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Dermatological findings in common rheumatologic diseases in children

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Abstract
The aim of this study is to outline the common dermatological findings in pediatric rheumatologic diseases. A total of 45 patients, nineteen with juvenile idiopathic arthritis (JIA), eight with Familial Mediterranean Fever (FMF), six with scleroderma (SSc), seven with systemic lupus erythematosus (SLE), and five with dermatomyositis (DM) were included. Control group for JIA consisted of randomly chosen 19 healthy subjects of the same age and gender. The age, sex, duration of disease, site and type of lesions on skin, nails and scalp and systemic drug use were recorded. $\chi 2$ test was used. The most common skin findings in patients with psoriatic JIA were flexural psoriatic lesions, the most common nail findings were periungual desquamation and distal onycholysis, while the most common scalp findings were erythema and scaling. The most common skin finding in patients with oligoarthritis was photosensitivity, while the most common nail finding was periungual erythema, and the most common scalp findings were erythema and scaling. We saw urticarial rash, dermatographism, nail pitting and telogen effluvium in one patient with systemic arthritis; and photosensitivity, livedo reticularis and periungual erythema in another patient with RF-negative polyarthritis. Vascular skin lesions like Raynaud's phenomenon, livedo reticularis, palmar erythema, periungual telangiectasia and nailfold abnormalities were common in SLE, DM and SSc. Patients with FMF displayed signs of atopy. Specific skin lesions can be the peculiar features of rheumatologic diseases in pediatric population. Since it is not always easy to perform biopsy in children to confirm skin involvement of a rheumatologic disease, skin findings can help both dermatologists and rheumatologists in diagnosis.

Keywords: Juvenile idiopathic arthritis, familial mediterranean fever, scleroderma, systemic lupus erythematosus, dermatomyositis

Introduction

Specific skin lesions can be the hallmark and initial involvement area of certain common rheumatologic diseases. Skin involvement can alert both the dermatologists and rheumatologists about systemic involvement and prognostic outcome. On the other hand, skin lesions may diminish after the initiation of systemic therapy or escalate as an adverse reaction to immune suppressants, biologics and targeted therapies for the treatment of rheumatologic diseases. Since it is not always easy to perform skin biopsy in pediatric population to confirm skin involvement of a rheumatologic disease, skin findings in such diseases can help physicians. In this regard, they may be helpful in the diagnosis, follow-up of the disease course and the adverse skin reactions to systemic drugs. On the other hand, there is scarce information in the literature on skin findings of rheumatologic diseases in children [1-8]. In this study, the involvement of the skin, nail and scalp of 19 patients with juvenile idiopathic arthritis (JIA) (Of these 19 patients, 1 of them had systemic arthritis, 1 of them had RF-negative polyarthritis, 7 of them had psoriatic arthritis, 6 of them had oligoarthritis, 2 of them had enthesitis-related arthritis and 2 of them had undifferentiated arthritis), 8 patients with familial mediterranean fever (FMF), 7 patients with systemic lupus erythematosus (SLE), 6 patients with scleroderma (SSc) [3 patients with diffuse SSc and 3 patients with limited SSc], and 5 patients with dermatomyositis (DM) were evaluated.

Material and Methods

The study was approved and performed in accordance with the guidelines of the institutional review board at our hospital and all patients signed informed consent forms before enrollment in the study (IRB#B.10.1.TKH.4.34.H.GP.0.01/107). Nineteen patients with JIA, 8 patients with FMF, 7 patients with SLE, 6 patients with SSc, and 5 patients with DM were included in this study. Nineteen controls were included in this study. Control group for JIA consisted of randomly chosen healthy subjects of the same age

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and gender without any skin and rheumatologic diseases.

The complete medical data of patients diagnosed with these disorders in 2017 were evaluated. Patients with JIA and FMF were diagnosed by rheumatologists clinically while the diagnosis of the patients with SLE, DM and SSc were verified by skin biopsy. Skin, nail and scalp findings were evaluated by a manual dermatoscope with x10 magnification in addition to naked eye. All photographs were taken from each patient using a contact & non-contact, polarized, manual dermatoscope (Dermlite DL4 PigmentBoost Plus X10; 3 Gen, San Juan Capistrano, CA, USA) equipped with magnetically attached smartphone. The age, sex, duration of disease, site and type of lesions on skin, nails and scalp and systemic drug use of patients prior to admission were recorded.

All statistical analyses were performed using SPSS version 15.0 (SPSS Inc, Chicago, IL, USA). For continuous variables, normality tests were performed with Kolmogorov-Smirnov test. Chi-Square test was used to compare (the categorical data) clinical differences with other healthy children. The level of significance for all analyses was set at $p<0.05$.

