



Trichoscopic, oral, and periungual fold findings as activity and damage markers in dermatomyositis patients and their correlation with myositis antibodies

Catalina Salgueiro¹ · María José Poblete¹ · Christian Robles-Silva² · Álvaro Abarzúa¹ · Cristián Vera-Kellet^{1,3}

Received: 11 December 2022 / Revised: 11 December 2022 / Accepted: 22 January 2023 / Published online: 9 February 2023
© The Author(s), under exclusive licence to Springer-Verlag GmbH Germany, part of Springer Nature 2023

Abstract

There is little clarity about the clinical manifestations of dermatomyositis (DM) in the periungual folds, scalp, and oral cavity and their association with disease activity and damage. The objective of this study was to compare the prevalence of trichoscopic, oral, and periungual changes between DM and healthy patients and assess their possible association with disease activity and damage. We conducted an observational, transversal, and analytical study between 2020 and 2021. Forty DM patients were matched by sex and age with 40 healthy individuals. On the same day, all patients had a clinical evaluation of the hands, periungual folds, scalp, and oral cavity. Photographs of these areas and peripheral venous blood tests, including myositis-associated (MAAs) and myositis-specific antibodies (MSAs), were taken. Two dermatologists blinded to their diagnosis, damage, and activity levels registered the lesions. The disease activity and damage were evaluated using the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI). The presence of mechanic's hands, Gottron's sign, and Gottron's papules in hands; capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds; thick tortuous capillaries in scalp; gingival telangiectasias in the oral cavity; and positive MSAs associated with severe cutaneous involvement in DM patients (Anti-TIF1g, Anti-MDA5, Anti-SAE1/2) were associated with a higher CDASI activity score. The presence of MSAs associated with intense muscle involvement in DM patients (Anti-Mi2a, Anti-Mi2b, Anti-NPX2, and Anti-SAE1/2) was related to a lower CDASI activity score. Gottron's sign and Gottron's papules in hands; capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds; basal erythema in scalp; and gingival telangiectasias in the oral cavity were associated with a higher CDASI damage score. There are trichoscopic, oral and periungual fold findings and some myositis-specific antibodies that correlate with disease activity and damage in DM patients.

Keywords Dermatomyositis · Dermoscopy · Nail-fold · Oral manifestations · Autoantibodies · Disease activity

Abbreviations

DM Dermatomyositis;
CDASI Cutaneous dermatomyositis disease area and severity index
Abs Autoantibodies

ESR Erythrocyte sedimentation rate
CK Total creatine kinase
RF Rheumatoid factor
LDH Lactate dehydrogenase
ANA Antinuclear antibody

✉ Cristián Vera-Kellet
cristianverakellet@gmail.com; cvera@med.puc.cl

- ¹ Department of Dermatology, Escuela de Medicina, Pontificia Universidad Católica de Chile, Av. Vicuña Mackenna 4686. Macul, 7820436 Santiago, Chile
- ² Escuela de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile
- ³ Connective Tissue Diseases Unit, Department of Dermatology, Facultad de Medicina, Pontificia Universidad Católica de Chile, Santiago, Chile

Introduction

Cutaneous manifestations of dermatomyositis (DM) are used as the first diagnostic approach [1–3]. Unfortunately, there is little clarity about the frequency of clinical findings in scalp, periungual folds, and oral cavity and their association with disease activity in DM, but they have been widely described in lupus erythematosus and scleroderma [4, 5].

Scalp involvement is frequent in DM, including psoriasiform changes, poikiloderma, alopecia, itching, and burning sensation. There are some studies on trichoscopic findings in DM patients, but no studies have compared these manifestations on healthy patients. [6–8].

The capillaroscopic results of periungual folds obtained with the dermatoscope are comparable to those described with other instruments such as videocapillaroscopy [9–13]. In addition, it has been observed that periungual telangiectasias and cuticle dystrophy are associated with the level of cutaneous activity in DM patients [12, 13].

The oral mucosa may exhibit manifestations of an underlying systemic disease, and it has been proposed in isolated case reports that gingival telangiectasias could be correlated with disease activity in DM patients [14, 15].

The Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) is a validated and sensitive measurement developed in 2008 that quantifies the skin activity and damage of DM patients [16].

Autoantibodies (Abs) have a crucial role in the pathogenesis of DM and have traditionally been divided into two subsets: myositis-associated Abs (MAAs) and myositis-specific Abs (MSAs) [1]. The presence of these Abs correlates with distinct clinical manifestations, malignancy, and interstitial lung disease, and are predictive of organ manifestations and potentially of prognosis. Anti-TIF1g, Anti-MDA5, and Anti-SAE1/2 are MSAs associated with severe cutaneous involvement in DM, and Anti-Mi2a, Anti-Mi2b, Anti-NPX2, and Anti-SAE1/2 are MSAs associated with intense muscle involvement in DM [1, 2, 17–21].

This study aims to compare the prevalence of trichoscopic, oral, and periungual fold findings between DM and healthy patients, and assess the association of these lesions with disease activity and damage level, to provide a clinical tool to guide therapy.

Materials and methods

This observational, transversal, and analytical study included 40 DM patients over the age of 15 treated at the Immunodermatology Clinic of the Pontificia Universidad Católica de Chile with compatible biopsies that fulfilled the EULAR/ACR 2017 criteria for the diagnosis. All DM patients with inflammatory diseases of the nails, scalp, or oral mucosa not related to DM were excluded.

There are no published studies on the prevalence of scalp lesions in a healthy population. However, according to Jasso-Olivares et al. [8], the prevalence of scalp lesions in DM was 77%. Therefore, considering a 40% difference in scalp involvement between DM and healthy patients that would be clinically relevant, a statistical power of 80% and a 95%

confidence level, the minimum sample size were 28 patients in each group.

DM patients were matched by sex and age with 40 healthy patients (control group), who were recruited from the General Dermatology clinic to compare the prevalence of scalp, periungual folds and oral findings between DM and healthy patients, and to identify clinical manifestations related to DM. The healthy patients included in the control group did not have scalp, nail or oral disease, or family history of autoimmune disease, or any personal signs or symptoms attributable to an autoimmune disease.

