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# Nail-fold Capillaroscopic Changes in Children with Juvenile Dermatomyositis and Specific Autoantibodies

## Jüvenil Dermatomiyozit Hastalarında Kapilleroskopik Değişiklikler ve Spesifik Otoantikorlarla İlişkisi

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## ABSTRACT

**Objective:** Microvascular changes observed during dermoscopy have been widely used for diagnosing and monitoring various connective tissue disorders, including juvenile dermatomyositis (JDM). This study investigated capillaroscopic changes in nail-folds, specifically, nail-fold capillary density (NFCD), in children with JDM.

**Methods:** A prospective study was conducted on children diagnosed with JDM between 2010 and 2021 who were examined via nail-fold capillaroscopy (n=14) during August and December 2021. Demographic and clinical data, myositis-specific autoantibodies for JDM, and capillaroscopic findings were prospectively collected. In addition to children with JDM (group JDM), we randomly selected 20 children with non-specific leg pain as the control group. Capillaroscopic findings were compared between the groups.

**Results:** The groups were similar in terms of age and sex characteristics (p=0.848 and p=0.635). Ten children had myositis-specific autoantibodies (71.4%). The median NFCD was significantly lower in the JDM group than in the controls (p=0.001). Children with JDM had a significantly higher frequency and amount of disorganized, tortuous, crossing, enlarged, and giant capillaries than healthy controls (p<0.05). There were significantly higher values of the neoangiogenesis score and MES in Group JDM than in Controls (p<0.001 and p<0.001). Children with positive autoantibodies had higher NFCD and lower interpapillary distance values (p<0.05).

**Conclusion:** Children with JDM exhibited remarkable morphological changes during nail-fold capillaroscopy. Higher neoangiogenesis scores, higher MES values, and decreased NFCD might play a role in diagnosing and differentiating childhood JDM.

**Keywords:** Juvenile dermatomyositis, dermoscopy, microscopic angioscopy, microvascular density, nails, capillaries

## ÖZ

**Amaç:** Dermoskopi sırasında gözlenen mikrovasküler değişiklikler, juvenil dermatomiyozit (JDM) de dahil olmak üzere çeşitli bağ dokusu bozukluklarının tanı ve takibinde yaygın olarak kullanılmaktadır. Bu çalışma, JDM'li çocuklarda kapilleroskopik tırnak kıvrımı değişikliklerini, daha doğrusu tırnak kıvrımı kılcal yoğunluğunu (NFCD) araştırdı.

**Gereç ve Yöntem:** 2010-2021 yılları arasında JDM tanısı alan ve Ağustos ve Aralık 2021'de tırnak kıvrımı kapilleroskopisi (n=14) ile incelenen çocuklar üzerinde prospektif bir çalışma yapıldı. Demografik ve klinik özellikler, JDM için miyozite özgü otoantikorlar ve kapillaroskopik bulgular ileriye dönük olarak toplandı. JDM'li çocukların (grup JDM) yanı sıra, spesifik olmayan bacak ağrısı olan 20 çocuğu da kontrol grubu olarak rastgele seçtik. Gruplar arasında kapillaroskopik bulgular karşılaştırıldı.

**Bulgular:** Gruplar yaş ve cinsiyet özellikleri açısından benzerdi (p=0,848 ve p=0,635). Miyozite özgü otoantikorları pozitif olan 10 çocuk (%71,4) vardı. Medyan NFCD, Grup JDM'de control grubuna göre anlamlı derecede düşüktü (p=0,001). JDM'li çocuklarda düzensiz, kıvrımlı, çapraz, genişlemiş ve dev kapillerlerin sıklığı ve sayısı sağlıklı kontrollere göre anlamlı derecede yüksekti (p<0.05). Grup JDM'de neoanjiyogenez skoru ve mikroanjiyopati değerlendirme skoru (MES) değerleri, kontrol grubuna göre anlamlı derecede yüksekti (p<0,001 ve p<0,001). Otoantikorları pozitif olan çocuklarda NFCD daha yüksek ve papiller mesafe değerleri daha düşüktü (p<0,05).

**Sonuç:** JDM'li çocuklarda tırnak kıvrımı kapilleroskopisi sırasında dikkat çekici morfolojik değişiklikler görüldü. Daha yüksek neoanjiyogenez skorları, daha yüksek MES değerleri ve azalmış NFCD, çocukluk çağı JDM'nin tanı ve ayrımında rol oynayabilir.