Results

A total of 45 patients were admitted to the hospital with diagnosis of a rheumatologic disease. Clinical characteristics such as the age, sex, duration of disease, prior systemic drug use and the location and type of lesions on skin, nails and scalp were summarized in Tables 1 and 2.

Nine (47.4%) patients with JIA were female while 10 (52.6%) patients were male. The mean age of the patients was 11.5±3.3. The control group for JIA consisted of randomly chosen 10 (52.6%) male and 9 (47.4%) female patients. The mean age of this group was 10.5±4.6. Of the 19 patients with JIA, one of them had systemic arthritis (adolescent-onset Still's disease), one of them had RF-negative polyarthritis, seven of them had psoriatic arthritis, six of them had oligoarthritis (persistent or extended), two of them had enthesitis-related arthritis and two of them had undifferentiated arthritis.The most common findings in JIA patients were erythema, scaling and pruritus on scalp, periungual desquamation and telogen effluvium (figure 1). Periungual desquamation was the most common nail finding of JIA in our patient group. The less frequent nail findings in the study group included distal onycholysis, pitting, leukonychia, Beau's lines, subungual hyperkeratosis, and splinter hemorrhages over the distal third of the nail plate (figure 1). The most common skin finding in patients with psoriatic JIA was flexural psoriatic lesions and the less common findings were periorbital hyperpigmentation and dactylitis. The most common nail findings in patients with psoriatic JIA were periungual desquamation and distal onycholysis, while the less common finding was splinter hemorrhages. The most common scalp findings in patients with psoriatic JIA were erythema, scaling and pruritus, while telogen effluvium was a less common finding. The most common skin finding in patients with oligoarthritis was photosensitivity, nail finding was periungual erythema, and scalp findings were erythema, scaling and pruritus. We saw transient urticarial rash over the trunk, legs and arms, dermatographism, nail pitting and telogen effluvium in one patient with systemic arthritis; and photosensitivity, livedo reticularis, periungual erythema and telogen effluvium in another patient with RF-negative polyarthritis.

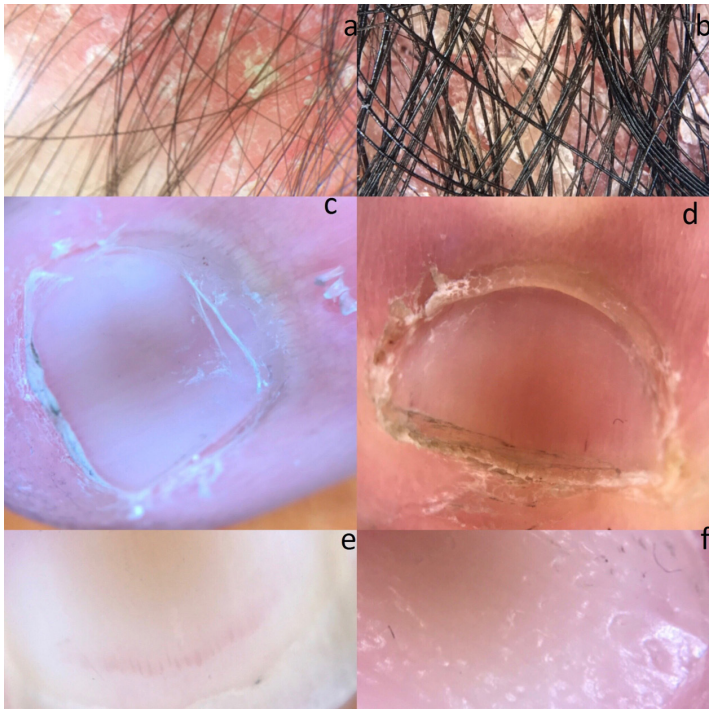

Figure 1. a&b; erythema and coarse squam on scalp, c&d; periungual desquamation in patients with JIA, e; distal splinter hemorrhages on onychodermal band and distal onycholysis, f; pitting in patients with JIA

Four patients with SLE were female and 3 patients were male. The mean age of the patients was 12.8 ±3. All but one of 7 patients showed photosensitivity, malar rash, nailfold capillary abnormalities and non-scarring alopecia clinically. Five patients had non-specific vascular lesions like Raynaud's phenomenon, livedo reticularis and palmar erythema (figure 2).