All DM and healthy patients underwent a complete medical history, physical examination, and careful examination of hands, periungual folds, scalp and oral cavity with the support of a Dermlite DL4 dermatoscope (3Gen Inc., San Juan Capistrano, CA, USA). Using a high-resolution digital camera with a 60 MMF macro lens, photographs were obtained of five areas of the hands (back of the right and left hand, the palm of the right and left hand, hands in prayer position), ten images of the fingers (dermoscopic images of the proximal periungual folds of each finger), eight areas of the scalp (dermoscopic images of the right and left sides of the frontal, parietal, temporal and occipital regions) and eight areas of the oral cavity (upper and lower gums, tongue, soft and hard palate, right and left buccal mucosa, the floor of the mouth and lips).

Hand manifestations were classified into four categories: Gottron's sign, Gottron's papules, inverse Gottron's sign, and mechanic's hands. Periungual fold findings were classified into five categories: capillary dilation, capillary tortuosity, cuticular hemorrhages, avascular areas, and cuticular hyperkeratosis. Scalp findings were classified into fourteen categories: basal erythema, peripilar cast, interfollicular scaling, poikiloderma, erosions or ulcers, thick tortuous capillaries (capillaries that have two branches, one afferent, and one efferent, the same size or wider than the hair shaft), fine or thick tortuous telangiectasias (capillaries that have only one branch, thinner or thicker than the hair shaft), out-of-focus linear telangiectasias (translucent), in-focus fine linear telangiectasias, bushy capillaries, vascular lake-like structures (ectatic vascular structures), tufting hairs, perifollicular pigmentation, and interfollicular pigmentation. Oral cavity lesions were classified into twelve categories: enanthema, gingivitis, hemorrhage, erosions, aphthous ulcers, leukoplakia, whitish streaks, ovoid palatal patch, gingival cobblestone, gingival telangiectasias, palatal telangiectasias (tortuous, glomerular, or punctate) and pigmentation.

Two dermatologists from our team, blinded to the diagnosis of DM and their activity and damage level, analyzed 2480 photographs from healthy and DM patients registering the presence of these characteristics.

On the same day of the clinical evaluation, each DM patient underwent peripheral venous blood tests that included blood cell counts, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), total creatine kinase (CK), rheumatoid factor (RF), lactate dehydrogenase (LDH), Antinuclear antibody (ANA), four MAAs (Anti-PM-Sc1100, Anti-PM-Sc175, Anti-Ku, and Anti-Ro52), and twelve MSAs (Anti-Mi2a, Anti-Mi2b, Anti-TIF1g, Anti-NPX2, Anti-SAE1/2, Anti-MDA5, Anti-Jo-1, Anti-SRP, Anti-PL-7, Anti-PL-12, Anti-EJ, and Anti-OJ). MAAs and MSAs were performed using the Immunoblot EUROBlotOne/Euroimmun myositis panel, and ANA was done by indirect immunofluorescence antibody technique on HEp-2 substrate.

The disease activity and damage were quantified using the Cutaneous Dermatomyositis Disease Area and Severity Index (CDASI) [16]. This clinician-scored instrument was developed by Werth et al. to estimate disease involvement in 15 different anatomical locations using three activity (erythema, scale, erosion/ulceration) and two damage (poikiloderma, calcinosis) measures. In addition, the presence and severity of Gottron's papules, periungual changes, and alopecia are also captured. Activity and damage scores range from 0 to 100 and 0 to 32, respectively, with higher scores indicating greater disease severity.

The lesions that presented association with DM were compared to laboratory results, and disease activity and damage according to the CDASI scores to determine associations between them.

Statistical analysis was performed with Software R version 4.1.1 [22]. The associations between the clinical manifestations studied and the presence of DM were made using Pearson's Chi-square test or Fisher's exact test. The variables that presented a statistically significant association with DM were then studied and associated with CDASI scores and laboratory results using the Mann–Whitney *U* test. A *p*-value < 0.05 was considered significant.

The ethics committee approved this study of the Pontificia Universidad Católica de Chile under the number 200803003. All patients signed written informed consent before enrollment.

Results

Forty DM patients and 40 healthy subjects were evaluated between November 2020 and September 2021. The main characteristics of DM patients and the treatment they were receiving at the time of the clinical evaluation are summarized in Tables 1 and 2.

Table 1 The main characteristics of DM patients included in the study

Sex	<i>n</i> (%)
Female	30 (75.0)
Male	10 (25.0)
Age	Mean ± SD (years)
Age at the time of evaluation	49.8 ± 17.31
Age of onset of the disease	46.0 ± 17.30
Antibodies	<i>n</i> (%)
ANA titers > = 1/80	34 (85.0)
Positive myositis specific or associated	36 (90.0)
Negative myositis antibodies	4 (10.0)
Myositis-specific antibodies	33 (82.5)
Myositis-associated antibodies	17 (42.5)
MSAs associated with severe cutaneous involvement*	24 (60.0)
MSAs associated with severe muscle involvement**	12 (30.0)
Myositis-associated antibodies	<i>n</i> (%)
Anti-PM-Sc1100	0 (0)
Anti-PM-Sc175	2 (5.0)
Anti-Ku	5 (12.5)
Anti-Ro52	11 (27.5)
Myositis-specific antibodies	<i>n</i> (%)
Anti-Mi2a	5 (12.5)
Anti-Mi2b	4 (10.0)
Anti-TIF1g	17 (42.5)
Anti-NPX2	2 (5.0)
Anti-SAE1/2	4 (10.0)
Anti-MDA5	3 (7.5)
Anti-SRP	0 (0)
Anti-synthetase antibodies	
Anti-Jo-1	1 (2.5)
Anti-PL-7	4 (10.0)
Anti-PL-12	0 (0)
Anti-EJ	1 (2.5)
Anti-OJ	2 (5.0)

*Myositis-specific Abs associated with severe cutaneous involvement: Anti-TIF1g, Anti-MDA5, Anti-SAE1/2

**Myositis-specific Abs associated with severe muscle involvement: Anti-Mi2a, Anti-Mi2b, Anti-NPX2, Anti-SAE1/2

Clinical manifestations in DM patients

When analyzing clinical manifestations in DM patients, it was found that thirty-four (85.0%) patients presented findings in hands, thirty-one (77.0%) patients had periungual involvement, forty (100%) patients had trichoscopic findings, and forty (100%) patients had oral lesions.

