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ORIGINAL ARTICLE PEDIATRICS

Towards a combined pediatric rheumatology-dermatology clinic: One-year experience

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ABSTRACT

OBJECTIVE: Dermatological findings may be the sole complaints of diseases in pediatric rheumatology practice. Evaluating patients with a multi-disciplinary approach may facilitate access to an accurate diagnosis. Herein, we reported our one-year experience in collaborative pediatric rheumatology-dermatology.

METHODS: Patients were initially evaluated separately in pediatric rheumatology-dermatology outpatient clinics. Subsequently, once a week, the final diagnoses of patients with suspected skin rash were collaboratively discussed by two pediatric rheumatologists and a dermatologist.

RESULTS: A hundred and one patients were included in this study. Of these 101 patients, 65 attended to dermatology outpatient clinic initially, while the remaining 36 applied to the pediatric rheumatology outpatient clinic. The most common mucocutaneous finding was squamous lesions in 30 patients, followed by erythematous lesions in 28 and mucosal ulcers in 14. Finally, 69 patients were diagnosed with a rheumatic disease while 32 had differential diagnoses apart from rheumatic diseases.

CONCLUSION: Patients with rheumatologic diseases frequently present with only mucocutaneous findings. Thus, a detailed examination of the mucosa, skin and its attachments is of paramount importance in rheumatology practice. We suggest that a close interaction between pediatric rheumatology-dermatology and the formation of consensus clinics are going to assist clinicians in making easier and accurate diagnoses.

Keywords: Combined clinic; dermatologic finding; rheumatologic disease.

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The skin has several roles in the body and acts as an immunologic organ. Rheumatologic diseases may present with multi-systemic involvement that requires collaboration among rheumatologists and many other disciplines. Dermatologic complaints may be the initial sign of rheumatologic diseases and examination of the skin and its attachments have special importance in rheumatology practice [1, 2]. In recent years, the importance of collaborative care has been increasingly perceived. Multidisciplinary evaluation of patients with a rheumatic disease provides not only an accurate diagnosis and prompt treatment but also better patient satisfaction. In the last decade, combined rheumatology-dermatology clinics have emerged as a new approach in the management of adult patients with psoriasis [3, 4]. However, experience regarding combined clinics in pediatric rheumatology is insufficient. Pediatric rheumatologists and dermatologists need to cooperate in combined clinics, especially when

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they deal with patients who have symptoms and signs involving skin and musculoskeletal system. At our center, we intended to provide a more holistic approach for children with rheumatologic disease, and for this purpose, during the last year, our patients with skin and musculoskeletal findings were collaboratively evaluated by two pediatric rheumatologists and a dermatologist. Herein, we aim to share our one-year experience of a combined pediatric rheumatology-dermatology clinic that may serve as a model while designing a combined clinic.

MATERIALS AND METHODS

Patients were initially examined separately in pediatric rheumatology-dermatology outpatient clinics. Patients who had attended the dermatology outpatient clinic with a rash suggesting a rheumatologic disease with additional symptoms, such as periodic fever, recurrent oral aphthous, purpuric rashes on lower extremities, were referred to the pediatric rheumatology clinic. Furthermore, all patients who had attended to the pediatric rheumatology outpatient clinic with rash that could not be attributed to a final rheumatologic diagnosis were consulted the same dermatologist. Subsequently, once a week, these patients were recalled and final diagnoses were jointly discussed by two pediatric rheumatologists (NAA and HES) and a dermatologist (ZT). Disagreements were resolved by consensus. Figure 1 shows a diagram of the assessment of the patients. Skin biopsy was performed in cases when the diagnosis could not be made by clinical and laboratory findings.

The institutional review board approved this study; reference number KAEK/2019.01.02, dated 01/02/2019.