**Anahtar Kelimeler:** Juvenil dermatomiyozit, dermoskopi, mikroskobik anjiyoskopi, mikrovasküler dansite, tırnaklar, kılcal damarlar

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## Introduction

Juvenile dermatomyositis (JDM) is a childhood autoimmune connective tissue disease associated with skin, skeletal system, and internal organ disturbances (1). It is categorized as a subgroup of idiopathic inflammatory myopathies according to the European League Against Rheumatism/American College of Rheumatology classification criteria (2,3). An immune response originating from the capillary endothelium of the endomysium is considered an early event causing skin involvement in JDM (3,4). Vascular skin changes play a diagnostic and prognostic role as they precede the development of subsequent myositis (5).

Dermoscopy has recently been used to observe the morphologic features of the skin using *in vivo* magnification (3,6). Standardized basic dermoscopic parameters of inflammatory, infiltrative, and infectious dermatoses have been described (3,7,8). Previous studies have reported that early microvascular changes in various rheumatic diseases can be assessed using nail-fold capillaroscopy (NFC) (3,6,9,10). In this way, NFC can differentiate the types of connective tissue disorders in patients with primary and secondary Raynaud's phenomenon and mixed connective tissue diseases, including polymyositis and dermatomyositis (2,11,12). Besides, it has been speculated that the findings of NFC are related to disease activity and the levels of myositis-specific and-associated autoantibodies for JDM (1,2,5,12,13).

We aimed to evaluate dermoscopic vascular changes via NFC in patients with JDM and to compare these findings with those of healthy children. We present the following case reports in accordance with the STROBE reporting checklist.

## METHODS

### Study

A prospective study including nail-fold capillaroscopic findings of children with JDM was performed in Erciyes University Faculty of Medicine, Department of Paediatric Rheumatology. This study was approved by the Erciyes University Clinical Research Ethics Committee (decision no: 2021/355, date: 05.05.2021). The analysis was performed in accordance with the principles of the Helsinki Declaration. Written informed consent was obtained from the parents of the children.

### Patients

Between August and December 2021, we performed nail-fold capillaroscopic examinations in 14 children with JDM. The diagnosis of probable or definitive JDM was based on the Bohan and Peter criteria, including disease onset before the age of 18 years and ≥24 months from symptom onset to follow-up (14). A predefined treatment protocol was applied to all children with JDM (1). Children with overlap syndrome, juvenile-onset mixed connective tissue disease, and juvenile polymyositis were excluded.

Blood samples were obtained for the anti-nuclear antibody (ANA), the extractable nuclear antigen antibody titer measurements, and the myositis-specific autoantibodies for JDM were measured at the last follow-up examinations. Anti-transcriptional factor-1-γ/p155/140, anti-NXP-2/anti-MJ, anti-SRP, anti-pl-7, anti-pl-12, anti-pm-Scl-75, Mi-2 alpha, Mi-2 beta, MDA5, SAE1, Ku, pm-Scl-100, Jo-1, SEJ, OJ, Ro-52, and anti-SRP were the autoantibodies in the category of the myositis-specific autoantibodies (14). We tested the presence of autoantibodies using an Immunoblot assay (Myositis Profile Euroline Blot test kit, Euroimmun, Lübeck, Germany) at the Biochemistry Laboratory of Erciyes University, Faculty of Medicine.

### Definitions

The first muscular or dermatological symptom was considered as the disease onset. The disease duration was calculated using the interval between the start and last follow-up examination. The Childhood Myositis Assessment Scale (CMAS) was used to assess proximal muscle strength, function, and endurance (15). The scale includes 14 physical maneuvers, with a maximum score of 52. Assessment of improvement or worsening between two quantitative evaluations of an individual patient is the primary function of the CMAS. A trained physical therapist applied the CMAS to all the children in the study (15).

The disease course was categorized as mono, polyphasic, or chronic (16). Remission within 36 months of diagnosis without relapse was defined as a monophasic course. Relapse of the disease at any time point after the previous remission was considered a polyphasic event. Chronic disease course was defined as persistent evidence of disease 36 months after diagnosis.

### NFC

After resting for at least 20 min at room temperature (20-24 °C), a video microscope (MEDL4N Dino-Lite Pro Capillary Scope, Dino-Lite Europe, NN Almere, the Netherlands) was used to perform capillaroscopic measurements as described previously (1,17). A pediatric rheumatologist with an experience of 3 years on NFC examined all fingers except the thumb.

We also investigated the morphological changes related to capillary dropout, branching and dilatation, areas of hemorrhage, avascular area, neovascular changes with neoangiogenesis score, capillary disorganization, and the number of vessels per millimeter (1,18-20). The measurements of the internal capillary diameter between 25 and 50 μm and ≥50 μm were defined as dilated and giant capillaries (18). We considered criteria for determining morphological changes as described previously (19).