Compared with the control group, DM patients had a statistically more frequent presence of mechanic's hands, Gottron's sign and Gottron's papules in hands (Fig. 1);

Table 2 Systemic and topical treatment received by DM patients at the time of clinical evaluation

Systemic treatment	N (%)
Hydroxychloroquine	19 (47.5)
Methotrexate	10 (25.0)
Mycophenolate mofetil	13 (32.5)
Azathioprine	4 (10.0)
Hydroxychloroquine and methotrexate	5 (12.5)
Hydroxychloroquine and mycophenolate mofetil	5 (12.5)
Hydroxychloroquine and azathioprine	3 (7.5)
Rituximab	1 (2.5)
Belimumab	1 (2.5)
Systemic corticosteroids	16 (40.0)
No systemic treatment	6 (15.0)
Topical treatment	N (%)
Topical corticosteroids on the scalp	20 (50.0)

capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds (Fig. 2); basal erythema, poikiloderma, thick tortuous capillaries, fine or thick tortuous telangiectasias, and in-focus fine linear telangiectasias in scalp (Fig. 3); and enanthema (mainly located in the palatal area), gingivitis, gingival cobblestones, gingival telangiectasias, and palatal telangiectasias on the oral cavity (Fig. 4)(Table 3).

Mechanic's hands and Gottron's papules in hands, capillary tortuosity in periungual folds, poikiloderma in scalp, and gingival telangiectasias in oral cavity were present exclusively in DM patients with a significant association. Inverse Gottron's sign in hands, tufted hairs on scalp, and

ovoid palatal patch in oral cavity were present exclusively in DM patients, but no significant association was found (Table 3).

Clinical manifestations and CDASI score

The presence of mechanic's hands, Gottron's sign and Gottron's papules in hands; capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds; thick tortuous capillaries in scalp; and gingival telangiectasias in the oral cavity were associated with a higher CDASI activity score (Table 4).

Gottron's sign and Gottron's papules in hands; capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds; basal erythema in scalp; and gingival telangiectasias in the oral cavity were associated with a higher CDASI damage score (Table 5).

High-CDASI-activity-score clinical manifestations and autoantibodies

When comparing MSAs or MAAs with the dermoscopic and clinical findings significantly associated with a higher CDASI activity score, it was found that the presence of MSAs related to severe cutaneous involvement in the literature (Anti-TIF1g, Anti-MDA5, Anti-SAE1/2) was associated with mechanic's hands (p -value = 0.005953) and Gottron's papules (p -value = 0.01310) in hands; capillary dilation (p -value = 0.02799), cuticular hemorrhages

Fig. 1 Hand findings associated with DM. Panel **a** Gottron's sign. Panel **b** Gottron's papules. Panel **c** Mechanic's hands



Fig. 2 Periungual folds findings on polarized dermatoscopy ($\times 10$) associated with DM. Panel **a** Capillary dilation (black arrows), capillary tortuosity (red arrows), and cuticular hyperkeratosis (yellow arrows). Panel **b** Capillary tortuosity (red arrows), cuticular hyperkeratosis (yellow arrows), and cuticular hemorrhages (blue arrows). Panel **c** Cuticular hyperkeratosis (yellow arrows), and cuticular hemorrhages (blue arrows). Panel **d** Capillary dilation (black arrows), and avascular areas (green arrows). Panel **e** Cuticular hyperkeratosis (yellow arrows), cuticular hemorrhages (blue arrows), and avascular areas (green arrows)

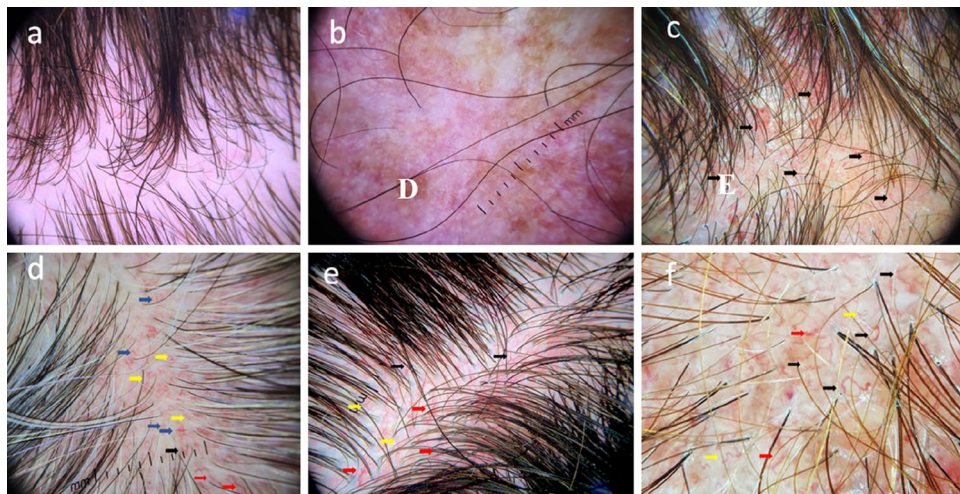
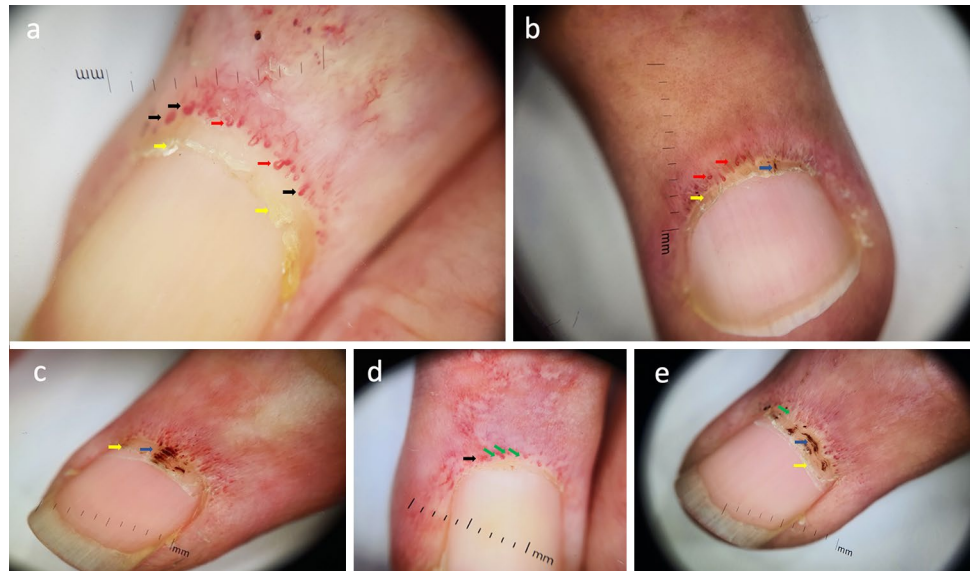


