Dermatology Practical & Conceptual

Cutaneous Manifestations and Their Corresponding Dermoscopic Features in Patients with Dermatomyositis

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ABSTRACT

Introduction: Dermoscopy is a noninvasive and easy to apply technique that allows in vivo magnification of the skin and thus observation of morphologic structures invisible to the naked eye. Recently, it gained popularity for evaluation of inflammatory skin conditions. In the field of connective tissue diseases, dermoscopy has been used mainly as a simple and accessible substitute of nailfold capillaroscopy.

Objectives: The aim of the present study is to expand the application of dermoscopy in patients with dermatomyositis (DM) beyond the usual nailfold examination. A clinico-dermoscopic correlation between clinical signs of skin affection and dermoscopic features is also suggested.

Methods: A total of 29 patients with DM were enrolled in this descriptive prospective study, conducted over a 3-year period. Dermoscopy was performed by a DermLite DL1 dermatoscope on polarization mode, attached to One Plus 3T camera. The following skin lesions were examined: periungual affection, scalp DM, Gottron papules, palmar papules, poikiloderma and auricular changes.

Results: Dermoscopy detected predominantly advanced nail fold capillary changes - giant capillaries (79%), microhemorrhages (46%) and avascular areas (25%). The most prevalent trichoscopic features were enlarged tortuous capillaries (64%), interfollicular scales (50%) and peripilar casts and tufting (36%). Among the other skin lesions assessed in this study - Gottron papules were present in 20 patients, poikiloderma in 11, palmar papules in 4 and auricular lesions in 4 patients.

Conclusions: The use of dermoscopy for clinical evaluation of skin lesions in DM enhances diagnostic accuracy and elucidates poorly known characteristics of the disease.

Introduction

Dermoscopy (epiluminescence microscopy) is a noninvasive and easy to apply technique that allows *in vivo* magnification of the skin and observation of morphologic features invisible to the naked eye. In the last decades dermoscopy has become a key tool for the evaluation of pigmented and nonpigmented skin tumors. Recently, an increasing number of publications have appeared that demonstrate the role of the handheld dermatoscope in an entirely new field - the field of inflammatory skin lesions. A huge break-through in this direction was the expert consensus published in 2020 on behalf of the International Dermoscopy Society providing a set of standardized basic dermoscopic parameters to follow when evaluating inflammatory, infiltrative and infectious dermatoses [1].

Nailfold capillaroscopy (NFC) is currently considered the gold standard for early assessment of microvascular changes in the nailfold area of patients with rheumatic diseases. As a simpler and more accessible method than NFC, dermoscopy has also been tested for gross analysis of capillary nailfold abnormalities in several studies comprising patients with collagen-vascular disorders [2,3].

Dermatomyositis (DM) is a rare autoimmune connective tissue disease that affects the skin, the skeletal muscles and the internal organs. Although its pathogenesis is not fully understood, the immune response is thought to originate from the capillary endothelium of the endomysium. An activation of the complement pathway and deposition of C5b-9 membrane attack complexes results in depletion of capillaries, ischemia, muscle and skin injury [4]. Therefore, vascular skin changes are fundamental for DM and a variety of vascular patterns might be observed via dermoscopy.

Objectives

The aim of the present study is to expand the application of dermoscopy in patients with DM beyond the simple nailfold examination. In order to do that, we have performed dermoscopy of other skin lesions, such as scalp DM, Gottron papules, palmar papules, poikiloderma and auricular lesions in the quest for subtle diagnostic clues, invisible to the unaided eye. In addition, we suggest a clinico-dermoscopic correlation between clinical signs of skin affection and dermoscopic features.

Due to the low incidence of DM in the population, dermoscopic data in different studies is combined with data from other connective tissue diseases which compromises the unique characteristics of the disease. The present study focuses only on patients with DM.