Statistical Analysis

Data were entered into an Excel file and imported into IBM SPSS (Version 22 for Windows, Armonk, New York, 2015) for statistical analysis. The variables were investigated using visual (histogram, probability plots) and analytic methods (Kolmogorov-Smirnov/Shapiro-Wilk's test) to determine whether or not they were normally distributed. Descriptive analyses were presented using proportions, mean, standard deviation (SD), medians, minimum (min), and maximum (max) values as appropriate.

RESULTS

In this study, 101 patients were evaluated collectively. Sixty-five patients initially applied to dermatology outpatient clinic while 36 patients attended to pediatric rheumatology outpatient clinic. Of 101 patients, 68 were female and the median (min–max) age was 11.5 (1.8–17.5) years.

The most common mucocutaneous findings were as follows: Squamous lesions (n=30), erythematous lesions (n=28) and mucosal ulcers (n=13). Finally, 69 patients were diagnosed with a rheumatic disease while 32 patients had differential diagnoses apart from rheumatic diseases (Table 1).

Thirty patients had papulosquamous lesions, which revealed with psoriasis. All of them were initially examined in a dermatology outpatient clinic. Among them, 12 patients had plaque psoriasis, 12 had guttate psoriasis, five had palmoplantar psoriasis and one had inverse psoriasis. Nine patients described morning stiffness lasting at least 15 minutes. On physical examination, arthritis was detected in 13 patients, enthesitis in five patients and sacroiliac tenderness in one patient. The final diagnoses of three patients were confirmed as psoriasis with a pathological examination of skin biopsies. 13 patients were finally diagnosed with psoriatic arthritis.

Fourteen patients with a complaint of oral ulcers were evaluated and all were initially evaluated in a dermatology outpatient clinic. They were finally diagnosed with Be-

Highlight key points

- Skin examination may provide many clues for differential diagnosis of rheumatological diseases.
- Collaboration between pediatric rheumatologists and dermatologists may facilitates the early diagnosis.
- Our preliminary results may help to develop combined pediatric rheumatology-dermatology clinics.

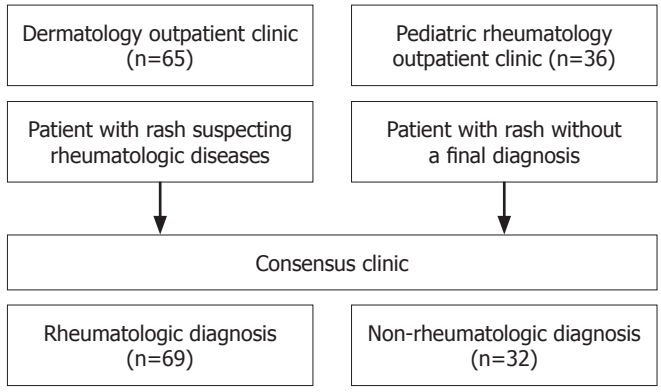

Figure 1. The assessment algorithm of the patients.

Table 1. Demographic and clinical characteristics of the patients

	Psoriasis (n=30)	BH (n=14)	SLE (n=13)	Scleroderma (n=7)	JDM (n=2)	DADA2 (n=3)	Non-rheumatologic conditions (n=32)
Gender (F/M)	20	10	10	6	0	2	20
Age, median (min–max)	10.7 (1.8–16.9)	15.1 (10.2–17.8)	13.6 (8.1–16.7)	9.8 (3.–17.5)	11.1/11.5	15.2 (8.2–15.6)	11.5 (3.1–17.5)
Type of skin lesion	Papulosquamous lesions	Oral aphthous	Malar rash (n=12) and lupus pernio (n=1)	Sclerotic lesion	Gottron's papule	Livedoid rash	Erythematous lesions, hyper-pigmented lesion, xerosis
Arthralgia, n	22	8	4	2	0	3	31
Arthritis, n	13	2	8	1	0	1	7

BH: Behcet's disease; DADA2: Deficiency of adenosine deaminase 2; F: Female; M: Male; max: Maximum; min: Minimum; n: Number of patients; SLE: Systemic lupus erythematosus; JDM: Juvenile dermatomyositis.

hçet's disease (BD). Among them, 57.1% patients had arthralgia, 35.7% patients had genital ulcers, 14.3% patients had abdominal pain and 14.3% patients had arthritis, concomitantly. On physical examination, pseudofolliculitis was observed in eight patients. Furthermore, three patients had erythema nodosum. Pathergy test was positive in three and HLA-B51 was positive in ten of them.