Fig. 3 Trichoscopic findings on polarized dermatoscopy ($\times 10$) associated with DM: Panel **a** Basal erythema. Panel **b** Poikiloderma (hypopigmentation, hyperpigmentation, telangiectasias and atrophy). Panel **c** Thick tortuous capillaries (black arrows). Panel **d** Thick tortuous capillaries (black arrows), in-focus fine linear telangiectasias

(blue arrows), fine tortuous telangiectasias (yellow arrows) and thick tortuous telangiectasias (red arrows). Panel **e** and **f** Thick tortuous capillaries (black arrows), fine tortuous telangiectasias (yellow arrows) and thick tortuous telangiectasias (red arrows)

(p -value = 0.01369), and avascular areas in periungual folds (p -value = 0.01998); and thick tortuous capillaries in scalp (p -value = 0.002949).

On the other hand, capillary dilation was associated with Anti-TIF1 antibodies (p -value = 0.04889), cuticular hemorrhages were associated with MSAs (p -value = 0.03276) and Anti-TIF1 antibodies (p -value = 0.01894), and avascular areas were associated with the lack of MAAs (p = 0.0489).

Finally, thick tortuous capillaries in the scalp were associated with Anti-TIF1 antibodies (p = 0.01194), and gingival telangiectasias were associated with the lack of

ANA antibodies (p = 0.021121) (Supplemental Tables 1 and 2).

CDASI score, autoantibodies, and laboratory abnormalities

The patients with positive Anti-NPX2 antibodies (2 patients) had a lower level of activity compared to those lacking Anti-NPX2 antibody (p -value = 0.02979) (Table 4 and Supplemental Table 3). The presence of positive Abs related to severe cutaneous disease was associated with higher activity (p -value = 0.03962). In contrast, Abs related

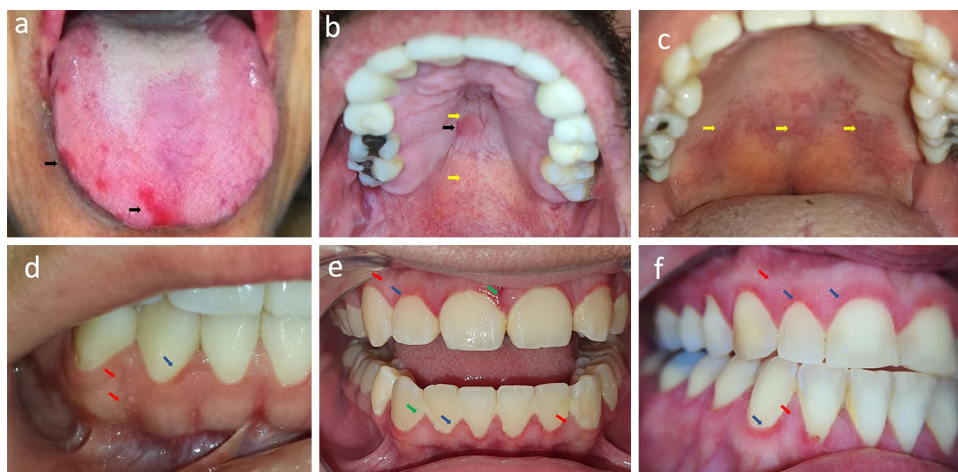


Fig. 4 Oral cavity findings associated with DM. Panel **a** Enanthema of the tongue (black arrows). Panel **b** Enanthema of the palate (black arrows) and palatal telangiectasias (yellow arrows). Panel **c** Palatal tortuous, glomerular, or punctate telangiectasias (yellow

arrows). Panel **d** Gingival Cobblestone (red arrows) and gingival telangiectasias (blue arrows). Panel **e** Gingivitis (green arrows) and gingival telangiectasias (blue arrows). Panel **f** Gingival Cobblestone (red arrows) and gingival telangiectasias (blue arrows)

to intense muscle involvement were associated with a lower activity level. (p -value = 0.04959) (Table 6).

Even though different laboratory abnormalities (anemia, leukopenia, thrombocytopenia, and LDH, ESR, RF, and total CK elevation) were related to CDASI activity and damage scores, the associations between these variables were not statistically significant.

Discussion

Gottron's sign and Gottron's papules were found to be significantly associated with DM, which coincides with the literature where they have been classified as pathognomonic of DM [17].