The parameters included the total number of capillaries over the nail-fold width, mean nail-fold capillary density (NFCD) per millimeter, capillary size, and numbers of dilated, giant, branching, and tortuous capillaries. Each patient's microangiopathy evaluation score (MES) was calculated as defined Sulli et al. (21). In this scale, the loss of capillaries, disorganization of the microvascular array, and capillary ramifications were semi-quantitatively evaluated with scores ranging from zero to three. The summation of the scores for each parameter revealed that the MES ranged from zero to nine.

### Groups

The study group included children with JDM (group JDM). We randomly selected 20 children with nonspecific leg pain as the control group (the control).

### Variables

Data on demographic characteristics (age, sex), clinical characteristics (age at diagnosis, CMAS scores at the admission and the last follow-up, disease duration, physical findings, and medications), and laboratory findings were collected and stored.

### Statistical Analysis

For descriptive statistics, mean±standard deviation was used to present continuous data with normal distribution. The median with minimum-maximum values was applied for continuous variables without normal distribution. Numbers and percentages were used as categorical variables. The Shapiro-Wilk, Kolmogorov-Smirnov, and Anderson-Darling tests analyzed the normal distribution of the numerical variables.

The Pearson chi-square and Fisher's exact tests were used to compare differences between categorical variables in 2x2 tables. The Fisher-Freeman Halton test was used for the RxC tables.

The Mann-Whitney U test was used to compare two independent groups in which numerical variables had no normal distribution.

Spearman's correlation coefficients were calculated to analyze the relationships of NFCD and MES with demographic (age) and clinical variables (age at diagnosis, CMAS at diagnosis, disease duration, duration for steroid use, and the length of follow-up).

Jamovi (Version 2.2.5.0) and JASP (Version 0.16.1) were used for statistical analysis. The significance level (p-value) was set at 0.05 in all statistical analyses.

## RESULTS

There were 14 and 20 children in the JDM and control. The mean age of children with JDM was 10.4±4.0 years. There were nine male and five female children in Group JDM. The groups were similar in terms of age and sex characteristics (p=0.848 and p=0.635). ANA positivity was more frequently detected in the JDM group than in the Controls (p<0.001).

The clinical characteristics of children with JDM are presented in Table 1. The median age at diagnosis was 11.5 years (range, 4-16 years). The median CMAS score at admission was 40, ranging from 30 to 50. The medication distribution in the JDM group is detailed in Table 1. The median follow-up time was 3 years (range, 0.1-11 years)

**Table 1.** Clinical characteristics of children with JDM (group JDM, n=14)

| | Value |
|---|---|
| **Age at diagnosis (year)§** | 11.5 (4-16) |
| **CMAS score at diagnosis§** | 40 (30-50) |
| **Disease duration (year)§** | 3 (0.1-11) |
| **Disease course‡** | |
| Monophasic | 10 (71.4) |
| Polyphasic | 3 (21.4) |
| Chronicity‡ | 1 (7.1) |
| **Medications** | |
| **Steroid‡** | 9 (81.8) |
| **Duration of steroid use (month)§** | 12 (9-24) |
| **DMARD‡** | 11 (78.6) |
| Mycophenolic acid | 3 (27.3) |
| Methotrexate | 6 (54.5) |
| Hydroxychloroquine | 2 (18.2) |
| Duration (month)§ | 2 (1-9) |
| **bDMARD‡** | 2 (14.3) |
| Etanercept | 1 (50) |
| Adalimumab | 1 (50) |
| **IVIG‡** | 6 (42.9) |
| **Follow-up (year)** | 3 (1-11) |
| **CMAS score at the last follow-up§** | 52 (50-52) |

‡n (%), §median (minnimum-maximum)
JDM: Juvenile dermatomyositis, CMAS: Childhood Myositis Assessment Scale, DMARD: Disease-modifying anti-rheumatic drugs, bDMARD: Biologic disease-modifying anti-rheumatic drugs, IVIG: Intravenous immunoglobulin

in group JDM. Nevertheless, the median CMAS score increased to 52 during the last follow-up examination.

Table 2 presents the frequencies of the ENA antibody profiles observed in children with JDM. In four children (28.6%), we detected four ENA antibodies: SM/RNP (n=1), SS-B (n=1), and PCNA (n=2). Ten children (71.4%) were negative for ENA antibodies in group JDM.

Although in four children (28.6%), there were negative results for the myositis-specific autoantibodies, TIF-1 (n=3), NXP-2 (n=2), p1-12 (n=1), p1-7 (n=1), pm-75 (n=1), SRP (n=1), and Jo-1 (n=1) were detected in group JDM. Thus, we noticed positive results for myositis-specific autoantibodies in ten children (71.4%) (Table 3). Children with positive and negative myositis-specific autoantibodies had similar demographic and clinical characteristics (p>0.05) (Table 3).