Thirteen patients were diagnosed with systemic lupus erythematosus (SLE). Among them, eleven patients had a malar rash, one patient had lupus tumidus and one patient had chilblain lesions. Of 13 patients, 10 were initially examined in the dermatology outpatient clinic while the remaining three were seen in pediatric rheumatology outpatient clinic. All patients had photosensitivity, four patients had cytopenia, four patients had non-erosive arthritis and one patients had neurologic involvement. Two patients also had discoid lesions on their face and scalp. The patient with chilblain lesions was finally diagnosed with lupus pernio. Anti-nuclear antibody (ANA) was positive in all of them, anti-double-stranded DNA was positive in 10 patients and eight patients had hypocomplementemia.

Seven patients with sclerotic lesions in different parts of the body were diagnosed with localized scleroderma. All applied to the dermatology outpatient clinic initially. The final diagnoses of these seven patients were confirmed as scleroderma by skin biopsy.

Three patients had a livedoid rash. All were initially seen by a dermatologist. Two had abdominal pain and one had recurrent fever attacks. All suffered from arthralgia and one had arthritis. They were finally diagnosed with adenosine deaminase 2 deficiency (DADA2) by genetic analyzes.

Two patients with Gottron's papules were diagnosed with juvenile dermatomyositis (JDM). One was initially examined in dermatology outpatient clinic and the other in pediatric rheumatology outpatient clinic.

Thirty-six patients attended to pediatric rheumatology outpatient clinic. Among them, 28 patients had erythematous lesions. All of them were initially examined in a pediatric rheumatology outpatient clinic because 20 of them were followed up with a familial Mediterranean fever (FMF) and eight patients with juvenile idiopathic arthritis (JIA). The final diagnoses were dermatitis in 22 patients, allergy in four and scabies in two patients. The final diagnoses of the remaining four patients were xerosis in three and atrophoderma in one patient.

DISCUSSION

The detailed examination of the skin and mucosal tissues may yield significant clues for the diagnosis of systemic dis-

eases. Herein, we evaluated 101 patients collaboratively, of whom 69 achieved accurate rheumatologic diagnoses by courtesy of integrated multidisciplinary collaborative care.

A collaborative approach promotes a timely and accurate diagnosis of patients whereas number of combined pediatric rheumatology-dermatology clinics is limited. A study conducted by Samycia et al. [5] reported 320 patients who were evaluated in a rheumatology and dermatology clinic during two years. They demonstrated that the most common rheumatologic diagnosis was SLE (18%), followed by rheumatoid arthritis (15%), psoriatic arthritis (13%), and undifferentiated connective tissue disease (8%). Furthermore, they showed that the most common dermatologic diagnoses were as follows: Dermatitis (17%), psoriasis (11%), cutaneous lupus (7%), alopecia (6%), and infections (5%) [5]. In their study, up to 50% of SLE patients presented with a non-lupus-specific facial rash. Consequently, they stated that skin problems in rheumatologic patients might not always be related to the underlying condition. In our study, the most common rheumatologic diagnosis was psoriasis, followed by BD and SLE. Furthermore, we demonstrated that 28 patients with a rheumatic disease presented nonspecific dermatologic features. Therefore, both rheumatologists and dermatologists should consider differential diagnoses while evaluating their patients.