Methods

Twenty-nine patients with classic DM (CDM, 22 patients), clinically amyopathic DM (CADM, 4 patients) and DM/overlap syndrome with another connective tissue disease (3 patients), such as systemic lupus erythematosus and systemic sclerosis were enrolled in this prospective descriptive study. The study was conducted in the Department of Dermatology and Venereology, Alexandrovska University Hospital, Sofia, Bulgaria between September 2018 and August 2021. Only patients with diagnosis of definite DM according to Bohan and Peter criteria for classic DM were included [5]. Patients with amyopathic DM were selected based on clinical skin findings, suggested by Sontheimer [6,7]. Additional examinations comprised serum enzyme levels, myositis-specific autoantibodies, electromyography, and skin biopsy. The mean age was 51 years (range: 19-77 years). Twenty patients were females (69%) and 9 patients were males (31%). A cancer-associated, or paraneoplastic DM was diagnosed in 10 patients (breast carcinoma in 2 patients, cervical carcinoma in 2, thyroid carcinoma in one, endometrial carcinoma in 2, ovarian carcinoma in one, cecal carcinoma in one, and bladder carcinoma in one). The disease duration ranged between 1 month and 23 years, and 69% of the patients had a maximal disease duration of 3 years.

All the 29 patients were included in the study after signing a written informed consent.

The dermoscopic examination was performed by a DermLite DL1 dermatoscope on polarization mode, attached to One Plus 3T camera.

Results

Affection of periungual skin was assessed by the following clinical parameters-ragged cuticles, erythema, nail fold telangiectasias visible by naked eye, erosions/ulcerations, skin atrophy. Ragged cuticles (N = 14,58%) and nail fold erythema (N = 14,58%) were relatively common in our patients. Some patients had only one of these parameters, and some of them had both. Macroscopic nail fold telangiectasias (N = 1), erosions/ulcerations (N = 2) and skin atrophy of the periungual area (N = 1) were a rare finding (supplementary Table S1; Figure 1).

Specific dermoscopic features were divided into 5 groups - subtle changes or disorganization of the normal capillary architecture, enlarged/giant capillaries, microhemorrhages (defined by > 2 microhemorrhages per digit or confluent hemorrhagic zones), loss of capillaries/large avascular areas, and ramified/bushy capillaries. Most of the patients had more pronounced nail fold capillary changes therefore only three patients presented with disorganization of the normal capillary architecture (13%). Enlarged/giant capillaries were the most common dermoscopic feature and occurred in nineteen patients (79%). Other signs like microhemorrhages, avascular areas and bushy capillaries were found in 46% (N = 11), 25% (N = 6) and 13% (N = 3) of the patients respectively (supplementary Table S1; Figure 11).

