Dermatology Practical & Conceptual

# A Comparison of Clinical, Demographic and Treatment Characteristics of Pediatric-Onset and Adult-Onset Patients Diagnosed With Localized Scleroderma

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## ABSTRACT

**Introduction:** Morphea localized scleroderma (LS) is a rare skin disease with unknown pathogenesis, which causes sclerosis of the dermis and subcutaneous tissue.

**Objectives:** It was aimed to compare the characteristics of patients with pediatric and adult-onset morphea.

**Methods:** A retrospective analysis was performed on the records of 183 adult morphea patients. The demographics, clinical and laboratory characteristics, and treatment options of the patients were recorded. Adult patients with morphea over the age of 18 were divided into two groups according to the age of onset and compared.

**Results:** Twenty-two percent (N = 41) of the patients had pediatric-onset morphea (POLS) and 77.6% (n=142) had adult-onset morphea (AOLS). While POLS had a higher head-neck involvement, AOLS had a higher breast involvement ($P < 0.001$ and $P = 0.043$). Patients with linear morphea were younger, and more frequently had at least one laboratory anomaly ($P = 0.016$ versus 0.024). Anti-dsDNA positivity and low hemoglobin (Hb) were observed more frequently in patients with breast involvement. Patients with inguinal involvement, on the other hand, had lower Hb and a higher rate of diabetes, and those patients were older ($P = 0.042$, 0.040, and 0.012, respectively).

**Conclusions:** Clinical characteristics and accompanying laboratory anomalies of the patients with morphea depend on the age of onset, involvement areas and the types of morphea, having such data readily available should guide the holistic approach for, and the monitoring process of, the disease.

## Introduction

Morphea localized scleroderma (LS) is an uncommon inflammatory skin disease which causes sclerosis on the dermis and subcutaneous tissue. The pathogenesis of morphea is unknown. Numerous factors can contribute to its etiology, including trauma, environmental factors like radiation, drugs and chemicals, infections, vascular dysregulation, and autoimmunity [1-3]. These stimuli are thought to cause the release of adhesion molecules and microvascular injuries. Vascular cell adhesion molecule-1, intercellular adhesion molecule-1 and E-selectin can induce T-cells and cause the release of transforming growth factor-beta (TGFβ) and profibrotic cytokines such as interleukin (IL) 4 and IL-6. As a result, excessive collagen production and decreased matrix metalloproteinases contribute to the development of the disease [2]. It is more common in women. The incidence of morphea ranges from 0.4 to 2.7 per 100,000 people and it is similar in children and adults. [2-5]. In children with morphea it is mostly seen between ages of 2 and 14 and in adults with morphea it is mostly seen in middle ages [6]. It can lead to functional and cosmetic issues. In the inflammatory phase, erythematous purple-colored patches and plaques are observed, which gradually transform into white, sclerotic lesions that are surrounded by violet halos. As lesions heal, post-inflammatory hyperpigmentation occurs [1,7].

Laxer et al. classified morphea into the following five types: plaque, generalized, linear, pansclerotic, and mixed [8].

There are few studies compare pediatric-onset morphea (POLS) and adult-onset morphea (AOLS) with variable results [9,10].

## Objectives

The aim of this study was to investigate the clinical and demographic characteristics of both POLS and AOLS patients who were followed up with the diagnosis of morphea in our clinic, evaluate the results, examine the clinical pattern and laboratory findings, and compare the treatment options with each other in the light of the literature data.

## Methods

Retrospective data analysis was performed on the records of patients who were clinically and histopathologically diagnosed with morphea between 2016 and 2022. The inclusion criterion for the study was that the patients be at least 18 years old at the time of diagnosis. Children under the age of 18 were excluded in the study. Morphea patients who were 18 years of age or older were divided into two categories: AOLS and POLS. Those with onset of morphea before the age of 18 were considered as AOLS, and those with onset of morphea after the age of 18 were considered as POLS. Demographics such as the age of onset of the disease, its duration, patient gender, the clinical type, dermatological examination results such as the anatomical region involved, extracutaneous signs, accompanying systemic findings and symptoms, triggering factors, laboratory findings, additional comorbid disease, history of autoimmune disease, and the administered treatment option were recorded. The first examination data was used in the analysis.