Previous studies have reported alterations in periungual folds in a high proportion of DM patients, such as elongated capillaries, avascular areas, disorganized vascular architecture, tortuous capillaries, dilated capillaries, and periungual hemorrhages [23]. However, these manifestations had not been yet compared with healthy patients. All the alterations in the periungual capillaries analyzed in this study were significantly associated with DM. As far as we know, this study is the first to correlate the periungual findings of the handheld dermatoscope as a nail-fold capillaroscopic instrument and the presence of any myositis antibodies in DM patients. It is also the first research to link the presence of cuticular hemorrhages with the presence of MSAs and Anti-TIF1 antibodies, capillary dilation with Anti-TIF1 antibodies, and avascular areas in periungual fold with the lack of MAAs; thick tortuous capillaries in scalp with Anti-TIF1 antibodies; and gingival telangiectasias with the lack of ANA antibodies. Naoki Mugii et al. [24] did not

find any association between Anti-TIF1, Anti-Mi2 or Anti-synthetase antibodies and enlarged capillaries, hemorrhages, disorganization of the normal capillary distribution, capillary loss, and tortuous, crossed, and ramified capillaries on videocapillaroscopy in 50 DM patients. The difference between our results and the findings of Mugii and his group could be explained by the fact that periungual abnormalities could be reversed and modified by treatment, as has been demonstrated in the capillaroscopy of 11 DM patients with anti-MDA5 antibodies at baseline and after treatment [25]. The physical examination at a given time can be affected by multiple factors, such as the activity of the disease and the use or not of systemic or topical therapy, among others.

The most frequently reported DM scalp dermoscopic findings are erythema, scaling, alopecia, poikiloderma, thick tortuous capillaries, bushy vessels, and linear vessels [7, 8, 23], which is similar to the conclusions of our study where all patients presented some trichoscopic alteration and the most frequently observed signs were basal erythema (87.5%), in-focus fine linear telangiectasias (77.5%), out-of-focus fine linear telangiectasias (67.5%), fine and thick tortuous telangiectasias (65.0%) and peripilar cast (62.5%). To the best of our knowledge, no studies have been published comparing trichoscopic findings between healthy and DM patients, and publications on trichoscopy in DM have included a maximum of 31 patients [8]. Therefore, this is the one with the highest number of cases. Chanprapaph et al. [26] recently compared trichoscopic findings in DM, systemic lupus, and systemic sclerosis patients. They found that perifollicular reddish-brown pigmentation and the presence of blood vessels with microaneurysms were present exclusively in DM patients. In addition, they observed that scalp desquamation favored the diagnosis of

Table 3 Summary of hand, periungual fold, trichoscopic and oral findings seen in healthy subjects and DM patients

	Healthy subjects, n (%)	DM patients, n (%)	p-value
<i>Hand findings</i>			
Gottron's sign	2 (5.0)	33 (82.5)	<0.0001
Gottron's papules	0 (0)	17 (42.5)	<0.0001
Mechanic's hands	0 (0)	9 (22.5)	<0.05*
Inverse Gottron's sign	0 (0)	3 (7.5)	0.2405*
<i>Periungual fold findings</i>			
Capillary dilation	2 (5.0)	21 (52.5)	<0.0001
Capillary tortuosity	0 (0)	19 (47.5)	<0.0001
Cuticular hemorrhages	9 (22.5)	22 (55.0)	<0.05
Avascular areas	1 (2.5)	19 (47.5)	<0.0001
Cuticular hyperkeratosis	11 (27.5)	26 (65.0)	<0.05
<i>Trichoscopic findings</i>			
Basal erythema	27 (67.5)	35 (87.5)	<0.05
Peripilar cast	26 (65.0)	25 (62.5)	0.8161
Interfollicular scaling	3 (7.5)	8 (20.0)	0.1045
Poikiloderma	0 (0)	6 (15.0)	<0.05*
Erosions or ulcers	1 (2.5)	4 (10.0)	0.3589*
Thick tortuous capillaries	9 (22.5)	19 (47.5)	<0.05
Fine or thick tortuous telangiectasias	13 (32.5)	26 (65.0)	<0.05
Out-of-focus linear telangiectasias	31 (77.5)	27 (67.5)	0.3166
In-focus fine linear telangiectasias	20 (50.0)	31 (77.5)	<0.05
Bushy capillaries	2 (5.0)	5 (12.5)	0.4315*
Vascular lake-like structures	3 (7.5)	8 (20.0)	0.1045
Tufting hairs	0 (0)	4 (10.0)	0.1156*
Perifollicular pigmentation	8 (20.0)	8 (20.0)	1
Interfollicular pigmentation	8 (20.0)	7 (17.5)	0.7745
<i>Oral findings</i>			
Enanthema	2 (5.0)	13 (32.5)	<0.05
Gingivitis	9 (22.5)	22 (55.0)	<0.05
Hemorrhage	1 (2.5)	0 (0)	1*
Erosions	0 (0)	1 (2.5)	1*
Aphthous ulcers	0 (0)	0 (0)	–
Leukoplakia	1 (2.5)	5 (12.5)	0.2007*
Whitish streaks	4 (10.0)	8 (20.0)	0.2104
Ovoid palatal patch	0 (0.0)	5 (12.5)	0.05474*
Gingival Cobblestone	15 (37.5)	29 (72.5)	<0.05
Gingival telangiectasias	0 (0.0)	21 (52.5)	<0.0001
Palatal telangiectasias	12 (30.0)	38 (95.0)	<0.0001
Pigmentation	5 (12.5)	7 (17.5)	0.5312

*p-value calculated with Fisher exact test

DM over lupus and systemic sclerosis, especially when it was perifollicular. In our study, no association was found between perifollicular reddish-brown pigmentation and DM, which could be explained by the difficulty in distinguishing a subtle pigmentation from the translucency of the hair shaft emerging from the skin or by differences in the phototypes of our patients, which modifies their skin pigmentary response to different inflammatory dermatoses, as has been

described in patients with DM and darker skin that present with predominant hyperpigmentation [27].