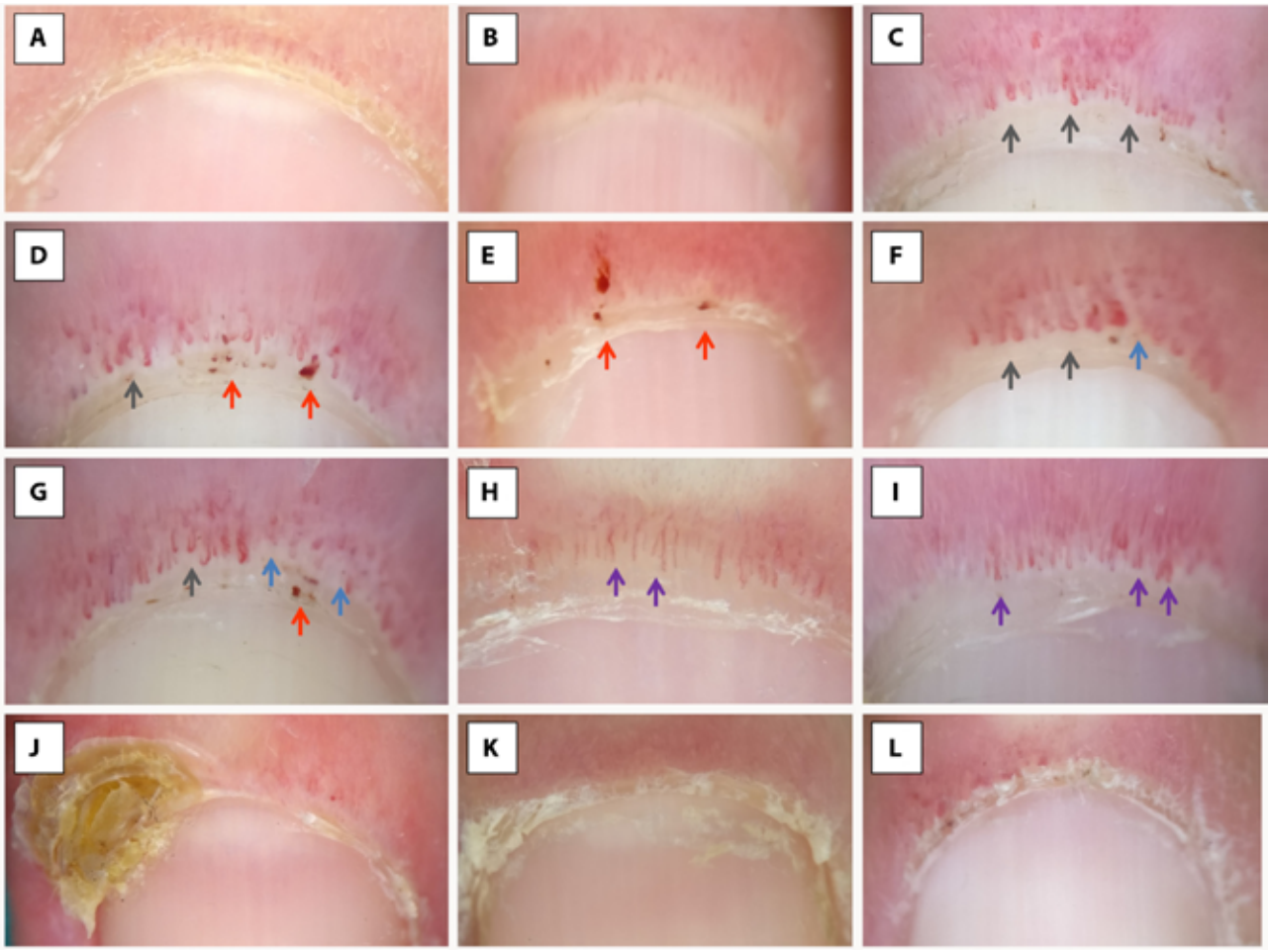

Figure 1. (A and B) Disorganization of the normal capillary architecture. (C-G) Enlarged/giant capillaries (gray arrows), microhemorrhages (red arrows), loss of capillaries/large avascular areas (blue arrows). (H and I) Ramified/bushy capillaries (purple arrows). (J) Nailfold erosion covered with crust. (K and L) Ragged cuticles.

Clinical signs of scalp affection comprised erythema (N = 12, 55%), scaling (N = 11, 50%), non-scarring alopecia (N = 14, 64%), as well as calcinosis cutis and erosions/ulcerations (none of the patients). Pruritus was reported by 16 patients (73%).

Dermoscopy of the scalp, also called trichoscopy, revealed enlarged tortuous capillaries in fourteen patients (64%), peripilar casts and tufting in eight patients (36%), interfollicular scales in eleven patients (50%), bushy capillaries in three patients (14%), interfollicular pigmentation in five patients (23%), perifollicular pigmentation in two patients (9%), vascular lake-like structures in seven patients (32%), punctuate hemorrhages in one patient (5%). Hair shaft abnormalities were also detected - broken hairs (n=2; 9%), pigtail hairs (n=1; 5%), in the absence of other conditions, that may present with such hair defects (e.g., fungal infection, alopecia areata etc.). Clinical and dermoscopic parameters concerning scalp affection are shown on supplementary Table S1 and Figure 2.

Dermoscopy of Gottron papules visualized erythema (N = 20, 100%), scaling (N = 20, 100%), skin atrophy (N = 1, 5%), irregularly arranged vessels (N = 6, 30%) and erosions/ulcerations (N = 1, 5%).

Poikiloderma, which by definition encompasses telangiectasias, dyspigmentation and atrophy, was found in eleven patients (38%) and palmar papules in four patients (14%).

Dermoscopic data about Gottron papules, poikiloderma and palmar papules is presented in supplementary Table S1 and Figure 3.