Disease subtyping was done according to the Laxer classification [8]. Anatomical localization was classified as involving the head, neck, extremities, body and/or multiple areas of involvement. The presence of sclerosis (skin fibrosis) and pigmentation on the skin were examined.

### Statistical Analysis

SPSS/IBM for Windows 21.0 was used to analyze the data. The sample was defined using descriptive statistics such as percentage, mean, median, standard deviation, and interquartile range (IQR). The chi-square significance test or Fisher Exact test was used to examine categorical data, and the Student t test or Mann-Whitney U test was used to examine numerical variables. A 95% significance level (or $\alpha$=0.05 error margin) was used to identify differences in the analyses.

## Results

One hundred eighty-three adult patients were included in the study. The female/male ratio was 5.1:1.

The demographic data of the patients are shown in Table 1. The most common type of morphea was the plaque type (Figures 1-3). Morphea types were similar in POLS and AOLS. Body and extremities were the most frequently affected body sites. Head and neck involvement was more frequent in patients with POLS whereas, breast involvement was more frequent in AOLS patients. Although arthralgia was more frequent among AOLS patients. Laboratory findings, systemic diseases and laboratory findings were similar among AOLS and POLS patients. Although, anti-dsDNA positivity and systemic steroid treatment frequencies were higher in AOLS patients the results did not reach statistically significant levels.

### The Evaluation of Demographical, Clinical, Laboratory Findings, and Treatment Based on the Type of Morphea

Sclerosis (tissue fibrosis) was significantly more common in patients with generalized morphea (30.6%, P = 0.001). In generalized morphea, topical treatments were significantly less common, whereas methotrexate (MTX) and systemic steroid treatments were used significantly more (P values <0.001, <0.001, and 0.028, respectively). Patients with

**Table 1.** General clinical, demographic characteristics of all patients and comparison of pediatric and adult-onset morphea patients.

| Characteristics | Total patient N =183 (100%) | Adult-onset N = 142 (77.6%) | Pediatric-onset N = 41 (22.4%) | P |
|---|---|---|---|---|
| Gender | | | | |
| Female | 153 (83.6) | 120 (84.5) | 33 (80.5) | 0.54 |
| Male | 30 (16.4) | 22 (15.5) | 8 (19.5) | |
| Age -*years (18-82)[a] | 41.1 (16.9) | 45.5 (34-59) | 21 (15.5-28) | <0.001 |
| Age of onset years[b] | 36 (19.5-55) | 43 (29.5-57) | 13.5 (12-17) | <0.001 |
| Duration of the disease years[b] | 2 (1-6) | 1 (1-2) | 8 (5.3-12.8) | <0.001 |
| Morphea type | | | | |
| Plaque | 124 (67.8) | 98 (69) | 26 (63.4) | 0.25 |
| Linear | 20 (10.9) | 12 (8.5) | 8 (19.5) | |
| Generalized | 33 (18) | 27 (19) | 6 (18.2) | |
| Mixed | 6 (3.3) | 5 (3.5) | 1 (2.4) | |
| Involved sites | | | | |
| Head/neck | 19 (10.4) | 7 (4.9) | 12 (29.3) | <0.001 |
| Body | 115 (62.8) | 94 (66.2) | 21 (51.2) | 0.080 |
| Extremity | 101 (55.2) | 79 (55.6) | 22 (53.7) | 0.82 |
| Inguinal areas | 23 (12.6) | 20 (14.1) | 3 (7.3) | 0.25 |
| Breast | 13 (7.1) | 13 (9.2) | 0 | 0.043 |
| Triggering factors (exp;vaccination, laser application, surgery, trauma, autoimmunity and psychological stress) | | | | |
| Yes | 12 (6.6) | 9 (6.3) | 3 (7.3) | 0.82 |
| No | 171 (93.4) | 133 (93.7) | 38 (92.7) | |
| Sclerosis presence | | | | |
| Yes | 62 (33.9) | 49 (34.5) | 13 (31.7) | 0.74 |
| No | 121 (66.1) | 93 (65.5) | 28 (68.3) | |
| Pigmentation presence | | | | |
| Yes | 55 (30.1) | 29 (30.3) | 12 (29.3) | 0.90 |
| No | 128 (69.9) | 99 (69.7) | 29 (70.7) | |
| Arthralgia presence | | | | |
| Yes | 19 (10.4) | 18 (12.7) | 1 (2.4) | 0.058 |
| No | 164 (89.6) | 124 (87.3) | 40 (97.6) | |

[a]main (standard deviation, sd), [b]median (interquartile range).

generalized morphea had significantly higher platelet levels (P = 0.010), neutrophil/lymphocyte ratios (NLR) (P = 0.029) and age of onset (P = 0.042).

