Reticulated atrophic skin lesions in an adult patient with dermatomyositis and antinuclear matrix protein-2 autoantibodies

Sir,

Dermatomyositis is an autoimmune connective tissue disorder classically manifesting with cutaneous inflammation and myopathy. Recently, multiple novel myositis-specific autoantibodies have been described which have been linked to different clinical phenotypes of the disease. Antinuclear matrix protein-2 autoantibodies, in particular, are associated with mild skin involvement, calcinosis, peripheral oedema, myalgia and dysphagia, as well as paraneoplastic dermatomyositis in adults.[1]

A 32-year-old man presented to our dermatology department with erythematous rash on the face [Figure 1a]; erythematous papules and crusted erosions on the chest [Figure 1b] and reticulated atrophic plaques on the right shoulder [Figure 1c] and both arms. The reticulated plaques were atrophic and whitish, situated on an erythematous base and were covered with few yellowish crusts. Dermoscopy demonstrated telangiectasias, skin atrophy and crusts [Figure 2]. The patient lacked the typical cutaneous features of dermatomyositis (heliotrope erythema, Gottron's sign/papules). He had noticed skin lesions for two months and also reported severe muscle weakness, myalgia and dysphagia to solids.

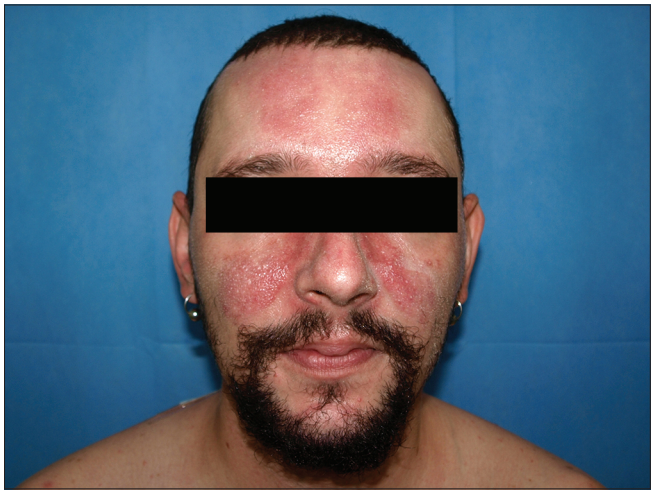

Figure 1a: Violaceous erythema on the face

Complete blood count was normal but serum biochemistry revealed elevated alanine aminotransferase (95 IU/L), aspartate aminotransferase (172 IU/L), gamma-glutamyl transfease (75 IU/L) and creatine kinase (3769 IU/L). Electromyography revealed myogenic injury of the proximal