The nail-fold capillaroscopic findings and their comparisons are presented in Table 4 and Figures 1-3. We detected significant differences between the groups (p<0.05). The median NFCD was significantly lower in the JDM group than in the Controls (p=0.001). The widths of the arteries, veins, and apical loop were substantially lower in children with JDM than in the Controls (p=0.001, p=0.001, and p=0.003). We found significantly higher values of the neoangiogenesis score and MES in the JDM group than in the controls (p<0.001 and p<0.001).

There were significant differences in the morphological capillary abnormalities via NFC between the groups (p<0.05) (Table 4). The children with JDM had significantly higher frequency and number of capillaries with disorganized, tortuous, crossing, enlarged, and giant features than the healthy controls (p<0.05).

We also compared the nail-fold capillaroscopic findings between children with and without myositis-specific

**Table 2.** Distribution of the extractable nuclear antigen antibody profile in children with JDM (group JDM, n=14)

| | Value |
|---|---|
| **Extractable nuclear antigen antibodies‡** | |
| Anti-SM/RNP | 1 (7.1) |
| Anti-SS-B (anti KU positive) | 1 (7.1) |
| Anti-PCNA | 2 (14.3) |
| Negative | 10 (71.4) |
| **Myositis-specific autoantibodies‡** | |
| Negative | 4 (28.6) |
| Anti-TIF1 | 3 (21.4) |
| Anti-NXP2 | 2 (14.2) |
| Anti-pl12 | 1 (7.1) |
| Anti-pl7 | 1 (7.1) |
| Anti-pm75 | 1 (7.1) |
| Anti-SRP | 1 (7.1) |
| Anti-histidyl tRNA synthetase (anti-Jo-1) | 1 (7.1) |

‡n (%), JDM: juvenile dermatomyositis, Ds-DNA: Double-stranded DNA, CCP: Cyclic citrullinated peptide, PCNA: Proliferating cell nuclear antigen protein

**Table 3.** Demographic and clinical characteristics of children with and without myositis-specific autoantibodies

| **Children** | | | |
|---|---|---|---|
| | **With positive autoantibodies (n=10)** | **Negative autoantibodies (n=4)** | **p-value** |
| **Age (year)†** | 12.8 (5.0-16.0) | 9.5 (4.0-11.0) | 0.118** |
| **Sex‡** | | | |
| Male | 6 (60.0) | 3 (75.0) | 0.999* |
| Female | 4 (40.0) | 1 (25.0) | |
| **ANA titer‡** | | | |
| Negative | 0 (0.0) | 2 (50.0) | 0.066* |
| Positive | 10 (100.0) | 2 (50.0) | |
| **Age at diagnosis (year)§** | 12.5 (5.0-16.0) | 9.5 (4.0-11.0) | 0.118** |
| **CMAS score at diagnosis§** | 39.5 (30.0-49.0) | 42.0 (38.0-50.0) | 0.135** |
| **Disease duration (year)§** | 3.0 (0.8-11.0) | 3.2 (0.1-6.5) | 0.618** |
| **Physical findings‡** | | | |
| Calcinosis | 1 (10.0) | 2 (50.0) | 0.176* |
| **Skin involvement** | | | |
| Mild | 5 (50.0) | 3 (75.0) | 0.999* |
| Moderate | 2 (20.0) | 0 (0.0) | |
| Severe | 3 (30.0) | 1 (25.0) | |
| **Muscular involvement‡** | | | |
| Mild | 7 (70.0) | 3 (75.0) | 0.999* |
| Moderate | 3 (30.0) | 1 (25.0) | |
| **Disease course‡** | | | |
| Monophasic | 7 (70.0) | 3 (75.0) | 0.999* |
| Polyphasic | 3 (30.0) | 0 (0.0) | 0.505* |
| Chronicity‡ | 0 (0.0) | 1 (25.0) | 0.286* |
| **Medications** | | | |
| **Steroid‡** | 7 (87.5) | 2 (66.7) | 0.491* |
| **Duration of steroid use (month)§** | 12.0 (9.0-24.0) | 12.0 (9.0-12.0) | 0.592** |
| **IVIG‡** | 4 (40.0) | 2 (50.0) | **0.999*** |
| **Follow-up (year)§** | 3.0 (1.0-11.0) | 3.2 (1.0-6.5) | 0.669** |
| **CMAS score at the last follow-up§** | 52.0 (50.0-52.0) | 52.0 (52.0-52.0) | 0.527** |