Patients with linear morphea had a significantly lower mean age and age of onset (P = 0.025 and P = 0.005). Patients with linear morphea had a significantly longer duration of disease (P = 0.016). At least one laboratory finding was abnormal in 21% of the patients with linear morphea (P = 0.024). In non-linear morphea, this rate was 8.1%.

Eighty percent of patients with plaque-type morphea were female while 91.5% of the patients with non-plaque morphea were female. Number of males was significantly higher in plaque-type morphea (P = 0.046). Sclerosis was found in 28.2% of patients with plaque-type morphea and in 45.8% of patients with non-plaque morphea. In plaque-type morphea, sclerosis was less prevalent (P = 0.019), and additional skin diseases were more prevalent (P = 0.049). Patients with plaque-type morphea received significantly less MTX therapy and significantly more topical treatments (P values 0.004 and <0.001, respectively).

## The Evaluation of Demographic, Clinical, and Laboratory Findings Based on Involved Sites

Pediatric onset was more frequent in patients with head and neck involvement (Figures 3 and 4). The mean age and age of onset of the disease in patients with head and neck involvement were significantly lower (P = 0.003 and P < 0.001).

Patients with body involvement used MTX and systemic steroids at a higher rate (P = 0.024). Their mean age and disease age of onset were significantly higher (both P = 0.005) and platelet levels were significantly higher (P = 0.017).