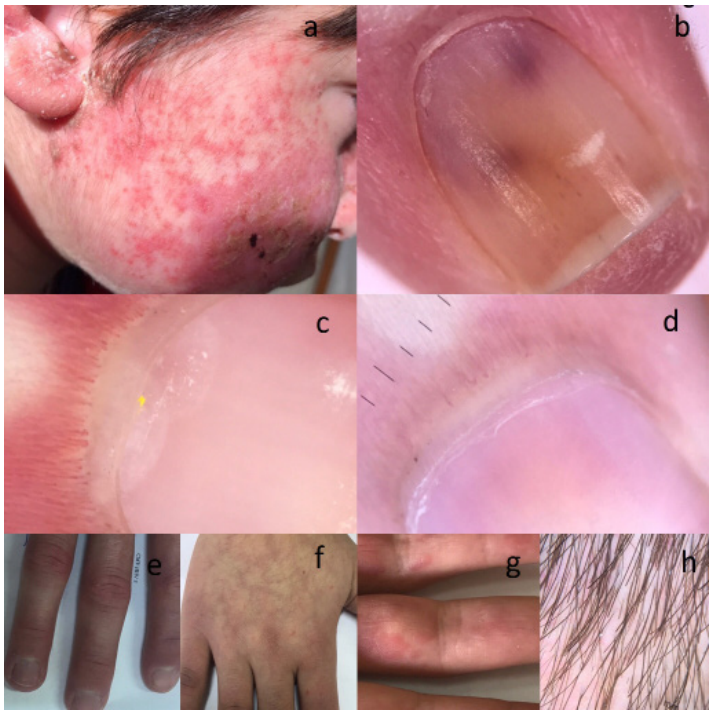

Figure 2. a&e; acute erythematous rash with fine scaling on face and hand, b; periungual erythema, blood spots and splinter hemorrhages on nails, c; dilated and tortious capillaries in nailfold, d; elongating and tortious capillaries alternating with loss of capillaries in nailfold, f; livedo reticularis on the dorsum of hand, g; palmar erythema, h; telogen effluvium with erythematous background in patients with SLE

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Table 1. Clinical characteristics such as the age, sex, duration of disease, prior systemic drug use and the location and type of lesions on skin, nails and scalp in patients with JIA.

JIA disease categories	Age	Gender	Duration (years)	Systemic medication	Skin findings	Nail findings	Scalp skin findings
JIA-1 Psoriatic	9	M	1	Mtx	Periorbital hyperpigmentation Kserosis Dactylitis	Distal onycholysis Periungual desquamation Nail Pitting Punctate leukonychia	Erythema, scaling and pruritus Telogen effluvium
JIA-2 Psoriatic	10	M	3	Mtx	Periorbital hyperpigmentation Kserosis Plaque psoriasis (flexural areas, dorsum of hand) Dactylitis	Periungual desquamation Splinter hemorrhages	Erythema, scaling and pruritus
JIA-3 Psoriatic	10	F	1	C/S	Plaque psoriasis (elbows and knees)	Distal onycholysis Subungual Hyperkeratosis Punctate leukonychia	Erythema, scaling and pruritus Telogen effluvium
JRA-4 Psoriatic	12	M	1	NSAİDs	Periorbital hyperpigmentation Plaque psoriasis (flexural areas)	Periungual desquamation Distal onycholysis Splinter hemorrhages Subungual hyperkeratosis	Erythema, scaling and pruritus, Telogen effluvium
JIA-5 Psoriatic	15	F	4	Mtx+NSAİDs	Plaque psoriasis (flexural areas and face)	Nail Pitting Punctate leukonychia	Telogen effluvium
JIA-6 Psoriatic	16	F	3	Sulfasalazine+Mtx	Sulfasalazine+Mtx	Nail Pitting Distal onycholysis Splinter hemorrhages Beau's lines	Erythema, scaling and pruritus Telogen effluvium
JIA-7 Psoriatic	3	F	1	NSAİDs+C/S	Dactylitis Plaque psoriasis (flexural areas)	Periungual desquamation	Erythema, scaling and pruritus
JIA-1 Oligoarthritis	13	F	1	NSAİDs	Vitiligo	None	None
JIA-2 Oligoarthritis	14	M	3	Sulfasalazine+Mtx	Photosensitivity Livedo reticularis Oral apthosis Raynaud's phenomenon	Periungual erythema Distal onycholysis	Erythema, scaling and pruritus
JIA-3 Oligoarthritis	13	M	1	Mtx	Periorbital hyperpigmentation Plaque psoriasis (flexural areas)	None Punctate leukonychia	Erythema, scaling and pruritus
JIA-4 Oligoarthritis	15	F	1	Mtx+NSAİDs	Photosensitivity Oral apthosis kserosis	Periungual desquamation Nail Pitting Distal onycholysis	None
JIA-5 Oligoarthritis	11	F	1	Sulfasalazine+C/S	Upper eyelid dermatitis	Periungual erythema	Erythema, scaling and pruritus
JIA-6 Oligoarthritis	6	M	1	Mtx+NSAİDs	None	Periungual erythema	Telogen effluvium
JIA-1 Undifferentiated arthritis	10	M	1	NSAİDs	Periorbital hyperpigmentation, Dennie-Morgan lines	Nail Pitting	None
JIA-2 Undifferentiated arthritis	10	F	2	Mtx	None	Periungual desquamation Punctate leukonychia	Erythema, scaling and pruritus Telogen effluvium
JIA-1 Enthesitis-related	15	F	1	C/S+Mtx	Photosensitivity Livedo reticularis	Splinter hemorrage Periungual desquamation	Erythema, scaling and pruritus
JIA-2 Enthesitis-related	13	M	2	NSAİDs	None	Periungual desquamation	Erythema, scaling and pruritus
JIA Polyarticular RF negative	15	M	1	Mtx+NSAİDs	Photosensitivity Livedo reticularis	Periungual erythema	Telogen effluvium
JIA Systemic arthritis	10	M	1	NSAİDs+C/S	Transient urticarial rash over the trunk legs and arms dermatographism	Nail Pitting	Telogen effluvium