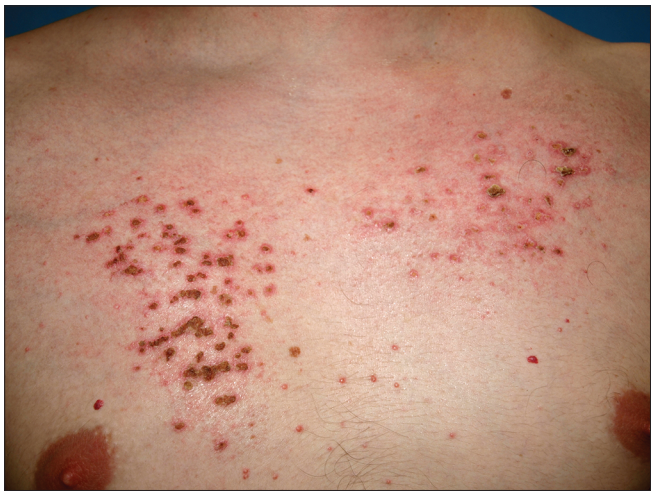

Figure 1b: Crusted erosions on the chest

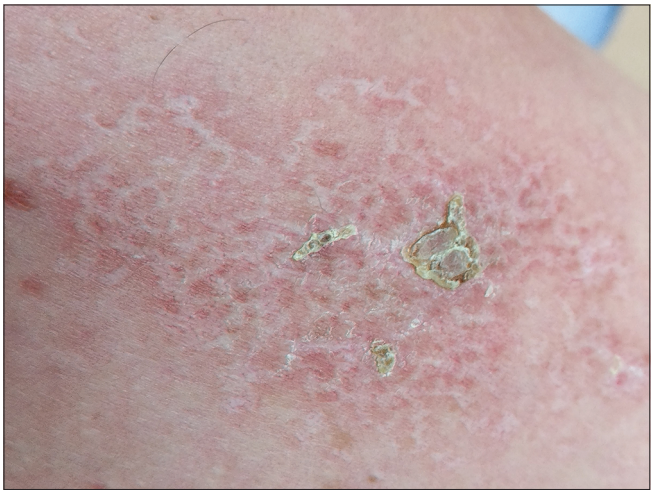

Figure 1c: Reticulated atrophic plaque on the right shoulder

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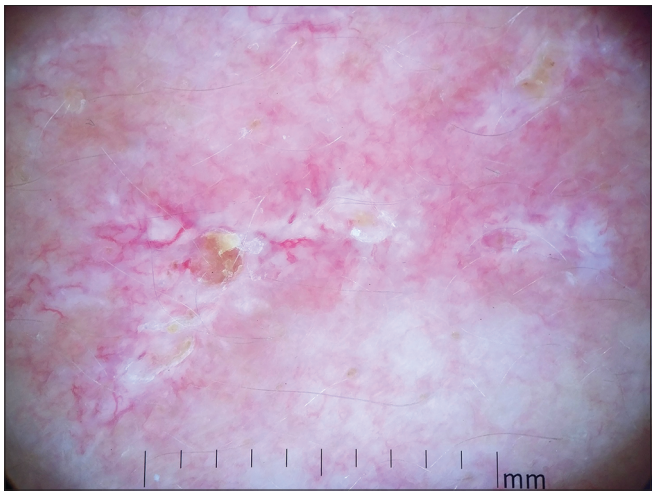

Figure 2: Dermoscopy of crusted and atrophic skin lesion (×10, DermLite DL1, OnePlus 3T Camera, polarisation mode)

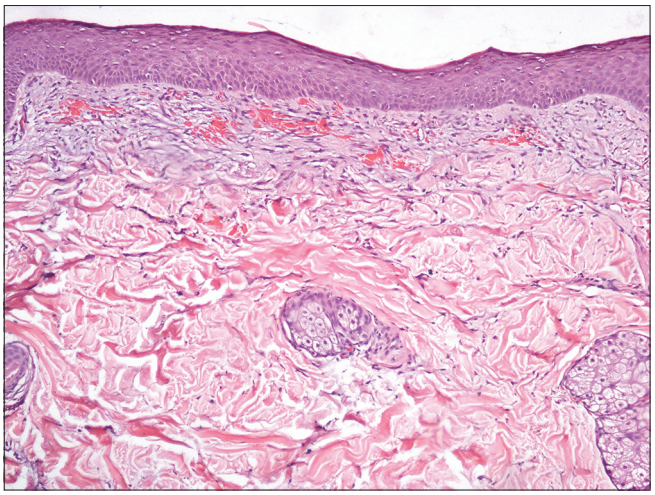

Figure 3: Histology from the right shoulder lesion shows atrophic epidermis, moderate fibrosis in the upper dermis with widening of some of the collagen bundles and erythrocyte extravasation in papillary dermis (H&E, ×100)

muscles of the upper and lower extremities compatible with inflammatory myopathy. Muscle biopsy was not performed.

Skin biopsy from the face demonstrated atrophic epidermis, follicular hyperkeratosis, sparse perivascular and periadnexal inflammatory infiltrate with perifollicular leukocytoclasia. Histology from chest erosion showed a lack of epidermis, fibrin crust with reactive hyperplasia in the surrounding epidermis and mild fibrosis in the upper dermis. Skin biopsy from the reticulated atrophic skin lesion on the right shoulder showed atrophic epidermis, moderate fibrosis in the upper dermis with widening of some of the collagen bundles and erythrocyte extravasation in papillary dermis [Figure 3]. A complete immunologic workup was done, including antinuclear antibodies, myositis specific [anti-Mi-2 antibody (Mi-2α), anti-Mi-2β antibody (Mi-2β), transcriptional intermediary factor 1γ (TIF1-γ), melanoma differentiation-associated protein 5 (MDA5), nuclear matrix protein 2 (NXP2), signal recognition particle (SRP), SUMO-activating enzyme subunit 1 (SAE1), EJ, OJ, PL-7, PL-12 and Jo-1] and associated autoantibodies (Ku, PM/Scl-75, PM/Scl-100 and anti-Ro52), as well as antiphospholipid, anti-La, anti-ribonucleoprotein (RNP), anti-Sm, anti-double-stranded-DNA autoantibodies and C3 and C4 factors of complement. It revealed high titres of antinuclear antibodies – 1:320 (positive >1:160; indirect immunofluorescence on Hep-2 cells) and antinuclear matrix protein-2 autoantibodies – strong positive, +++ (immunoblot assay). All the other autoantibodies, C3 and C4 were within reference ranges. A diagnosis of dermatomyositis was confirmed.