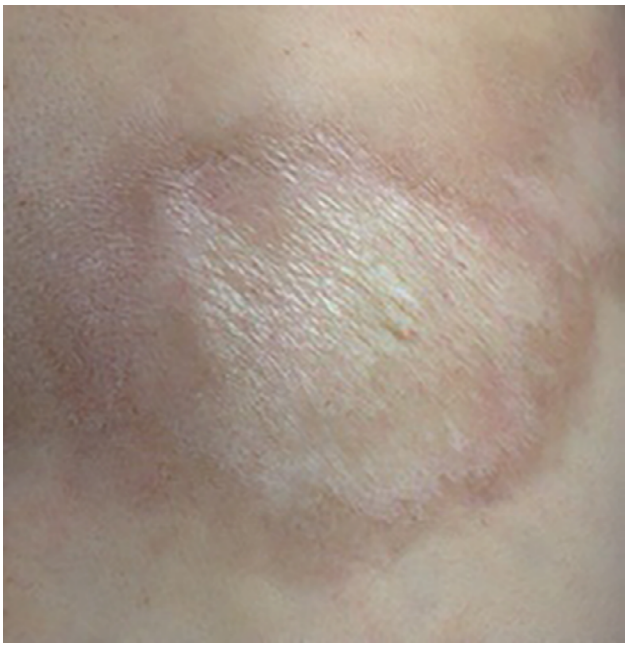

**Figure 1.** Plaque type morphea.

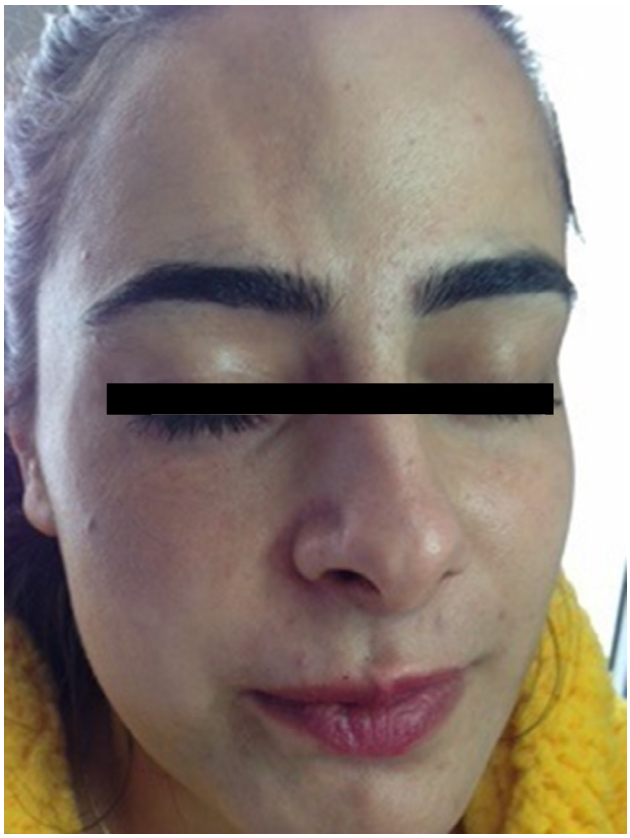

**Figure 2.** En coup de sabre.

All patients with breast involvement were adult-onset. Anti-dsDNA positivity was seen in 37.5% of the patients with breast involvement and in 9.2% of the patients without breast involvement. Anti-dsDNA positivity was significantly higher in the patients with breast involvement (P = 0.014). Hemoglobin (Hb) levels were 12.4 (SD 1.4) in patients with breast involvement and 13.6 (SD 1.4) in patients without

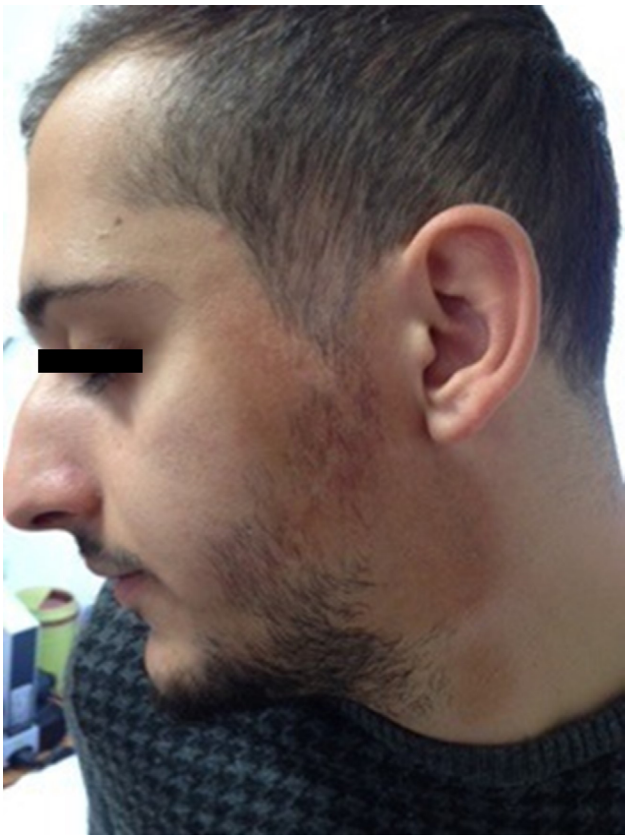

**Figure 3.** Plaque type morphea on the face.

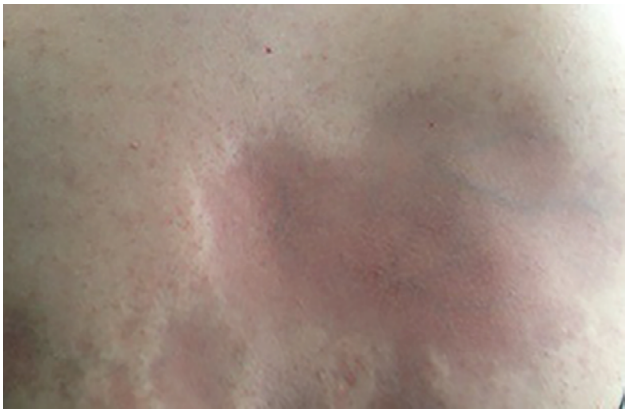

**Figure 4.** Atrophic morphea.

breast involvement. Patients with breast involvement had a significantly lower Hb (P = 0.017).

All of the patients with inguinal involvement were female. The patients were significantly older (P = 0.012), and their Hb levels were lower than the patients without inguinal involvement (P = 0.042). Sclerosis was more common in patients with inguinal involvement (P = 0.014). The incidence of diabetes in patients with inguinal involvement was higher (P = 0.040)

Table 1 shows the clinical characteristics of the patients including triggering agents and presence of arthralgia. Table 2 illustrates the laboratory findings and the Table 3 shows the accompanying comorbidities. The treatments given to pediatric morphea and adult morphea were shown Table 4.