†: mean standard deviation, ‡: n (%), §: median (min-max)
JDM: Juvenile dermatomyositis, ANA: Anti-nuclear antibody. CMAS: Childhood Myositis Assessment Scale, DMARD: Disease-modifying anti-rheumatic drugs, bDMARD: Biologic disease-modifying anti-rheumatic drugs, IVIG: Intravenous immunoglobulin, CMAS: Childhood Myositis Assessment Scale

autoantibodies. There were significant differences in the capillary density calculations and the length of the intercapillary distances between the groups (p<0.05). Capillary density was significantly higher in children with positive autoantibodies (p=0.021), contrary to the significantly lower values of intercapillary distance (p=0.023). The other findings were similar between the groups (p>0.05) (Table 5).

We found significantly higher values of the neoangiogenesis score and MES in the JDM group than in the controls (p<0.001 and p<0.001). The measurements of NFCD and MES were not significantly correlated with age, age at diagnosis, CMAS at diagnosis, disease duration, duration of steroid use, and length of follow-up (p>0.05) (Table 6).

## DISCUSSION

This study demonstrated significant microvascular changes in the qualitative and semi-quantitative evaluation of nail-fold capillaroscopic findings in children with JDM compared with healthy children. Higher neoangiogenesis scores, MES values, and decreased NFCD were remarkable findings in JDM. Morphological capillary abnormalities were detected more frequently in children with JDM than in healthy controls. The NFCD and intercapillary distance

**Table 4.** Nail-fold capillaroscopy findings of the groups

| | Group JDM (n=14) | Control (n=20) | p-value |
|---|---|---|---|
| **Capillary density (capillary count/mm)§** | 8.0 (5.0-11.0) | 10.0 (8.0-11.0) | **0.001**** |
| **Capillary length (µm)§** | 299.5 (221.0-628.0) | 329.0 (232.0-470.0) | 0.649** |
| **Arterial width (µm)§** | 14.0 (10.0-71.0) | 11.0 (9.0-15.0) | **0.001**** |
| **Venous width (µm)§** | 17.0 (13.0-82.0) | 14.0 (10.0-17.0) | **0.001**** |
| **Apical loop width (µm)§** | 21.5 (14.0-79.0) | 14.5 (12.0-33.0) | **0.003**** |
| **The intercapillary distance (µm)§** | 114.0 (92.0-251.0) | 112.0 (98.0-225.0) | 0.752** |
| **Morphological changes** | | | |
| **Number of overall capillaries (per linear mm)§** | 0.812 (0.500-1.000) | 0.0 (0.0-0.125) | **<0.001**** |
| **Capillary disorganization§** | 0.438 (0.250-0.625) | 0.0 (0.0-0.250) | **<0.001**** |
| **Capillary tortuosity‡** | | | |
| <%50 | 13 (92.9) | 0 (0.0) | **<0.001*** |
| None | 1 (7.1) | 20 (100.0) | |
| **Number of tortuous capillaries (per linear mm)§** | 0.500 (0.0-0.750) | 0.0 (0.0-0.0) | **<0.001**** |
| **Crossing capillaries‡** | | | |
| >%50 | 1 (7.1) | 0 (0.0) | **0.011*** |
| <%50 | 10 (71.4) | 6 (30.0) | |
| None | 3 (21.4) | 14 (70.0) | |
| **Number of capillaries crossing (per linear mm)§** | 0.375 (0.0-0.750) | 0.0 (0.0-0.0) | **<0.001**** |
| **Dilated (enlarged) capillaries‡** | 8 (57.1) | 4 (20.0) | **0.036*** |
| **Number of enlarged capillaries (per linear mm)§** | 0.625 (0.0-0.750) | 0.0 (0.0-0.0) | **<0.001**** |
| **Giant capillaries‡** | 3 (21.4) | 0 (0.0) | 0.061* |
| **Number of giant capillaries (per linear mm)§** | 0.0 (0.0-0.625) | 0.0 (0.0-0.0) | **0.005**** |
| **Avascular areas‡** | 3 (21.4) | 0 (0.0) | 0.061* |
| **Microhemorrhage‡** | 5 (35.7) | 0 (0.0) | **0.007*** |
| **Microhemorrhage area§** | 0.0 (0.0-0.375) | 0.0 (0.0-0.0) | **0.004**** |
| **Pericapillary edema‡** | 2 (14.3) | 0 (0.0) | 0.162* |
| **Neoangiogenesis‡** | 7 (50.0) | 0 (0.0) | **0.001*** |
| **Neoangiogenesis score§** | 0.562 (0.250-0.750) | 0.0 (0.0-0.125) | **<0.001**** |
| **MES§** | 1.875 (1.250-2.300) | 0.0 (0.0-0.375) | **<0.001**** |