On the other hand, aneurysmal blood vessels refer to dilated linear, serpentine, or tortuous capillaries with an ectatic portion resembling an aneurysm and have also been described as ectatic vascular lakes/structures. In our study, aneurysmal blood vessels were present in eight DM and three healthy patients, and no significant association

Table 4 Clinical findings in hands, periungual folds, scalp, and oral cavity in DM patients that are significantly associated with a higher CDASI activity score

Clinical findings	CDASI activity score (points)		<i>p</i> -value
	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Hands			
Gottron's sign	33 (3–75)	22 (3–25)	< 0.01
Gottron's papules	52 (28–75)	23 (3–51)	< 0.0001
Mechanic's hands	48 (25–75)	28 (3–74)	< 0.05
Periungual folds	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Capillary dilation	37 (22–75)	17 (3–56)	< 0.001
Capillary tortuosity	39 (22–75)	24 (3–56)	< 0.05
Cuticular hemorrhage	38 (4–75)	20 (3–56)	< 0.05
Avascular areas	39 (23–75)	22 (3–56)	< 0.001
Cuticular hyperkeratosis	37 (3–75)	23 (3–52)	< 0.05
Scalp	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Thick tortuous capillaries	37 (12–75)	25 (3–74)	< 0.05
Oral cavity	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Gingival telangiectasias	39 (8–75)	25 (3–59)	< 0.05

Table 5 Clinical findings in hands, periungual folds, scalp, and oral cavity in DM patients that are significantly associated with a higher CDASI damage score

Clinical findings	CDASI damage score (points)		<i>p</i> -value
	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Hands			
Gottron's sign	12 (1–21)	3 (0–15)	< 0.05
Gottron's papules	15 (6–21)	5 (0–16)	< 0.0001
Periungual folds	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Capillary dilation	13 (2–21)	4 (0–17)	< 0.001
Capillary tortuosity	15 (6–21)	4 (0–17)	< 0.0001
Cuticular hemorrhage	13 (0–21)	4 (1–17)	< 0.001
Avascular areas	15 (2–21)	5 (0–17)	< 0.0001
Cuticular hyperkeratosis	13 (2–21)	4 (0–14)	< 0.05
Scalp	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Basal erythema	11 (1–21)	3 (0–12)	< 0.05
Oral cavity	Lesion present Median (min–max)	Lesion absent Median (min–max)	
Gingival telangiectasias	13 (1–18)	5 (0–21)	< 0.05

could be found. In addition, the presence of peripilar cast in DM and healthy patients was similar (62.5 and 65.0%, respectively), which may be because 50% of the patients with DM were receiving topical corticosteroids on the scalp at the time of the clinical evaluation, decreasing the prevalence of this sign in this group of patients. In contrast, interfollicular desquamation was more frequent in DM, but no significant association was found. Scaling on the scalp is frequent in patients who consult in the context of mild to moderate seborrheic dermatitis. It could be aggravated by psychological stress from the COVID-19 pandemic when our study was done, explaining the frequency in the

control group [28]. Finally, our results showed that in-focus capillaries were significantly associated with the diagnosis of DM, which has not been reported yet in literature to our knowledge.

The most described oral findings in DM are gingival telangiectasias and ovoid palatal patch [29, 30]. Gingival telangiectasias were present exclusively in DM patients in our study. There are many case reports in which gingival telangiectasias have been the initial signs of DM, suggesting an essential role in early diagnosis, especially in juvenile DM patients [31–34]. Gingival telangiectasias could be associated with cutaneous activity and refractory

Table 6 CDASI activity score according to the presence or absence of antibodies

Antibodies	CDASI activity score (points)		<i>p</i> -value
	Antibody positive Median (min–max)	Antibody negative Median (min–max)	
Anti-Mi2	25.5 (3–42)	33 (3–75)	0.1446
Anti-TIF1g	35 (3–75)	26 (3–74)	0.3112
Anti-NPX2	5 (3–7)	30.5 (3–75)	0.02979
Anti-SAE1/2	40 (8–55)	29 (3–75)	0.7522
Anti-MDA5	56 (29–57)	28 (3–75)	0.1297
Anti-Ku	42 (12–59)	29 (3–75)	0.6975
Anti-PMscl100	–	29 (3–75)	–
Anti-PMscl75	14 (3–25)	30.5 (3–75)	0.1446
Anti-Jo1	52 (52–52)	29 (3–75)	0.3404
Anti-SRP	–	29 (3–75)	–
Anti-Pl17	42 (15–57)	28.5 (3–75)	0.4568
Anti-Pl12	–	29 (3–75)	–
Anti-EJ	22 (22–22)	29 (3–75)	0.4354
Anti-OJ	37 (15–59)	29 (3–75)	0.7562
Anti-Ro52	26 (3–75)	32 (3–74)	0.7048
Myositis-specific	29 (3–75)	25 (9–74)	0.9716
Myositis-associated	26 (3–75)	34 (4–74)	0.2338
Anti-synthetase	42 (15–59)	28.5 (3–75)	0.3631
Any myositis antibody	29 (3–75)	38 (24–74)	0.4705
Anti-TIF1g, Anti-MDA5, Anti-SAE1/2*	36 (3–75)	23.5 (3–74)	0.03962
Anti-Mi2, Anti-NPX2, Anti-SAE1/2**	24.5 (3–55)	34.5 (0–75)	0.04959
ANA	28.5 (3–75)	46.5 (24–74)	0.1296

*Myositis-specific Abs related to severe cutaneous involvement in the literature: Anti-TIF1g, Anti-MDA5, Anti-SAE1/2

**Myositis-specific Abs related to severe muscle involvement in the literature: Anti-Mi2a, Anti-Mi2b, Anti-NPX2, Anti-SAE1/2

disease, representing a possible site for evaluating DM activity and response to therapy, similar to periungual folds [14, 31]. Gingival telangiectasia has been reported in 20% of adults with DM [35], which contrasts with our study, where a prevalence of 52.5% was seen. This difference may be explained by the use of high-quality photographs to find our results. This is the first time that gingival telangiectasias are related to a higher CDASI activity and damage score in DM patients.

The presence of palatal telangiectasias was also associated with DM, which could be explained by the same pathophysiological phenomena underlying the development of gingival telangiectasias, thus adding a new area to explore that could guide the diagnosis.

Gingivitis has been described as epiphenomena to capillary changes rather than a primary feature in DM [36]. However, our study showed a statistically significant association of DM with the presence of gingival telangiectasias and gingivitis, just as demonstrated in patients with lupus [4].