**Table 2.** Laboratory findings of all patients with morphea and comparison of pediatric and adult-onset morphea patients.

| Characteristic | Total patient N (%) | Adult-onset N (%) | Pediatric-onset N (%) | P |
|---|---|---|---|---|
| Borelia | | | | |
| Positive | 5 (6) | 3 (4.8) | 2 (9.5) | 0.42 |
| Negative | 79 (94) | 60 (95.2) | 19 (90.5) | |
| ANA | | | | |
| Positive | 51 (38.6) | 41 (38.3) | 10 (40) | 0.88 |
| Negative | 81 (61.4) | 66 (61.7) | 15 (60) | |
| Anti-dsDNA | | | | |
| Positive | 13 (11.1) | 13 (13.7) | 0 (0) | 0.066 |
| Negative | 104 (88.9) | 82 (86.3) | 22 (100) | |
| Hb[a] | 13.5 (1.4) | 13.5 (1.4) | 13.5 (1.7) | 0.71 |
| Leukocyte[a] | 6.7 (1.8) | 6.8 (1.8) | 6.1 (1.4) | 0.14 |
| Platelet[a] | 276.7 (67.8) | 275.1 (66.6) | 288.9 (77.6) | 0.78 |
| NLR[b] | 1.9 (1.5-2.6) | 1.9 (1.5-2.6) | 1.9 (1.3-2.2) | 0.33 |
| Eosinophil[b] | 0.14 (0.08-0.25) | 0.14 (0.09-0.25) | 0.15 (0.07-0.29) | 0.95 |
| Eosinophil (%)[b] | 2.1 (1.2-3.4) | 2.1 (1.2-3.4) | 2.6 (1-3.2) | 0.77 |
| Sedimentation[b] | 10 (6-19) | 10 (6-19) | 9 (5.3-17) | 0.52 |
| CRP[b] | 0.06 (0.01-7.5) | 0.07 (0.01-10) | 0.03 (0.005-2) | 0.17 |
| Vit D deficiency | 38 (20.76%) | 30 (21.12) | 14 (19.5) | 0.19 |
| Iron deficiency | 47 (25.68%) | 33 (23.23) | 14 (34.1) | 0.11 |
| Vit $B_{12}$ deficiency | 18 (9.8%) | 12 (8.45) | 6 (14.6) | 0.46 |

NLR = neutrophil lymphocyte ratio.
[a]main (standard deviation, sd), [b]median (interquartile range).

## Conclusions

In our study, the characteristics of pediatric and adult-onset morphea patients were compared, and some of them showed similarities with the literature, while some findings showed differences [8-19].

According to the previous studies, disease onset was between the ages of 2 and 14 for children and 43.3 to 44.8 for adults [8,11,13,15,17,18,20]. We found that the average age at which the disease manifested itself was 36-43 for AOLS, and 13.5 for POLS.

The incidence of linear morphea in children range from 15.3% to 81% [2,4,5,11,17,18,21]. In this study, linear morphea was found to be 19.5% for POLS and 8.5% for AOLS Supporting the fact that the linear type starts more frequently in the childhood [6,11,14]. Those patients with the pediatric onset naturally had a longer duration of illness. Some researchers state that the disease is more active (20%-59%) in patients with POLS [2,5,11]. The linear type was also reported to be more sclerotic than in adult-onset patients, and in the present study, we found that there was more sclerosis in both linear and generalized types. Sarocino et al found laboratory anomalies as 40.4% in linear type [18]. In this study, at least one laboratory anomaly was present in 21% of the patients, especially in the group with linear morphea. Since the platelet and ESR levels are significantly lower in this type compared to the generalized type, there may be a lower risk of systemic involvement and a lower tendency to thrombosis. The fact that the extremity involvement is around 50% in both types of onset suggests that the disease has a progressive course.

In various studies, the incidence of plaque type in children ranges from 15.3% to 69.2% [6,14-17]. In the present study, the plaque type was found to be high in both groups. This rate was similar to the rates in our previous research with children cases [17]. A significant data here was that the plaque type was higher in men, but sclerosis was less common than other types. Morphea appears to have a milder course in men. This finding also supports the view that autoimmune diseases are more frequent and more severe in women.