Wong et al. [6] designed a study to develop a standardized screening tool for rheumatologists to assess skin manifestations. Rheumatologists and rheumatology fellows were trained by a dermatologist, and thereafter, they evaluated 100 patients [6]. They reported that 81% of dermatologic diagnoses made by the rheumatologists matched with the diagnoses that were made by the dermatologist. The most common diagnosis was psoriasis, followed by dermatitis [6]. Finally, they suggested that a standardized integumentary assessment tool might increase the rate of accurate diagnosis by rheumatologists [6]. In our clinic, we assess all new patients with the same questionnaire prepared by the collaboration of rheumatologists and dermatologists that provides a more standardized approach and includes queries about skin, musculoskeletal system, and systemic findings.

In previous studies, it was stated that close interactions between rheumatologists and dermatologists might enhance the correct diagnosis and optimum care of the patients. For instance, Reich et al. [7] evaluated the prevalence of psoriatic arthritis among patients attending a dermatologist and reported a high prevalence of undiagnosed cases of active psoriatic arthritis among patients with psoriasis. Thus, they suggested that encouraging multidisciplinary evaluation and management was required for these patients [7]. In present study, thirty patients were diagnosed with psoriasis by a dermatologist. However, after the consensus clinic, 13 of them were finally diagnosed with psoriatic arthritis. Furthermore, 14 patients with BD, 10 patients with SLE, seven patients with scleroderma, three patients with DADA2 and one patient with JDM attended to dermatology clinic at first. The presence of skin lesions as the sole symptom is exceptional and systemic findings may accompany these aforementioned diseases, so a collaborative care providing a more comprehensive examination hinders additional morbidity among these patients. Furthermore, in pediatric patient care, the dosage of drugs is adjusted according to the weight of the chidren, and the side effect profile may be distinct as well. Therefore, all children receiving systemic therapy should also be under the supervision of a pediatrician. We believe that integrated multidisciplinary collaborative care provides earlier accurate diagnosis and better monitoring and treatment of patients. In this way, patients' satisfaction will also improve. Urruticoechea-Aranaa et al. [8] evaluated patients' satisfaction with psoriasis who were followed up in a combined rheumatology-dermatology clinic and reported that almost all patients were pleased with the combined clinic. Similarly, Campagna et al. [9] demonstrated that three quarters of patients with autoimmune diseases were quite satisfied with this collaborative approach.

Ardoin et al. [10] conducted a survey about research priorities in childhood-onset lupus. Three quarters of the participants reported that collaboration with dermatologists is required while managing childhood-onset lupus. Although clinicians believe in the necessity of collaborative clinics, unfortunately, there are no combined pediatric rheumatology-dermatology clinic data. To our knowledge, this is the first study evaluating children with cutaneous and musculoskeletal findings at a consensus pediatric dermato-rheumatoidoutpatient-clinic. However, our study is limited by its single-center design and small sample size.

In early 2020, two dermatologists reviewed an article in two parts: "The systemic autoinflammatory disorders

for dermatologists". They mentioned that those diseases have some form of skin manifestation as a primary clinical feature, and therefore are important for a dermatologist to bear in mind. In this way, the majority of these syndromes can be effectively controlled and quality of life can be dramatically improved by protecting from the development of irreversible complications such as AA amyloidosis [11, 12].

In conclusion, the presentation of rheumatic diseases with almost all kinds of skin and mucosal features is a well-recognized issue. The criteria put forward for several rheumatic diseases–involve dermatological features as classification items. Thus, a detailed and careful examination of skin and mucosal tissues by rheumatologists and dermatologists collaboratively is essential for accurate diagnosis and differential diagnosis of rheumatologic diseases. Our preliminary results may provide a road map for combined pediatric rheumatology-dermatology clinics in the future.

Ethics Committee Approval: The Faculty of Health Science, Kanuni Sultan Süleyman Training and Research Hospital Clinical Research and Ethics Committee granted approval for this study (KAEK/2019.01.02, date: 01.02.2019).

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