The ovoid palatal patch is a sign described in 2016 by Fiorentino [37]. In our study, this sign was present exclusively in DM patients (5 patients). Still, no significant association with the disease was found (p -value = 0.05474), possibly due to the low number of patients with this finding.

Other oral manifestations associated with DM described in the literature are erythema, ulcers, and leukoplakia-like lesions [38, 39]. However, in our study, no statistically significant associations were found with these findings, even though erythema could be included as a form of enanthema. In contrast, a significant association was observed for enanthema (any rash on a mucous membrane) and gingival cobblestones with DM, which has not been previously reported in the literature.

Finally, the presence of mechanic's hands, Gottron's sign, and Gottron's papules in hands; capillary dilation, capillary tortuosity, cuticular hemorrhage, avascular areas, and cuticular hyperkeratosis in periungual folds; thick tortuous capillaries in scalp; and gingival telangiectasias in the oral cavity suggest active disease given their association with a

higher CDASI activity score. Assessing these clinical signs in daily practice is more accessible and more straightforward than applying the CDASI, so this information could be a helpful tool when determining DM activity.

Patients with MSAs associated with severe cutaneous involvement (Anti-TIF1g, Anti-MDA5, and Anti-SAE1/2) had with higher CDASI activity scores. Conversely, patients with MSAs associated with intense muscle involvement in DM (Anti-Mi2a, Anti-Mi2b, Anti-NPX2, and Anti-SAE1/2) had a lower activity level than those with these negative MSAs, giving clinicians new information related to disease activity. Given that most DM patients were undergoing treatment at the time of the clinical evaluation, some manifestations could have been negativized, explaining the lack of association between certain findings and the antibodies studied.

This study is the first to compare trichoscopic, oral and periungual findings between DM and healthy patients. In addition, most of the DM patients had scalp, oral and periungual involvement, confirming that these areas should be consistently examined to identify patients with an active DM, guiding therapeutic decisions. Lastly, one of the limitations of our investigation was that biopsies of the described findings were not taken, which would have allowed a better characterization of these signs.

Funding sources

Research Grant from the Department of Dermatology and The School of Medicine of Pontificia Universidad Católica de Chile, Santiago, Chile.

Supplementary Information The online version contains supplementary material available at <https://doi.org/10.1007/s00403-023-02554-0>.

Acknowledgements The authors wish to acknowledge the Department of Dermatology and The School of Medicine of Pontificia Universidad Católica de Chile for the Research Grant and all patients of this study who have given written informed consent to the publication of their case details.

Author contributions The authors confirm contribution to the paper as follows: study conception and design: Catalina Salgueiro, María José Poblete, Álvaro Abarzúa, Cristián Vera-Kellet; data collection: Catalina Salgueiro, María José Poblete, Cristián Vera-Kellet. Author; analysis and interpretation of results: Catalina Salgueiro, María José Poblete, Christian Robles-Silva, Álvaro Abarzúa, Cristián Vera-Kellet. draft manuscript preparation: Catalina Salgueiro, María José Poblete, Christian Robles-Silva, Cristián Vera-Kellet. All authors reviewed the results and approved the final version of the manuscript.

Data availability The datasets generated during and/or analysed during the current study are available from the corresponding author on reasonable request.

Declarations

Competing interests The authors declare no competing interests.

Conflicts of interest The authors have no conflict of interest to declare.

Ethical statement IRB approval status: Reviewed and approved by Ethics Committee of Pontificia Universidad Católica de Chile under the number 200803003. Reprint requests: Cristián Vera-Kellet.

References

- DeWane ME, Waldman R, Lu J (2020) Dermatomyositis: Clinical features and pathogenesis. *J Am Acad Dermatol* 82(2):267–281
- Aussy A, Boyer O, Cordel N (2017) Dermatomyositis and immune-mediated necrotizing myopathies: a window on autoimmunity and cancer. *Front Immunol*. 8:992
- Furst DE, Amato AA, Iorga ŞR, Gajria K, Fernandes AW (2012) Epidemiology of adult idiopathic inflammatory myopathies in a U.S. managed care plan. *Muscle Nerve*. 45(5):676–683
- Del Barrio-Díaz P, Reyes-Vivanco C, Cifuentes-Mutinelli M, Manríquez J, Vera-Kellet C (2020) Association between oral lesions and disease activity in lupus erythematosus. *J Eur Acad Dermatol Venereol* 34(2):349–356
- Denton CP, Khanna D (2017) Systemic sclerosis. *The Lancet* 390(10103):1685–1699
- Miteva M, Tosti A (2012) Hair and scalp dermatoscopy. *J Am Acad Dermatol* 67(5):1040–1048
- Tilstra JS, Prevost N, Khera PEJ (2009) Scalp dermatomyositis revisited. *Arch Dermatol* 145(9):1062–1063
- Jasso-Olivares JC, Tosti A, Miteva M, Domínguez-Cherit J, Díaz-González JM (2017) Clinical and dermoscopic features of the scalp in 31 patients with dermatomyositis. *Skin Appendage Disord* 3(3):119–124
- Bergman R, Sharony L, Schapira D, Nahir MA, Balbir-Gurman A (2003) The handheld dermatoscope as a nail-fold capillaroscopic instrument. *Arch Dermatol* 139(8):1027–1030
- Bauersachs RM, Löbner F (1997) The poor man's capillary microscope. A novel technique for the assessment of capillary morphology. *Ann Rheum Dis*. 56(7):435–437
- Elmas ÖF, Okçu M, Demirbaş A, Akdeniz N (2020) Handheld dermatoscopy as an easy-to-use capillaroscopic instrument in rheumatoid arthritis: A cross-sectional study. *Turk J Med Sci* 50(6):1540–1545
- Bertolazzi C, Cutolo M, Smith V, Gutierrez M (2017) State of the art on nailfold capillaroscopy in dermatomyositis and polymyositis. *Semin Arthritis Rheum* 47(3):432–444
- Barth Z, Witczak BN, Flatø B, Koller A, Sjaastad I, Sanner H (2018) Assessment of Microvascular Abnormalities by Nailfold Capillaroscopy in Juvenile Dermatomyositis After Medium- to Long-Term Followup. *Arthritis Care Res (Hoboken)* 70(5):768–776
- Ghali FE, Stein LD, Fine J, David D, Burkes EJ, McCauliffe DP (1999) Gingival telangiectases: an underappreciated physical sign of juvenile dermatomyositis. *Arch Dermatol*. 135(11):1370–4
- Mugii N, Hasegawa M, Matsushita T et al (2011) Association between nail-fold capillary findings and disease activity in dermatomyositis. *Rheumatology (Oxford)* 50(6):1091–1098
- Anyanwu CO, Fiorentino D, Chung L et al (2015) Validation of the Cutaneous Dermatomyositis Disease Area and Severity Index: characterizing disease severity and assessing responsiveness to clinical change. *Br J Dermatol* 173(4):969–974