Several unusual findings which deserve attention were also noticed. Four of the patients had auricular lesions (prominent telangiectasias, venous lake-like structures, erosions). One patient presented with significant perifollicular erythema on the body and extremities (supplementary Table S1 and Figure 3).

Conclusions

Periungual changes are a well-known marker for cutaneous affection in DM. As a standard practice, periungual lesions are examined via NFC. NFC pattern in DM is generally similar to this found in systemic sclerosis patients and therefore is referred to as a “scleroderma-like pattern”. NFC represents a valuable tool for the clinician as studies confirm that changes in nail-fold capillaries reflect disease activity in

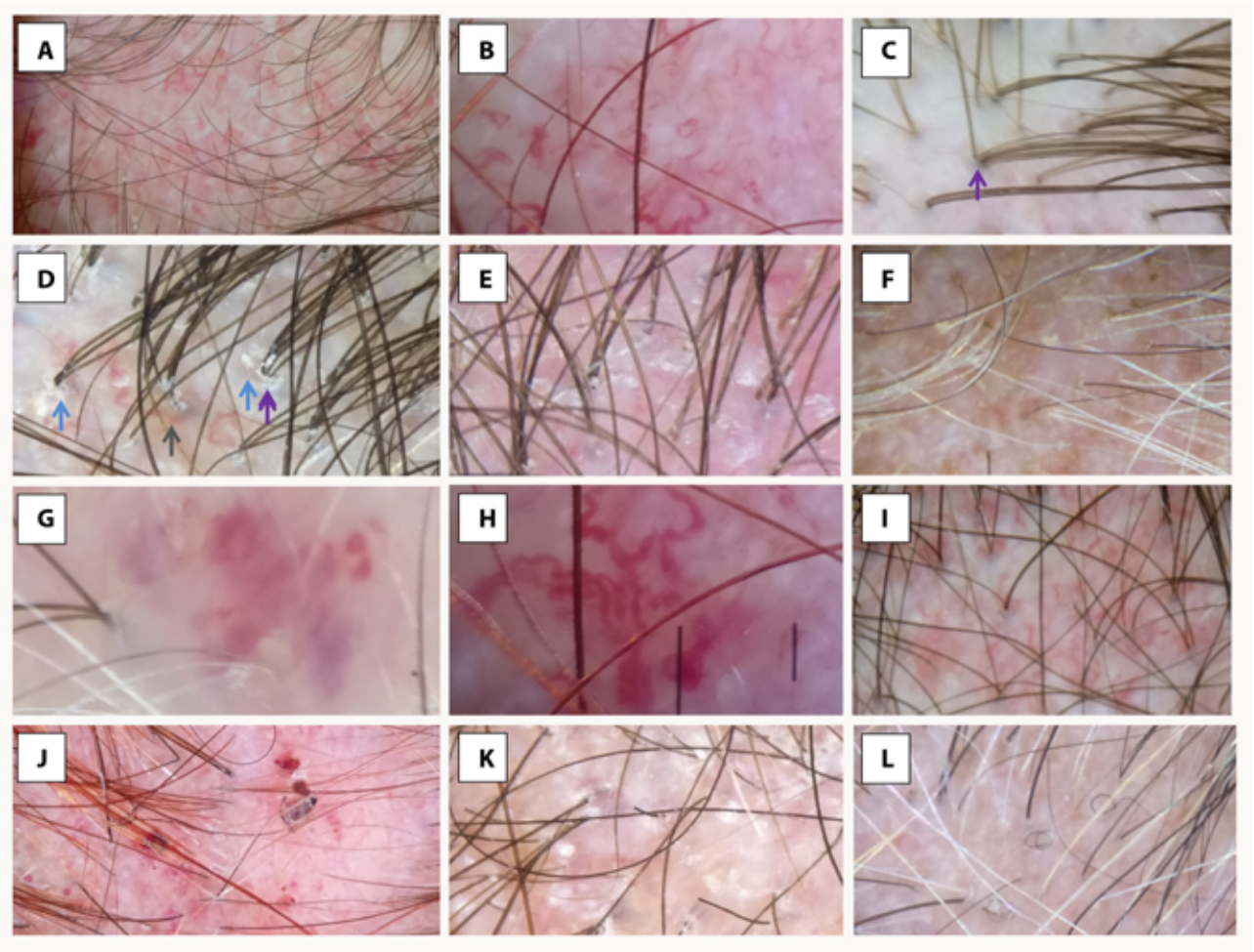

Figure 2. (A and B) enlarged tortuous capillaries. (C and D) Tufting (purple arrows), peripilar casts (blue arrows), perifollicular pigmentation (gray arrow). (E) Interfollicular scales. (F) Interfollicular pigmentation. (G) Vascular lake-like structure. (H) Bushy capillaries. (I) Perifollicular erythema. (J) Punctuate hemorrhages. (K) Broken hairs; (L) Pigtail hairs.

DM [8,9]. Data on periungual dermoscopy in DM patients is scarce and it is unclear whether dermoscopy might be used as a prognostic method for disease activity, in the same way as NFC, in those patients.

Our dermoscopic findings correspond roughly to those reported in the literature [10-12]. Most of the patients presented with scleroderma-like pattern suggestive for active disease. The most frequent nailfold dermoscopy features were respectively enlarged or giant capillaries and microhemorrhages. The enlarged/giant capillaries are regarded as an abnormal angiogenic response, secondary to peripheral ischemia. Hemorrhages most probably result from capillary injury caused by ischemia-reperfusion. Signs of early (disorganization of the normal capillary architecture) or late scleroderma-like pattern (large avascular areas/ loss of capillaries and ramified/bushy capillaries) were less frequent in our patient group.

