

Unexpected photosensitivity – a patient with dermatomyositis and concomitant solar urticaria

Editor

Dermatomyositis (DM) is a connective tissue disease with rare incidence, characterized by progressive muscle weakness and typical cutaneous manifestations. Its pathogenesis is still incompletely understood, but next to genetic predisposition environmental factors seem to be implicated in the development of DM.¹ Photosensitivity is one important factor for the onset and exacerbation of DM, which is corroborated by the distribution of the cutaneous features on sun-exposed areas and disease photoaggravation.^{2,3}

Here, we report about a 64-year-old male patient with recently diagnosed DM and specific photosensitivity. The patient suffered from symmetrical proximal weakness and showed typical skin features [heliotrope rash (Fig. 1a), Gottron papules, nailfold changes]. With antinuclear antibody titres of 1 : 12 800, specific anti-Mi-2-antibodies⁴ and elevated muscle enzymes of 585 U/L, the diagnosis was confirmed promptly.

Further, the patient complained about itching and burning skin lesions manifesting immediately after sun exposure. To investigate UV radiation as a possible trigger of disease flare, we performed photoprovocation tests. At the non-irradiated buttocks area, increasing doses of broadband UVA- and UVB-rich radiation were applied.

Surprisingly minutes after provocation, the patient developed erythema and wheals in the UV-exposed areas but no DM-specific skin lesions (Fig. 1b). Hence, we diagnosed a hitherto unknown solar urticaria. The minimal urticarial dose (MUD) was <5 J/cm² with UVA irradiation and <25 mJ/cm² with UVB irradiation (Table 1).

For the treatment of DM, an immunomodulatory medication with systemic corticosteroids (prednisolone: starting dose 120 mg/day, tapering to 10 mg/day) and azathioprine (100 mg/

day) was initiated. Interestingly, regarding solar urticaria, no therapeutic effect or major change in MUD-UVA could be seen (Table 1). Because of relapsing DM symptoms, the treatment needed to be escalated. The patient received a therapy with mycophenolate mofetil (MMF; 1000 mg/day) and high-dose intravenous immunoglobulins (IVIg; 2 g/kg bodyweight every 6 weeks). Initially, only a small increase in the MUD-UVA was observed still not increasing >5 J/cm². Though after 1 year with MMF (2000 mg/day) and continuous IVIg cycles, we observed a significant decrease in photosensitivity (MUD-UVA: 20 J/cm²; MUD-UVB: 75 mJ/cm²; Table 1).

Both, DM and solar urticaria, are rare conditions. To our knowledge, this is the first description of a case showing an association between both diseases. Photosensitivity is known for a subgroup of DM patients. Dourmishev *et al.* reported on 36.8% of his DM patients complaining about increased cutaneous photosensitivity. He demonstrated a reduced MED to UVB among DM and LE patients compared to healthy controls.² Besides, a significant association between UV radiation and induction of anti-Mi-2 antibodies among American women was described by Love *et al.*³ suggesting that UV radiation modulates the immune system and could promote autoimmunity.

Surprisingly in our case, the patient's photosensitivity was primarily not associated with DM, but was due to solar urticaria. Solar urticaria is an idiopathic, probably IgE mediated photodermatosis, that is defined by the development of classical urticarial lesions minutes after sun exposure with short duration.⁵ The therapeutic management of solar urticaria includes sun protection as well as phototherapy/hardening,⁶ oral antihistamines, omalizumab^{7,8} and immunosuppressive agents. The use of IVIg in therapy-refractory patients is described to be successful in reaching partial remission.^{9,10} Aubin *et al.*¹⁰ performed a multicentric study in patients with refractory solar urticaria and showed clinical improvement in 67% of tested patients.

In our patient, we initially observed only a small change in the MUD under a regimen including IVIg, but the long-term



Figure 1 Clinical pictures. (a) Patient (skin type III) with dermatomyositis and typical heliotrope rash and shawl sign. (b) Phototesting with increasing doses of UVA- (upper row) and UVB-rich irradiation (lower row) manifesting solar urticaria in December 2016.

Table 1 Results of follow-up photoprovocation tests

Time	Therapy	MUD-UVA (tested doses)	MUD-UVB (tested doses)	MED-UVB
Dec 2016	No therapy	<5 J/cm ² (5, 10, 15, 20, 25, 30 J/cm ²)	<25 mJ/cm ² (25, 50, 75, 100, 125, 150 mJ/cm ²)	50 mJ/cm ²
Feb/May 2017	Prednisolone; azathioprine (100 mg/day) since 2/5 months	<5 J/cm ² (Feb 2017) (5, 10, 15, 20, 25, 30 J/cm ²) 1 J/cm ² (May 2017) (0.5, 1, 2, 3, 4, 5, J/cm ²)	50 mJ/cm ² (25, 50, 75, 100, 125, 150, mJ/cm ²)	75 mJ/cm ²
Jun 2017	IVIG (75 g/6 weeks); mycophenolate mofetil (1000 mg/day)	2 J/cm ² (0.5, 1, 2, 3, 4, 5 J/cm ²)	n.d.†	n.d.†
Jan 2018	IVIG (75 g/6 weeks); mycophenolate mofetil (2000 mg/day) since 5 months	3 J/cm ² (0.5, 1, 2, 3, 4, 5 J/cm ²)	50 mJ/cm ² (15, 25, 50, 75, 100, 150 mJ/cm ²)	75 mJ/cm ²

†Due to the persistent pigmentation of the preceding UVB irradiations, no further UVB irradiation was performed.

Determination of the minimal urticarial dose (MUD) during follow-up photoprovocation tests (UVA 700 Waldmann, emission spectrum 325–490 nm, maximum at 365 nm; UVB Philips TL20W/12RS fluorescent test lamps, main emission spectrum between 275 and 365 nm, maximum at 313 nm) to assess the potential suppressive effect of the different immunomodulatory therapies for DM on solar urticaria.

follow-up (12 months) showed a significant increase in the MUD and therefore a remarkable suppressive effect of IVIG on the solar urticaria. In patients with severe solar urticaria, this treatment option should be considered.

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DOI: 10.1111/jdv.15379

Prevalence and persistence of oral HPV infection in Italy

Editor

Oral human papillomavirus (HPV) persistent infection may cause numerous tumours^{1,2} but little is known about the HPV clearance time and the variables that may influence the HPV persistence.³

We aimed to assess the prevalence of the oral HPV infection, the HPV persistence rate and the variables influencing the persistence.

We retrospectively studied 274 patients all without overt signs of oral HPV infection, visited from December 2015 to December 2017 at the Dermatology Clinic in Genoa: 241 had a sexually transmitted infection (STI) and 33 participated in a screening programme among the healthy population. For 125 of the patients included in this study, molecular/clinical data have been previously described.⁴ No patient had comorbidities, except two HIV-positive patients. Samples from the oral cavity (dorsal/ventral tongue, buccal mucosa) were collected with ThinPrep liquid-based cytology preparation system and underwent polymerase chain reaction to detect HPV types 6, 11, 16, 18, 31, 33, 35, 39, 40, 42, 43, 44, 45, 51, 52, 53, 58. All patients gave consent to the study. Statistical analysis was performed (Fisher's test) considering *P*-value < 0.05 as statistically significant.

Overall, 90/274 patients (33%; 43 women, 47 men) proved positive for oral HPV [37 low-risk (LR)-HPV, 53 high-risk