JIA: Juvenile idiopathic arthritis, M: Male, F: Female, Mtx: Methotrexate, NSAİDs: non-steroidal anti-inflammatory drugs, C/S: Corticosteroids

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Table 2. Clinical characteristics such as the age, sex, duration of disease, prior systemic drug use and the location and type of lesions on skin, nails and scalp in patients with SLE, DM, SSc and FMF

SLE	Age	Gender	Duration	Systemic medication	Skin findings	Nail findings	Scalp skin findings
SLE-1	16	F	2	C/S	Photosensitivity, Malar rash Raynaud's phenomenon, Livedo reticularis, Chillblain lupus	Subtle nailfold abnormalities, Periungual erythema	Non-scarring alopecia
SLE-2	8	M	7	C/S	Photosensitivity Malar rash, Palmar erythema Periungual telangiectasia	Subtle nailfold abnormalities, Splinter hemorrhage Blood spots	Non-scarring alopecia, Erythema, scaling and pruritus
SLE-3	14	F	2	C/S	Photosensitivity Red poikiloderma, Malar rash Raynaud's phenomenon, Livedo reticularis, Edematous hyperemic rash with scaling on trunk, arms and dorsal aspects of hands sparing knuckles	Subtle nailfold abnormalities	Non-scarring alopecia
SLE-4	14	F	6	C/S	Photosensitivity, Malar rash Raynaud's phenomenon	Distinct nailfold abnormalities	Non-scarring alopecia
SLE-5	16	F	5	C/S	Photosensitivity, Malar rash Raynaud's phenomenon Livedo reticularis	Subtle nailfold abnormalities	Non-scarring alopecia
SLE-6	12	M	2	C/S	Raynaud's phenomenon Livedo reticularis, Malar rash Periungual telangiectasia	Subtle nailfold abnormalities	None
SLE-7	10	M	2	C/S	Photosensitivity Malar rash, Raynaud's phenomenon	Distinct nailfold abnormalities	Non-scarring alopecia
DM							
DM-1	12	F	6	Mtx	Photosensitivity, Violet poikiloderma, Gottron's papules	Nail fold erythema, Distinct nailfold abnormalities, Cuticular dystrophy	Violaceous erythema, scaling and pruritus
DM-2	16	F	2	Mtx + C/S	Photosensitivity Violet poikiloderma, Upper eyelid eczema Heliotrope rash, Gottron's papules	Distinct nailfold abnormalities, Nail fold erythema, Cuticular dystrophy	Violaceous erythema, scaling and pruritus
DM-3	14	F	7	Mtx	Photosensitivity Violet poikiloderma, Calsinosis cutis Gottron's papules, Gottron's sign	Distinct nailfold abnormalities, Nail fold erythema, Cuticular dystrophy	Violaceous erythema, scaling and pruritus
DM-4	8	M	1	Mtx	Photosensitivity Violet poikiloderma, Gottron's papules	Nail fold erythema, Distinct nailfold abnormalities	None
DM-5	11	M	2	Mtx	Violet poikiloderma, Gottron's papules Gottron's sign	Nail fold erythema, Distinct nailfold abnormalities	Erythema, scaling and pruritus
SSc							
Diffuse SSc-1	9	F	2	Mtx+C/S	Raynaud's phenomenon, Pitting edema of the digits Taut and shiny skin Loss of substance from finger pads Pruritus	Distinct nailfold abnormalities, Total leukonychia, Periungual erythema	None
Diffuse SSc-2	16	F	4	Mtx+C/S	Raynaud's phenomenon, Erythema nodosum Taut and shiny skin, Salt and pepper sign, Digital pitted scar loss of substance from finger pads, Pruritus	Distinct nailfold abnormalities, Total leukonychia, Periungual erythema	Erythema, scaling and ruritus elogen effluvium
Diffuse SSc-3	14	F	4	Mtx+C/S	Raynaud's phenomenon, Taut and shiny skin, Salt and pepper sign, Telangiectasia, Loss of substance from, Finger pads Pruritus	Distinct nailfold abnormalities, Leukonychia totalis, Periungual erythema	None
Limited SSc (CREST 1)	13	F	1	Mtx+C/S	Raynaud's phenomenon, Calcinosis cutis Sclerodactyly, Telangiectasia on face and hands	Distinct nailfold abnormalities	None
Limited SSc (CREST 2)	7	F	1	C/S	Raynaud's phenomenon, Calcinosis cutis Sclerodactyly, Telangiectasia on face and hands	Distinct nailfold abnormalities, Distal onycholysis	None
Limited SSc (CREST 3)	12	F	1	C/S	Raynaud's phenomenon Calcinosis cutis, Sclerodactyly, Telangiectasia on face and hands	Distinct nailfold abnormalities	Non-scarring alopecia
FMF							
FMF-1	8	M	4	colchicine	Kserosis, Periorbital hyperpigmentation	Distal onycholysis, Leukonychia punctata	None
FMF-2	14	F	10	colchicine	Upper eyelid eczema	Longidutinal white lines, Leukonychia punctata	Anagen effluvium
FMF-3	7	F	1	colchicine	Kserosis	Pitting, Distal onycholysis, Leukonychia punctata	Anagen effluvium
FMF-4	7	M	4	colchicine	Kserosis, Pitriasis alba, Periorbital hyperpigmentation	Distal onycholysis,, Leukonychia punctata	None
FMF-5	12	F	2	colchicine	Kserosis	Pitting, Leukonychia punctata	Anagen effluvium
FMF-6	11	F	1	colchicine	Kserosis Periorbital hyperpigmentation	Leukonychia punctata	Anagen effluvium
FMF-7	7	F	1	colchicine	Kserosis Periorbital hyperpigmentation	Leukonychia punctata	Telogen effluvium
FMF-8	8	M	2	colchicine	Kserosis	Pitting,	Telogen effluvium