Scalp involvement is a generally overlooked clinical characteristic of DM hence its actual frequency remains unrecognized. In the majority of manuals and review articles scalp DM is described as an erythematous, scaly and sometimes pruriginous condition of the scalp, resembling seborrheic dermatitis or psoriasis. Often nonscarring alopecia is also present [13-15]. We found only one study that examines scalp dermoscopy in DM patients [16]. In this study the authors performed trichoscopy on 31 patients with DM and concluded that in their patient group the most consistent finding was the presence of enlarged capillaries (71.4%), followed by peripilar casts (57.1 %) and tufting, and interfollicular scaling (50%). Our results demonstrated predominance of the same dermoscopic features - enlarged tortuous capillaries in fourteen patients (64%), peripilar casts and tufting in eight patients (36%), interfollicular scales in eleven patients (50%). In addition, we detected some hair shaft abnormalities (broken hairs, pigtail hairs) in the absence of other conditions that may present with such hair defects. We did not find any previous publications about hair shaft abnormalities in patients with DM.

Enlarged capillaries are a trichoscopic finding characteristic for connective tissue diseases. They can be easily differentiated from other vascular patterns, such as prominent arborizing vessels in seborrheic dermatitis or twisted capillary loops in psoriasis [17].

Hair tufting usually is regarded as a sign of scarring alopecia. The presence of more than six hairs suggests a