‡n (%), §median (min-max)
MES: Microangiopathy evolution score, JDM: Juvenile dermatomyositis

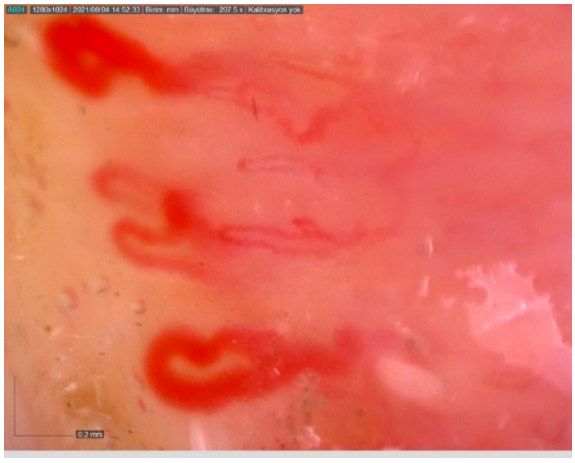

**Figure 1.** Examples of abnormal capillary shapes (morphology); Giant capillaries (homogeneous enlargement of all three limbs, normal shape, and apical diameter ≥50 µm)

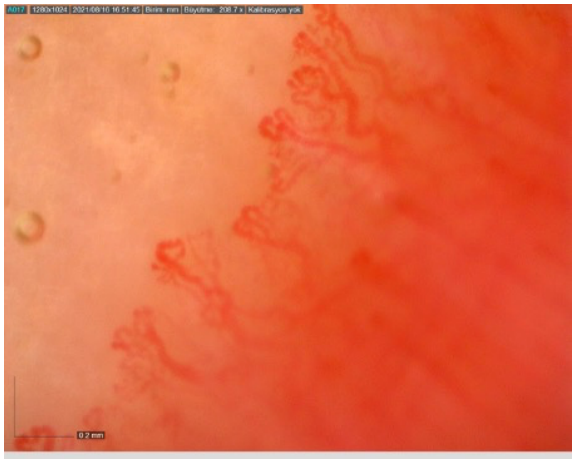

**Figure 2.** Examples of abnormal capillary shapes (morphology); Capillary "ramifications"

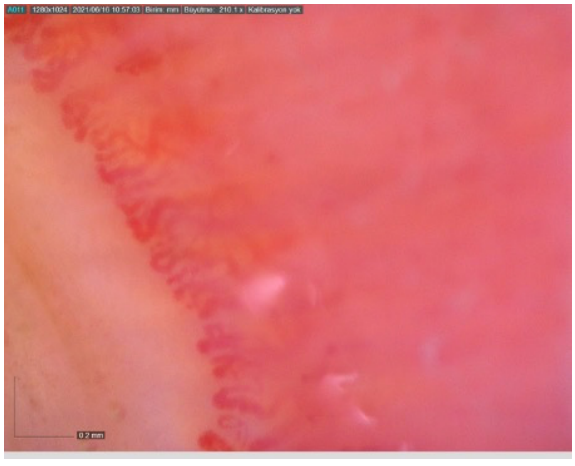

**Figure 3.** Examples of abnormal capillary shapes (morphology); "Meandering" capillaries

values differed significantly between children with and without myositis-specific autoantibodies. Although their clinical presentations were similar, higher NFCD values in children with positive autoantibodies might be explained by possible pathophysiological mechanisms.

Decrements in NFCD, increments in avascular areas of the relevant regions, minor (tortuous, crossed, and enlarged capillaries), and significant capillary morphological changes (mega, meandering, branching, bushy, bizarre, and disorganized polymorphic capillaries) were common findings observed in several chronic inflammatory and connective tissue diseases during childhood and adulthood, such as sclerosis, scleroderma, eczema, psoriasis, dermatomyositis, cutaneous lupus erythematosus, mixed connective tissue diseases, coronavirus (SARS-CoV-2) infection, and chronic hepatitis (2,11,17,19-25). We also detected these microangiopathic changes in children with JDM. Depending on the underlying diseases, various findings were recommended to diagnose or survey their evolution. Although authors are expected to specify each disease's most demonstrative pathological changes, they failed in these comparative studies (2,11,17,21). These microvascular abnormalities may be regarded as evidence of endothelial tissue damage due to microvascular involvement rather than pathognomonic signs. In addition to including several capillaroscopic parameters in this study, we detected significant differences in the capillaroscopic evaluations between the JDM and healthy control groups. Nevertheless, the lack of comparison between JDM and other pediatric rheumatological diseases led to the detection of specific capillaroscopic findings of JDM.