SLE: Systemic Lupus Erythematosus, DM: Dermatomyo¬sitis, SSc: Scleroderma, FMF: Familial Mediterranean Fever, M: Male, F: Female, Mtx: Methotrexate, C/S: Corticosteroids

Three patients with DM were female and the remaining 2 were male patients. The mean age of this group was 12.2±3. All five patients displayed photosensitivity, violet poikiloderma, Gottron's papules, distinct nailfold abnormalities and nail fold erythema (figure 3).

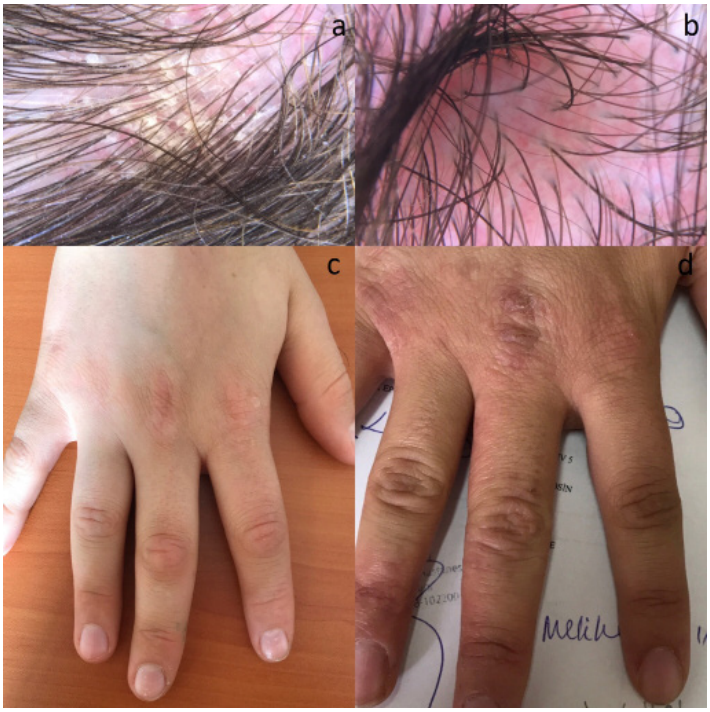

Figure 3 a&b; violaceous erythema and coarse squam on scalp, c&d; Gottron's papules in patients with DM