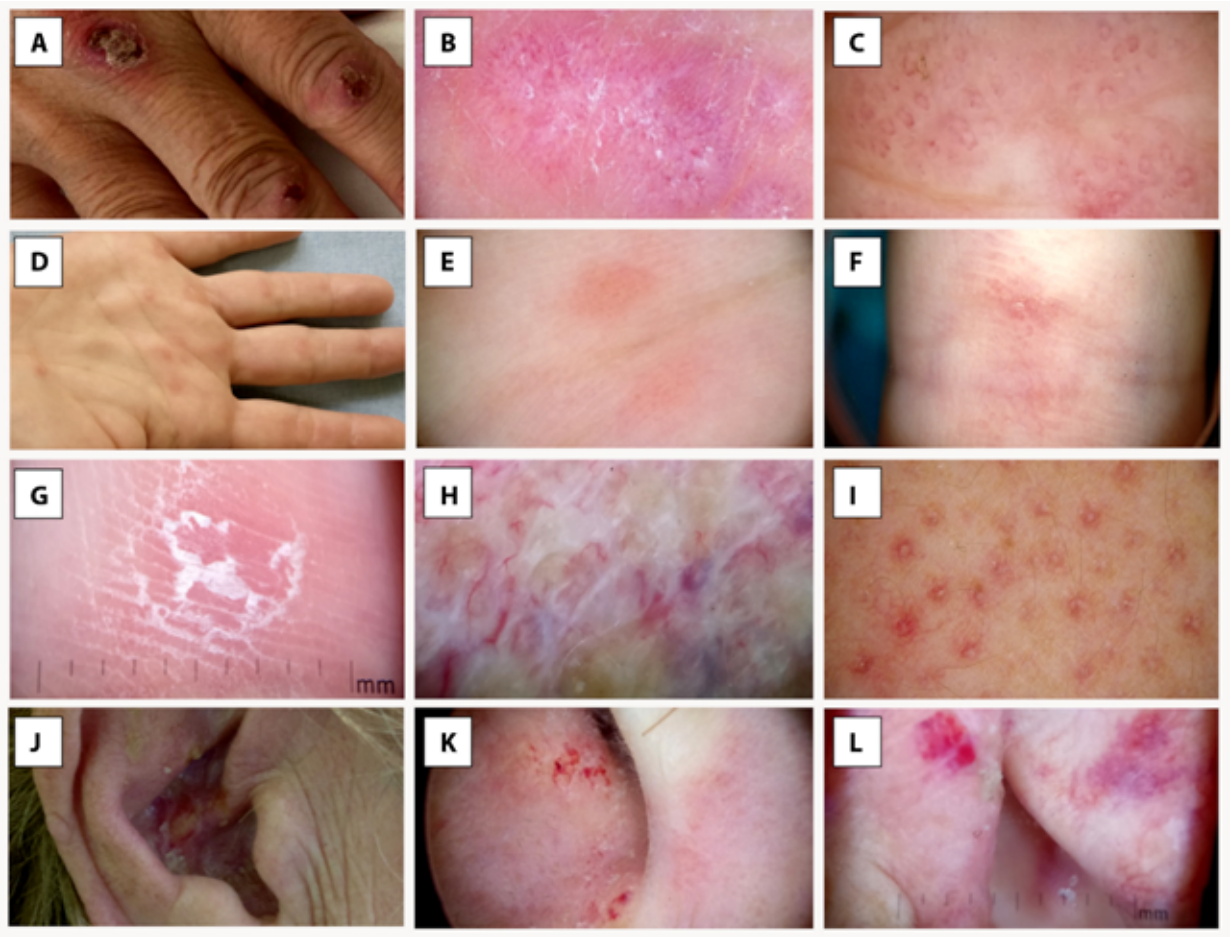

Figure 3. (A) Ulcerative Gottron papules. (B and C) Dermoscopy of Gottron papules. Note the circular appearance of the enlarged vessels in (C).Clinical (D) and dermoscopic pictures of palmar papules (E-G). Note how palmar papules are located on the two opposite sides of the joint.,(H) Poikiloderma on dermoscopy. (I) Perifollicular erythema on dermoscopy. (J) Auricular erosions. (K and L) Telangiectasias and vascular lake-like structures of the auricle on dermoscopy.

diagnosis of folliculitis decalvans. Hair tufts in lichen planopilaris contain fewer than six hairs (small tufts of four to five hairs)[17]. For the purpose of this study, we consider tufting as more than 3 hair shafts emerging together from the same follicular opening, similarly to inflammatory scalp diseases.

Tufts tend to be surrounded by peripilar casts or tube-like layers of scales enclosing the hair shafts. The latter are also considered indicative of scarring alopecia [17].

Broken hairs are relatively non-specific and are seen in a variety of scalp diseases, such as alopecia areata, trichotillomania, tinea capitis, and trichorrhexis nodosa. In alopecia areata, broken hairs may develop by two mechanisms. One is inflammation-driven transverse fracture of terminal hair shafts and the other, rapid regrowth of incompletely destroyed hair shafts [14].

Pigtail or circular hairs appear as a result from fast hair regrowth before full recovery of the hair follicle. They are considered typical for alopecia areata although sometimes found in cicatricial alopecia [14].