17. Mainetti C, Terziroli Beretta-Piccoli BSC (2017) Cutaneous manifestations of dermatomyositis: a comprehensive review. *Clin Rev Allergy Immunol* 53(3):337–356
18. Marasandra Ramesh H, Gude SS, Venugopal S, Peddi NC, Gude SS, Vuppalapati S (2022) The role of myositis-specific autoantibodies in the dermatomyositis spectrum. *Cureus* 14(3):1–10
19. Hodgkinson LM, Wu TT, Fiorentino DF (2021) Dermatomyositis autoantibodies: how can we maximize utility? *Ann Transl Med* 9(5):433–433
20. Chen X, Zhang L, Jin Q et al (2022) The clinical features and prognoses of anti-MDA5 and anti-aminoacyl-tRNA synthetase antibody double-positive dermatomyositis patients. *Front Immunol* 13:1–10
21. Gono T, Kuwana M (2020) Current understanding and recent advances in myositis-specific and -associated autoantibodies detected in patients with dermatomyositis. Vol 16. Taylor & Francis
22. R Core Team (2021). R: A language and environment for statistical computing. R Foundation for Statistical Computing, Vienna, Austria. URL <https://www.R-project.org/>
23. Żychowska M, Reich A (2022) Dermoscopy and trichoscopy in dermatomyositis—a cross-sectional study. *J Clin Med* 11(2)
24. Mugii N, Hasegawa M, Matsushita T et al (2011) Association between nail-fold capillary findings and disease activity in dermatomyositis. *Rheumatology (Oxford)* 50(6):1091–1098
25. Hamaguchi Y, Mugii N, Matsushita T, Takehara K (2021) Long-term changes in nail fold capillary abnormalities and serum fibroblast growth factor 23 levels in dermatomyositis patients with anti-melanoma differentiating antigen 5 antibody. *J Dermatol* 48(1):106–109
26. Chanprapaph K, Limtong P, Ngamjanyaporn P, Suchonwanit P (2022) Trichoscopic signs in dermatomyositis, systemic lupus erythematosus, and systemic sclerosis: a comparative study of 150 patients. *Dermatology* 238(4):677–687
27. Neema S, Kothari R, Kashif AW, Vashisht D, Vasudevan B (2022) Dermoscopy of dermatomyositis in dark skin. *Dermatol Pract Concept* 12(1):e2022013
28. Mahadi AR, Rafi MA, Shahriar T et al (2022) Association between hair diseases and COVID-19 pandemic-related stress: a cross-sectional study analysis. *Front Med (Lausanne)* 9:1–9
29. Castillo RL, Femia AN (2021) Covert clues: the non-hallmark cutaneous manifestations of dermatomyositis. *Ann Transl Med* 9(5):436–436
30. Milani-Nejad N, Kaffenberger J (2020) Gingival telangiectasia of dermatomyositis. *Br J Dermatol* 183(4):e93
31. Rider LG, Atkinson JC (2009) Gingival and periungual vasculopathy of juvenile dermatomyositis. *N Engl J Med* 360(15):e21
32. Gonçalves LM, Bezerra-Júnior JRS, Gordón-Núñez MA et al (2011) Oral manifestations as important symptoms for juvenile dermatomyositis early diagnosis: A case report. *Int J Paediatr Dent* 21(1):77–80
33. Shikino K, Hanazawa N, Noda K, Ikusaka M (2021) Gingival telangiectases due to dermatomyositis. *J Gen Fam Med* 22(1):49–50
34. Savioli C, Silva CAA, Fabri GMC et al (2010) Gingival capillary changes and oral motor weakness in juvenile dermatomyositis. *Rheumatology* 49(10):1962–1970
35. Márton K, Hermann P, Dankó K, Fejérdy P, Madléna M, Nagy G (2005) Evaluation of oral manifestations and masticatory force in patients with polymyositis and dermatomyositis. *J Oral Pathol Med* 34(3):164–169
36. Khan H, Mehta P, Gupta L (2021) Juvenile dermatomyositis with gingival vasculopathy. *Clin Rheumatol* 40(8):3369–3370
37. Bernet LL, Lewis MA, Rieger KE, Casciola-Rosen L, Fiorentino DF (2016) Ovoid palatal patch in dermatomyositis: A novel finding associated with Anti-TIF1 γ (p155) antibodies. *JAMA Dermatol* 152(9):1049–1051
38. Geist SMRY, Tanaka TI (2014) Oral lichen planus in a dermatomyositis patient that resolved after intravenous immunoglobulin therapy. *Oral Surg Oral Med Oral Pathol Oral Radiol* 118(4):e111–e114
39. Healy CM, Tobin AM, Kirby B, Flint SR (2006) Oral lesions as an initial manifestation of dermatomyositis with occult malignancy. *Oral Surg Oral Med Oral Pathol Oral Radiol Endodontology* 101(2):184–187

Publisher's Note Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Springer Nature or its licensor (e.g. a society or other partner) holds exclusive rights to this article under a publishing agreement with the author(s) or other rightsholder(s); author self-archiving of the accepted manuscript version of this article is solely governed by the terms of such publishing agreement and applicable law.