Schmeling et al. (1) investigated the association between NFCD and JDM. They showed that NFCD was a marker for skin and muscle disease activity and a reflection of activity changes depending on the clinical visits of children with JDM. NFCD was also proposed as a dynamic marker of global disease activity of adult dermatomyositis (5). Besides, the early occurrence of nail-fold abnormalities was considered a universal finding in JDM (1,26). This feature may be necessary for the diagnostic efficacy of NFCD in JDM. Barth et al. (18) showed that decreased NFCD and increased neovascularization were significantly associated with higher disease activity and impaired muscle function in adult patients with long-lasting JDM. The same research team reported that NFCD was a predictive marker of lung involvement in long-term JDM (14). It is believed that the grades or extensiveness of nail-fold microangiopathic changes in various rheumatological or connective tissue diseases reflect the severity of the underlying pathologies (22). These changes might be helpful during the onset of

**Table 5.** Nailfold capillaroscopy findings of children with and without myositis-specific autoantibodies

| | Children | | |
|---|---|---|---|
| | **With positive autoantibodies (n=10)** | **Negative autoantibodies (n=4)** | **p-value** |
| **Capillary density (capillary count/mm)§** | 8.5 (5.0-11.0) | 7.0 (6.0-7.0) | **0.021**** |
| **Capillary length (µm)§** | 299.5 (221.0-628.0) | 327.5 (232.0-470.0) | 0.999** |
| **Arterial width (µm)§** | 13.0 (10.0-71.0) | 26.0 (22.0-30.0) | 0.089** |
| **Venous width (µm)§** | 16.0 (13.0-82.0) | 31.5 (28.0-38.0) | 0.088** |
| **Apical loop width (µm)§** | 16.5 (14.0-79.0) | 30.0 (26.0-37.0) | 0.088** |
| **The intercapillary distance (µm)§** | 111.0 (92.0-251.0) | 154.5 (135.0-188.0) | **0.023**** |
| **Morphological changes** | | | |
| **Number of overall capillaries (per linear mm)§** | 0.9 (0.5-1.0) | 0.8 (0.8-1.0) | 0.884** |
| **Capillary disorganization§** | 0.4 (0.2-0.6) | 0.5 (0.4-0.6) | 0.462** |
| **Capillary tortuosity‡** | | | |
| <%50 | 9 (90.0) | 4 (100.0) | 0.999* |
| None | 1 (10.0) | 0 (0.0) | |
| **Number of tortuous capillaries (per linear mm)§** | 0.5 (0.0-0.8) | 0.6 (0.5-0.6) | 0.271** |
| **Crossing capillaries‡** | | | |
| >%50 | 1 (10.0) | 0 (0.0) | 0.999* |
| <%50 | 7 (70.0) | 3 (75.0) | |
| None | 2 (20.0) | 1 (25.0) | |
| **Number of capillaries crossing (per linear mm)§** | 0.4 (0.0-0.6) | 0.4 (0.4-0.8) | 0.183** |
| **Dilated (enlarged) capillaries‡** | 5 (50.0) | 3 (75.0) | 0.580* |
| **Number of enlarged capillaries (per linear mm)§** | 0.6 (0.0-0.8) | 0.6 (0.5-0.8) | 0.770** |
| **Giant capillaries‡** | 1 (10.0) | 2 (50.0) | 0.176* |
| **Number of giant capillaries (per linear mm)§** | 0.0 (0.0-0.6) | 0.1 (0.0-0.2) | 0.620** |
| **Avascular areas‡** | 1 (10.0) | 2 (50.0) | 0.176* |
| **Microhemorrhage‡** | 3 (30.0) | 2 (50.0) | 0.580* |
| **Microhemorrhage area§** | 0.0 (0.0-0.2) | 0.1 (0.0-0.4) | 0.320** |
| **Pericapillary edema‡** | 1 (10.0) | 1 (25.0) | 0.505* |
| **Neoangiogenesis‡** | 3 (30.0) | 4 (100.0) | 0.070* |
| **Neoangiogenesis score§** | 0.5 (0.2-0.8) | 0.8 (0.5-0.8) | 0.103** |
| **MES§** | 1.9 (1.2-2.3) | 2.1 (1.6-2.1) | 0.277** |