Paraneoplastic screening, comprising abdominal sonography, chest X-ray, tumour markers (PSA – prostate-specific antigen, CA19-9 – carbohydrate antigen 19-9 and AFP – α-fetoprotein) and serum immunofixation, excluded the presence of malignancy.

Echocardiography revealed no abnormality. Phototesting demonstrated an increased photosensitivity toward short wave ultraviolet B. Serology for Lyme disease was negative.

Treatment with methylprednisolone 40 mg/daily and methotrexate 15 mg weekly resulted in improvement of muscle symptoms. A moderate potency corticosteroid cream was applied on skin lesions other than reticulated atrophic plaques which led to subsidence of skin rash except for the facial livid erythema and the reticulated atrophic lesions.

The history of dysphagia and myalgia was typical for antinuclear matrix protein-2 phenotype.[1] However, some unusual skin lesions were also present. Erosions are generally considered a marker for paraneoplastic dermatomyositis but no evidence of malignancy was found in our patient. We plan to keep the patient under follow up as malignancy may be diagnosed even 3-5 years after diagnosis of dermatomyositis. The localised reticulated atrophic plaques were the most striking finding and several conditions were discussed in differential diagnosis as follows. Poikiloderma in dermatomyositis affects the sun-exposed skin but it is usually a late finding.[2] Poikiloderma atrophicans vasculare is characterised by a limited number of patches with a reticulate pattern and is considered a variant of mycosis fungoides.[2] Histology of skin lesions in our patient ruled out the possibility of mycosis fungoides. Cutaneous leucocytoclastic vasculitis, when reported in dermatomyositis, affects paraneoplastic cases and the lesions lack a reticulated pattern.[3] Livedoid vasculopathy resolves with characteristic atrophie blanche like scarring. In association with dermatomyositis, it usually affects elderly individuals with malignancies.[4] Morphea and atrophoderma of Pasini and Pierini do not exhibit reticulated pattern and the clinical presentation differs. In addition, serology for Lyme disease was negative in our patient. A few cases of dermatomyositis with Degos-like skin changes have

been described and only one with positive antinuclear matrix protein-2 autoantibodies.[5]

We were unable to find any previous reports that described similar reticulated atrophic skin lesions in dermatomyositis. Further follow-up would be needed to elaborate the correct diagnosis for the reticulated atrophic plaques in our patient. The question whether these specific lesions might be related to the antinuclear matrix protein-2 phenotype remains open.

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Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest
There are no conflicts of interest.

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Lichenoid pseudovesicular papular eruption on nose through the dermatoscope

Sir,
Lichenoid pseudovesicular papular eruption on nose (LIPEN) is a newly described entity in dermatology characterized by relatively asymptomatic grouped vesicle-like/pseudovesicular papules on the nose and adjoining facial areas and occasionally on the extensors of arms and dorsum of hands. These lesions histopathologically show a dense focal and/or nodular lichenoid infiltrate in the upper dermis.[1] We report four cases with newly described dermatoscopic features.

All four patients presented to the dermatology outpatient department of All India Institute of Medical Sciences Bhopal and Kubba Skin Clinic, New Delhi with asymptomatic erythematous papules on the nose and upper lip. The age of the patients (one man, three women) ranged between 21 and 43 years (mean = 32.3 years) . There was no history of photosensitivity, episodic flare of erythema, excessive sweating over the papules or any other history suggestive of connective tissue disease or rosacea. Cutaneous examination showed pinhead-sized erythematous to skin-coloured shiny papules on the tip and alae of the nose and upper lip below the nasolabial fold [Figure 1]. In two of our cases, the forehead and malar areas were involved with confluent plaques and other two cases had extra-facial involvement with tiny papules on the dorsum of the hand, upper back and forearm [Table 1].

A dermatoscopic evaluation (Heine Delta 20 dermatoscope,® 10×, polarised) of all the four cases showed round to oval, variably sized, yellowish-orange to red clustered clods with

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