermatitis. She was treated with prednisone 50 mg daily tapering followed by rituximab (1 gx2) and intravenous immunoglobulins (0.4 g/kg/day for 5 days) with normalisation of muscle strength and dysphagia, but her skin rash remained very active. Over time, her skin rash proved resistant to a mean prednisone dose of 25 mg/day and (sequentially) ciclosporin 3 mg/kg/day, methotrexate 20 mg/week, mycophenolate mofetil 2 g/day, and tofacitinib 5 mg twice daily. Attempts to reduce the prednisone dose failed because of worsening of skin lesions with an intolerable burning sensation and itching.

We therefore chose to treat her with anifrolumab, an inhibitor of type I interferon receptor subunit-1 approved for the treatment of SLE, because DM rash is akin to lupus dermatitis both histologically and because of the pathogenic role of type I interferon in both disorders. Prior to anifrolumab, skin rash was severe with an active CDASI (Cutaneous Dermatomyositis Disease Area and Severity Index) of 34 (Fig. 1). Concomitant medications included prednisone 25 mg/day, cholecalciferol and alendronate. Anifrolumab was administered at the standard dose of 300 mg e.v. One month later, active CDASI decreased to 13, her symptoms (burning and pruritus) significantly improved, and prednisone was cautiously tapered to 17.5 mg/day. At her subsequent visit, two months after starting anifrolumab active CDASI further decreased to 4 (Fig. 1), and prednisone was tapered to 12.5 mg/day (Fig. 1). No safety signals emerged during therapy with anifrolumab.

Anifrolumab has shown considerable efficacy in both cutaneous lupus and in the rash of DM even in refractory patients. In our patient, active CDASI before therapy with anifrolumab was 34 (a score >19 indicates severe skin rash) (1). Response to anifrolumab was dramatic, with the CDASI decreasing to 13 after a single dose and 4 after two doses (a reduction in 4-5 points is considered clinically significant) (2), while prednisone could be tapered to 12.5 mg/day.

A review of the literature revealed fourteen cases of DM-resistant treated with anifrolumab, all for skin manifestations (2-9) and one for active muscle disease as well (2). Of these, three had juvenile and eleven adult-onset DM, while two patients had paraneoplastic DM (5;9), although in one patient neoplasm had been eradicated prior to therapy with anifrolumab (5). CDASI reduction (when reported) was ~20 points over 1-8 months (2;2;3;5;6;8). Muscle disease responded to anifrolumab in one patient with active myositis (in addition to skin rash) (2). No treatment-related adverse events have been reported.

Of the patients described, two had failed tofacitinib (4;7), one responded well but relapsed while continuing on tofacitinib (2), while another relapsed after withdrawal of tofacitinib (due to a diagnosis of neoplasm) (5). The response to anifrolumab in tofacitinib failures is all the more remarkable since tofacitinib is considered quite effective for treating DM rash even when other medications have failed (10).

Of the four cases for which myositis-specific autoantibodies were reported, three were anti-TIF1gamma- (3;5;6) and one was anti-NXP-positive (4), consistent with the association of anti-TIF1gamma with more severe skin disease (as in our case).

In conclusion, anifrolumab appears to be highly effective for refractory DM rash with an excellent safety profile. Anifrolumab should be considered in DM-resistant skin rash.

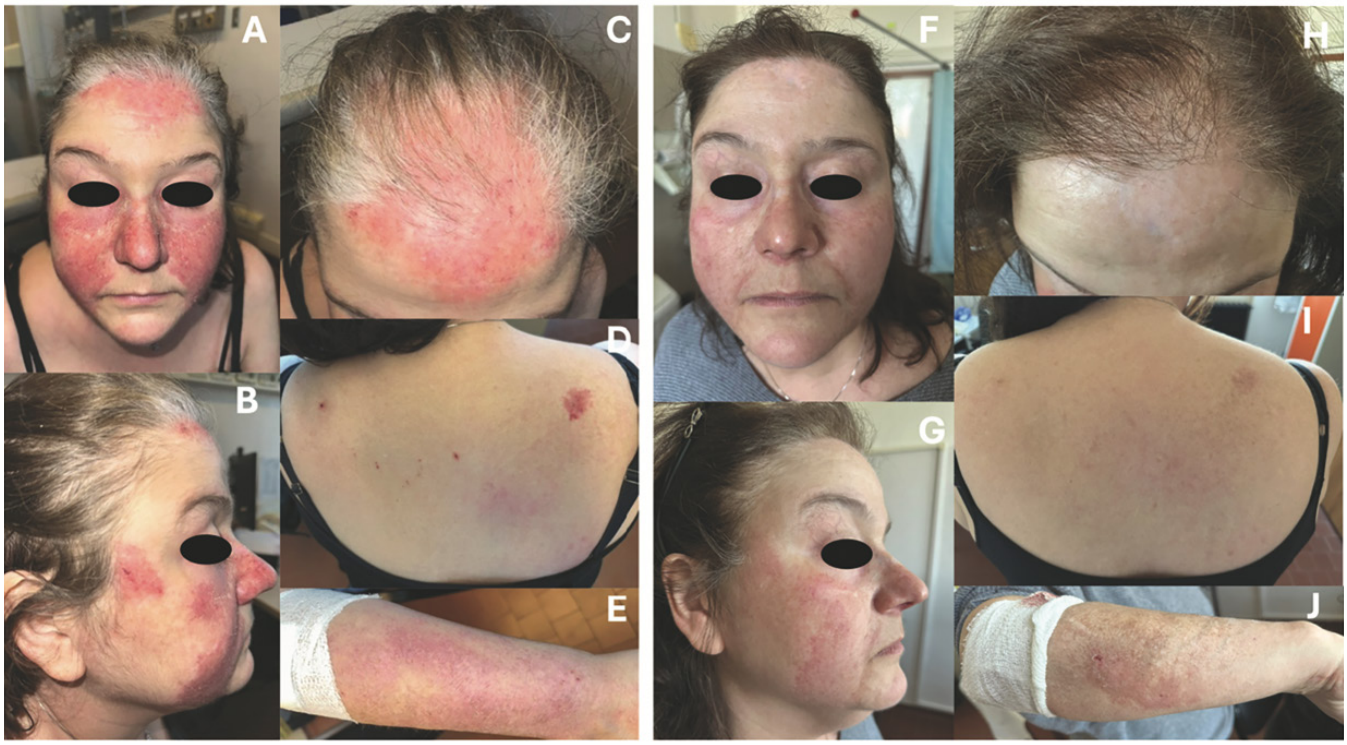

Fig. 1. Skin rash of the patient before (left panel, A-E) and after (right panel (F-J) 2 infusions of anifrolumab (A-F: face, frontal view; B-G: face, lateral view; C-H: scalp; D-I: upper back; and E-J-: right arm).

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