In the expert consensus on dermoscopic parameters in general dermatology the topic about scalp dermoscopy is not addressed. Further prospective studies are needed to elaborate a guideline for evaluation of trichoscopic features in inflammatory diseases, especially considering rare diseases such as DM [1].

Dermoscopic picture of Gottron papules and Gottron sign is nonspecific. Irregular capillaries and scales on an erythematous background could be easily visualized. However, single peculiar cases are described. Hasegawa reported the presence of punctuate hemorrhages, detected by dermoscopy, on the elbow of a patient with Gottron sign [11]. The same patient was positive for anti-melanoma differentiation-associated protein 5 (MDA-5) autoantibody and later-on developed a rapidly progressive interstitial lung disease. Hasegawa suggested that punctate hemorrhages might be related to vascular injury in the context of the aggressive interstitial pneumonia. The correlation between anti-MDA 5 antibody specificity in DM patients and the presence of skin ulcers is well known. Despite that, ulcerative Gottron papules/Gottron sign are a rather rare phenomenon [18]. In our patient group we had a case with anti-MDA 5 autoantibodies who exhibited ulcerated Gottron papules, noticeable

by naked eye, along with ulcerations all over the body and extremities (Figure 3).

Palmar papules or inverse Gottron papules are tender and often painful lesions on palmar surfaces with a predilection towards the metacarpal and interphalangeal joints. They are described also as a part of the MDA-5 antibody phenotype. Sometimes they are two separate papules on the two opposite sides of the joint [19]. Dermoscopy of palmar papules is virtually unexplored. In our patient group we had four patients with palmar papules examined by dermoscopy. We noticed the presence of dotted vessels on an erythematous base, sometimes with an orange hue. Scales were also observed (Figure 3). Palmar papules are considered a result of vasculopathy as histopathological data from previous studies demonstrates vasculopathy of small and medium-sized dermal vessels [19]. We did not perform skin biopsy of palmar papules. Interestingly, only one of our cases had MDA-5 autoantibodies in the serum. The other three were positive for transcriptional intermediary factor 1 gamma (TIF1-γ), small ubiquitin-like modifier activating enzyme (SAE1) and Mi-2 Beta autoantibodies, respectively.

Poikiloderma in DM is often a late finding. Typical distribution involves the sun-exposed skin of the neck and chest (V-sign) or the sun-protected skin of the upper back (shawl sign) and lateral thighs (holster sign). Seldom, it might have a more generalized character [20]. Dermoscopy enables better visualization of the three distinctive components, forming poikiloderma - cutaneous atrophy, telangiectasias, and macular pigmentary changes (Figure 3).

Antihelix/helix violaceous macules, erythematous auricular papules and even small ulcerations of the ears have been described previously in association with anti - MDA 5 antibody phenotype and have been proposed as a prognostic marker for fatal pulmonary disease [21-23]. The latter findings might be explained by an underlying vasculopathy. As tempting as it may seem to bind the auricular involvement with a particular autoantibody profile, we do not consider that ears, as a target organ in DM, are specifically affected only in patients with anti MDA-5 antibodies. In our opinion, auricular involvement is a known, although rarely noticed and little studied aspect of the clinical picture in DM [24]. In our study group, we have three patients with prominent auricular telangiectasias and one patient with both auricular vascular lake-like structures, and erosions (Figure 3). As the patient with auricular erosions is TIF1-γ positive, we suggest that in her case a different pathogenetic mechanism, other than vasculopathy, might be implicated in erosion formation. A possible interpretation might be a vacuolar degeneration of the basal layer of the epidermis.

Notable perifollicular erythema on the skin of the body and extremities was observed in one patient. The patient had a darker skin color (type IV according to Fitzpatrick classification (Figure 3).

The use of dermoscopy for clinical evaluation of skin lesions in DM enhances diagnostic accuracy and elucidates poorly known characteristics of the disease.

The present study had some obvious limitations due to its small sample size. Larger studies could help to distinguish clinical and dermatoscopic hallmarks in various subtypes of DM. The role of dermoscopy as a prognostic tool for disease activity and outcome in patients with DM remains to be investigated.

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