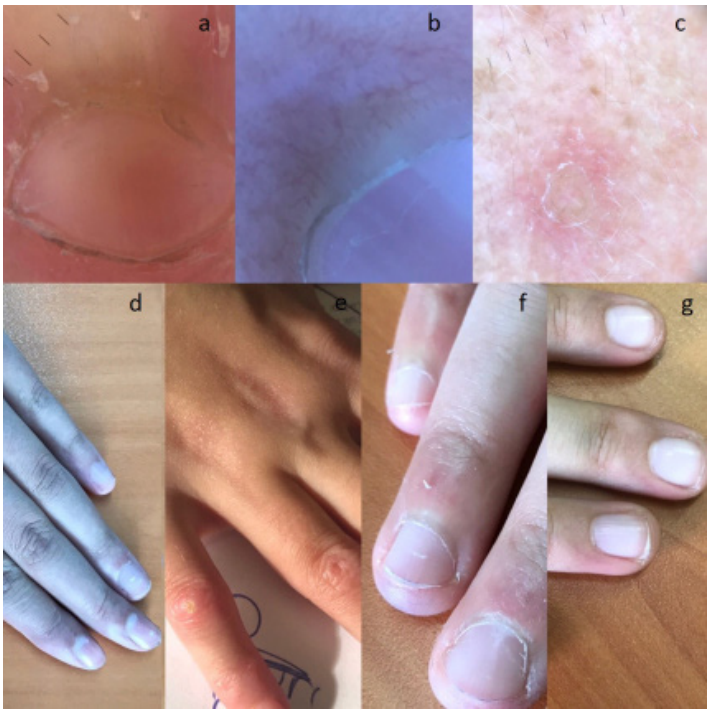

Figure 4. a; loss of capillaries on nailfold b; arborizing vessels on nailfold, c; salt and pepper sign on scalp, d; loss of substance from finger pads, e; digital pitted scar on the 5th finger, f&g; leukonychia totalis, punctata and periungual erythema in patients with SSc.

Five patients with SSc were female and 1 patient was male. The mean age of the patients was 11.8±3.3. Three patients with limited SSc had Raynaud's phenomenon, calcinosis cutis, sclerodactyly, telangiectasia on face and hands and distinct nailfold abnormalities while all three patients with diffuse SSc displayed Raynaud's phenomenon, taut and shiny skin, loss of substance from finger pads, total leukonychia, periungual erythema and pruritus (figure 4) . Telangiectasias involving the face, lips and palms were more common in patients with limited SSc, but were also noticed in patients with diffuse disease. We saw capillary loss alternating with dilated loops in nailfold and arborising vessels in distal finger with the dermatoscope. Distinct nailfold changes like prominent large, tortuous capillaries and areas of marked avascularity were as well seen in patients with DM in our study (figure 4).

Five patients with FMF were female while the remaining 3 were male patients. The mean age of this group was 9.25±2.7. Almost all the patients with FMF had signs of atopy; kserosis, pitriasis alba, periorbital hyperpigmentation, leukonychia punctata and pitting. Four of the patients with FMF suffered from anagen effluvium.

Discussion

The skin, joints and musculoskeletal system are often involved concomittantly in rheumatologic diseases [1]. Rheumatologic diseases are the second most common autoimmune diseases in childhood after diabetes mellitus [2] and there is scarce information in the literature on skin findings in this population [1-8]. In this study, we aimed to evaluate the clinical and dermatoscopic skin, nail and scalp findings of common rheumatologic diseases in the pediatric population.

The biggest number of patients referred to our clinic was JIA while the smallest groups were SLE and DM. This may be due to the fact that SLE or DM are diseases with an occasional later onset in adolescence or adulthood while JIA starts in childhood [2,3]. The skin, nail and scalp findings in common pediatric rheumatologic diseases are listed into subheadings and discussed below.

Juvenile Idiopathic Arthritis

Juvenile idiopathic arthritis is a disease of unknown cause characterized by the presence of a chronic arthritis persisting for more than 6 weeks in children or adolescents under 16 years of age [9]. JIA is the most common rheumatologic disease encountered in the pediatric population that is almost exclusively seen in this age group, so it is important to recognize the skin findings of JIA, which are not known well, in this particular patient population [2,3].

Seven disease categories have been suggested for JIA by the Pediatric Task Force of the International League of Associations for Rheumatology. These categories are systemic arthritis (adolescent-onset Still's disease), RF-negative polyarthritis, RF-positive polyarthritis, psoriatic arthritis, oligoarthritis (persistent or extended), enthesitis-related arthritis and undifferentiated arthritis [9].

In our study, plaque psoriasis was seen in 36.8% of JIA patients. Psoriatic JIA may occur in concurrence with psoriasis. On the other hand, without the presence of any skin lesions, it is defined as the arthritis with any two of the following: psoriasis in a first-degree relative, dactylitis, and nail pitting or onycholysis. Plaque psoriasis, the most common type of psoriasis in JIA [6], was the most frequent type in our study as well.