‡n (%), §median (minimum-maximum)
MES: Microangiopathy evolution score

**Table 6.** Correlation analysis of NFCD and MES with numerical demographic and clinical parameters

| | | Capillary density | MES |
|---|---|---|---|
| **Age** | r | 0.182 | 0.074 |
| | p | 0.532 | 0.803 |
| **Age at diagnosis** | r | 0.175 | 0.053 |
| | p | 0.550 | 0.857 |
| **CMAS upon diagnosis** | r | -0.175 | 0.217 |
| | p | 0.550 | 0.457 |
| **Disease duration** | r | -0.056 | -0.149 |
| | p | 0.848 | 0.612 |
| **Duration of steroid use** | r | 0.216 | -0.361 |
| | p | 0.458 | 0.205 |
| **Follow-up** | r | -0.077 | -0.140 |
| | p | 0.794 | 0.634 |

Spearman's rho correlation coefficient was used
NFCD: Nail-fold capillary density, MES: Microangiopathy evolution score, CMAS: Childhood Myositis Assessment Scale

the disease or during clinical follow-up examinations. In addition, several scores based on capillaroscopic changes in the nail-folds were considered sensitive tools for quantifying and monitoring microvascular damage (18).

Pizzorni et al. (22) reported a positive association between MES and skin telangiectasia in systemic sclerosis. They also thought that vascular damage might be a predictive factor for future organ involvement. Avcı et al. (23) found no relationship between capillaroscopic changes and nail involvement in psoriasis and eczema. The present study found significantly higher MES values in children with JDM than in healthy children. Nevertheless, no follow-up data showed changes in the nail-fold capillaroscopic changes according to treatment duration or duration in this study. In addition, there were no significant correlations between

NFCD and MES and other clinical parameters. Previous studies have shown a relationship between NFCD, age, and disease duration (14,27). We did not find any correlation between these parameters. Our study group included only children with a mean age of 10.4±4.0 years, contrary to Sanner's study in which older patients (24.9±12.7 years) were investigated (27). Heterogeneous demographic and clinical characteristics may be factors for such conflicting findings.

NFCD is one of the most common capillaroscopic findings in several connective tissue disorders. Many researchers have reported significant decreases, as in this study (1,5,14,18,23 24). Although the extent of the avascular area was significantly different between patients with psoriasis and eczema and healthy controls, the relatively low number of children with the avascular area might be the reason for not reaching statistical differences in the present study (23). Therefore, large-scale studies are needed to describe the pathognomonic microangiopathic findings of JDM.

This study found significant differences in the NFC findings between children with and without myositis-specific autoantibodies. Children with positive autoantibodies had higher NFCD and lower intercapillary distance values. Previous studies have investigated the possible association between NFCD measurements and the positivity of several myositis-specific autoantibodies. Sugimoto et al. (13) found that the scores during nail-fold video capillaroscopy were inversely correlated with anti-melanoma differentiation-associated gene 5 antibody titers in patients with dermatomyositis. Liu et al. (28) reported higher arteriolar densities in patients with dermatomyositis and positive anti-nuclear matrix protein-2 antibodies. They also showed thickened vascular walls, thrombosis, and lipid accumulation in these patients. Positive myositis-specific autoantibodies are associated with a higher degree of perivascular inflammation in children with juvenile overlap myositis (29). Another study evaluated the impact of anti-Jo-1 antibodies on the pathology of the perimysium and neighboring muscle fibers in patients with myopathy (30). They found that capillary density was normal, contrary to reduced capillary density in dermatomyositis. The case series design of the paper should be considered when evaluating the findings. Thus, the impact of positive myositis-specific autoantibodies on capillaroscopic changes in patients with JDM remains unclear. Future studies are needed to clarify these controversial issues.

### Study Limitations

The relatively small number was the study's main limitation. The small number of children with positive myositis-specific autoantibodies prevented a more dynamic analysis. In addition, the variances between the duration of the disease and the time of NFC might be a limiting factor in obtaining more accurate findings. Prospective studies that control the disease characteristics, including duration, severity, and different phenotypic involvements, might be helpful in obtaining more powerful results.

## CONCLUSION

In conclusion, children with JDM exhibited remarkable nail-fold capillaroscopic changes that might be useful in diagnosing and differentiating connective tissue disorders in children.

### ETHICS

**Ethics Committee Approval:** This study was approved by the Erciyes University Clinical Research Ethics Committee (decision no: 2021/355, date: 05.05.2021).

**Informed Consent:** Written informed consent was obtained from the parents of the children.

### FOOTNOTES

### Authorship Contributions

Surgical and Medical Practices: Ş.D., S.N.T., Concept: Ş.D., Design: Ş.D., Data Collection or Processing: Ş.D., S.N.T., Analysis or Interpretation: Ş.D., Literature Search: Ş.D., Writing: Ş.D., S.N.T., A.P.K., M.H.P.

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