Psoriatic lesions in JIA are often smaller, thinner and less scaling than adult psoriasis and tend to develop more often on the face and flexural areas [10] similar to the patients in this study. The scalp is also frequently involved and is often the first site of presentation in children [6] as was the case in our study group. In this study, the most common findings in JIA patients were erythema, scaling and pruritus on scalp (63.1%, p=0.0001), telogen effluvium (47.3%, p=0.003) and periungual desquamation (42.1%, p=0.007). In our opinion, the presence of periungual desquamation and telogen effluvium were associated with the adverse effects of systemic administered drugs, especially methotrexate [6]. Periungual desquamation was the most common nail finding of JIA in our patients that contrasts with the literature where distal onycholysis has been reported as the most frequent finding. The less frequent nail findings in the study group included, distal onycholysis (31.5%, p=0.03), pitting (31.5%, p=0.03), periungual erythema (21%, p=0.14), splinter hemorrhages (21%, p=0.14), and dactylitis (15.7%, p=0.2) which are consistent with the findings in the literature [11,12], [fig 1]. On the other hand, punctate leukonychia (26.3%, p=0.4) and periorbital hyperpigmentation (26.3%, p=0.2) were also common in JIA while they were not statistically significant. In our opinion, the presence of these signs was associated with concomitant atopy.

Systemic Lupus Erythematosus

The cutaneous lupus erythematosus (LE) is classified into two groups as specific and non-specific. Within the category of specific skin lesions, it is divided into acute, subacute and chronic subgroups [13]. Acute specific cutaneous lesions that have been associated with SLE [14] were also observed in our study group.

In our study, one of the patients had edematous hyperemic rash with scaling on trunk, arms and dorsal aspects of hands sparing the knuckles. In lupus, the face is most commonly affected; but often times, lesions may be more widespread in distribution sparing the knuckles [14] as seen in one of our patients.

In the current study, six of seven patients with SLE exhibited photosensitivity, malar rash, subtle nailfold capillary abnormalities and non-scarring alopecia clinically. Nonspecific vascular lesions like Raynaud's phenomenon, livedo reticularis, palmar erythema and periungual telangiectasia are common in patients with LE, particularly in those who have SLE. Other non spesific lesions that can be seen in LE are purpurae, urticarial papules or ulcerations, livedo reticularis, thromboses, ulcerations, lesions resembling Degos' disease, sclerodactyly, calcinosis, rheumatoid nodules, papulonodular mucinosis and anetoderma [14]. The pathogenesis of these skin changes in SLE are mostly vascular such as nail-fold abnormalities (large and tortuous capillaries together with areas of avascularity), more severe complications such as vasculitis (urticarial vasculitis, leukocytoclastic vasculitis and nodular vasculitis) and other forms of vasculopathy (livedo reticularis, atrophie blanche, Degos' disease, ulcerations, and thromboses) [14,15]. In our study, five of the patients displayed common non-spesific vascular lesions like Raynaud's phenomenon, livedo reticularis, palmar erythema and periungual telangiectasia [figure 2].

Dermatomyositis

Common skin findings of DM like photosensitivity, violet poikiloderma, Gottron's papules, distinct nailfold abnormalities, nail fold erythema and cuticular dystrophy were seen in five of six patients with DM in our study [figure 3].

In our study, poikiloderma in DM was characterized by a violet color, whereas it was red-pink in patients with SLE and it was distributed around the eyes and extensor surfaces, including the knuckles. Gottron's papules (lichenoid lesions on knuckles) and Gottron's sign (violaceous discoloration of the knuckles, elbows and/or knees) were located on knuckles and elbows in our patients.

One of our female patients with DM had calcinosis cutis on her legs in the deep fascia and intramuscular connective tissue which was thought to be secondary to recurrent trauma to her legs and this was convenient with the fact that it occurs most often at sites of friction and trauma (15). Calcinosis cutis consists of painful, hard, irregular nodules that eventually drain chalky material to the skin surface, and it is known to be more prevalent in juvenile dermatomyositis, affecting between 25% and 70% of patients [15-17].

The scalp and cutaneous lesions of DM were pruritic and had a violet background with arborizing vessels with dermatoscopy in our patients while the lesions were not pruritic in patients with SLE. According to the literature, this pruritus can significantly affect the patients' quality of life and may help in distinguishing DM from SLE [14,18,19] .

Alopecia appears to be a significant sign in the course of SLE. The involvement of the scalp is also a common finding in DM, and is often characterized by a diffuse, violaceous, scaly, non-scarring and symptomatic hair loss [7] that was observed as violaceous erythema, scaling and pruritus in our study group.

Histopathologic examination of a heliotrope, violet erythema reveals an interface dermatitis. More inflammatory lesions (i.e., Gottron papules) show lichenoid infiltrate and acanthosis of the epidermis as well. This may explain the violet colour of lesions on the skin and scalp.

Scleroderma

SSc has two major clinical subtypes: Limited and diffuse. Limited SSc is characterized by fibrotic skin changes that are limited to the fingers, hands and face, and includes the CREST syndrome. In diffuse SSc, generalized fibrotic skin changes spread to involve the forearms, arms, trunk, face and lower extremities. CREST syndrome describes the clinical features in a subset of patients with limited SSc: calcinosis, Raynaud's phenomenon, esophageal involvement, sclerodactyly and telangiectasia [14]. In our study we evaluated the skin findings of 3 patients with CREST syndrome and 3 patients with diffuse SSc. All patients with limited SSc displayed Raynaud's phenomenon, calcinosis cutis, sclerodactyly, telangiectasia on face and hands, and distinct nailfold abnormalities; while all patients with diffuse SSc had Raynaud's phenomenon, taut and shiny skin, loss of substance from finger pads and pruritus. In our study, late atrophic phase with a beaked nose and microstomia were not observed in the pediatric population. The term sclerosis describes induration as a result of excessive deposition of collagen and subsequent tissue fibrosis. When cutaneous involvement proceeds, an edematous phase of the affected sites often takes place, especially on the fingers. Similar changes may be seen on the forearms, legs, feet, face and

trunk as well [15]. This edema results in taut and shiny appearance of our patients. This is followed by gradual thickening of the skin in which the initial inflammation is replaced by interstitial fibrosis caused by abnormal collagen metabolism and subsequent sclerodactyly. The impaired acral blood flow in sclerodactyly may lead to digital pits and ulcers as seen in our patients.

A rare skin sign in patients with SSc is hyperpigmentation. Variants of skin discoloration in patients with SSc include "salt and pepper sign" which describes localized areas of depigmentation with sparing of the perifollicular skin. The leukoderma of scleroderma is characterized by salt and pepper sign and arborising vessels were as well seen by dermatoscope in the scalp skin of one of our patients with diffuse SSc and this was parallel to the findings reported in the literature [15,20].

Telangiectasias involving the face, lips and palms are more common in patients with limited SSc but may also be seen in patients with diffuse disease, similar to our patients. Capillary abnormalities in the proximal nail fold have been reported in more than 90% of SSc patients [14,21]. We noted capillary loss alternating with dilated loops in nailfold and arborising vessels in distal finger with the dermatoscope. Distinct nailfold changes like prominent large, tortuous capillaries and areas of marked avascularity were also seen in patients with DM in our study. Parallel to the previous reports [13], our patients with SLE had more subtle nailfold capillary abnormalities than patients with scleroderma or dermatomyo¬sitis.

Familial Mediterranean Fever

FMF is a disease characterized by recurrent attacks of fever accompanied by peritonitis, pleuritis and arthritis. Skin lesions of FMF are known as erysipelas, such like the erythema that occur mainly on the legs, between the ankle and the knee, or on the dorsum of the foot [22,23]. Vasculitides are rare, approximately 5% of individuals with FMF have been reported to have Henoch-Schonlein purpura and about 1% to have polyarteritis nodosa [24,25]. In addition to these, recurrent urticaria and atypical hyperemic recurrent skin lesions had been reported as rare manifestations of FMF [26,27].

In contrast with the reports in the literature, almost all the patients with FMF displayed signs of atopy; kserosis, pityriasis alba, periorbital hyperpigmentation, leukonychia punctata and pitting in our study.

Common hair disorders in autoimmune connective tissue diseases are known to be telogen hair loss, diffuse thinning or fragility of hair and scarring alopecia. In addition to this, some drugs used to treat autoimmune connective tissue diseases may cause hair loss like methotrexate and colchicine, as observed in our study. Four of eight patients with FMF suffered from anagen effluvium that was thought to be the adverse effect of colchicine [28-31].

To sum up, when we see a child with arthralgia accompanied with pruritus on scalp with fine scales and periungual desquamation, one may consider JIA as the diagnosis; and when arthralgia is accompanied by signs of atopy like kserosis, pityriasis alba, periorbital hyperpigmentation, leukonychia punctata and pitting, FMF should be considered in the differential diagnosis. Vascular skin lesions like Raynaud's phenomenon, livedo reticularis, palmar erythema, periungual telangiectasia and nailfold abnormalities viewed by dermatoscopy are common in childhood rheumatologic diseases.

This study has several limitations. The study population included a relatively small sample of rheumatologic diseases. Further clinical trials with larger patient populations where comparison of pediatric findings with adult rheumatologic diseases and detailed evaluation of nail findings with dermatoscopy and comparison of these findings with capillaroscopy are carried out are needed to gather more conclusive data on the dermatological findings in pediatric rheumatological diseases.

Conclusion

Specific skin lesions can be the peculiar features of common rheumatologic diseases like JIA, SLE, DM, SSc and FMF in the pediatric population. Since it is not always easy to perform biopsy in children to confirm skin involvement of a rheumatologic disease, clinical and dermatoscopic skin findings can help both dermatologists and rheumatologists in diagnosis.

Competing interests
The authors declare that they have no competing interest.

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Ethical approval
Consent of ethics was approved by the local ethics committee.

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