Mixed type, in which more than one type of morphea coexists, was found at a rate of 7.6%-23% in various studies in children [2,13,15-17]. Saracino et al reported mixed type morphea in 27.1% of the adults with morphea, in 28% of POLS and 25.9% of AOLS [18]. In our study, we found that mixed type morphea was 3.3% lower in total, 3.5% lower in AOLS and 2.4% lower in POLS.

**Table 3.** Comparison of presence of additional disease of adolescent- and adult-onset morphea patients.

| Comorbid diseases | Adult-onset N = 142 patients | Pediatric onset N = 41 patients | Total N = 183 |
|---|---|---|---|
| Skin diseases N (%) | | | 13 (7.1) |
| Vitiligo | 3 (2.11) | - | 3 (1.63) |
| Psoriasis | 3 (2.11) | - | 3 (1.63) |
| A.Areata | 2 (1.4) | - | 2 (1.09) |
| LSA | 3 (2.11) | - | 3 (1.63) |
| BCC | 1 (0.7) | - | 1 (0.5) |
| Verruka vulgaris | 1 (0.7) | - | 1 (0.5) |
| Rheumatological diseases, N (%) | | | 7 (3.18) |
| Ankylosing spondylitis | 1 (0.7) | - | 2 (1.09) |
| Rheumatoid arthritis | 3 (2.11) | - | 2 (1.09) |
| Scleroderma | 1 (0.7) | - | 1 (0.5) |
| Sacroileitis | 1 (0.7) | - | 1 (0.5) |
| Spondyloarthropathy | 1 (0.7) | - | 1 (0) |
| Thyroid disease N (%) | 15 (10.56) | 1 (2.4) | 16 (8.74) |
| Diabetes mellitus N (%) | 11 (7.74) | 3 (7.31) | 14 (7.65) |
| Hypertension, N (%) | 15 (10.56) | 2 (4.9) | 17 (7.1) |
| Asthma N (%) | 3 (2.11) | 2 (4.9) | 5 (1.73) |
| Fibromyalgia, N (%) | 2 (1.4) | - | 2 (1.09) |
| Ulcerative colitis, N (%) | 1 (0.7) | - | 1 (0.5) |
| Autoimmune hepatitis, N (%) | 1 (07) | - | 1 (0.5) |
| Familial Mediterranean Fever, N (%) | 2 (1.4) | - | 2 (1.09) |
| Ferritin deficiency anemia N (%) | 33 (23.23) | 14 (34.1) | 47 (25.68) |
| Vitamin $B_{12}$ deficiency anemia N (%) | 12 (8.45) | 6 (14.6) | 18 (9.8) |
| Vitamin D deficiency N (%) | 30 (21.12) | 8 (19.5) | 38 (20.76) |
| Depression N (%) | 2 (1.4) | 1 (2.4) | 3 (1.63) |
| Other diseases migraine, vertigo, renal failure N (%) | 3 (1.4%) | - | 3 (1.5%) |
| Total N (%) 101 (55.19%) | 137 (96.47%) | 36 (87.80%) | 173 (95%) |

**Table 4.** Comparison of treatments given to pediatric morphea and adult morphea.

| Treatments used | Total patients | Adult-onset | Pediatric-onset | P |
|---|---|---|---|---|
| Topical treatments, N (%) | 120(65.6) | 89 (62.7) | 31 (75.6) | 0.13 |
| Methotrexate, N (%) | 50(27.3) | 39 (27.5) | 11 (26.8) | 0.94 |
| Colchicine, N (%) | 31(16.9) | 21 (14.8) | 10 (24.4) | 0.15 |
| Systemic steroid, N (%) | 12(6.6) | 12 (8.5) | 0 | 0.054 |
| Phototherapy, N (%) | 9(4.9) | 7 (4.9) | 2 (4.9) | 0.99 |
| Hydroxychloroquine, N (%) | 7(3,8) | 5 (3.5) | 2 (4.9) | 0.69 |
| Others N (%) Zinc, Salazopyrin, Vitamin D Mycophenolate mofetil, Azathioprine, Isotretinoin, | 10 (1.09-0.54) | 7 (4.9) | 3 (7.3) | |
| Untreated N (%) | 22 (15.38) | | 6 (4.22) | 0.84 |

Although the affected areas were similar in both groups of patients, special region involvement such as the inguinal region was more common in AOLS. Breast involvement was observed only in AOLS. This particular group of patients were older and female.

There are multiple localizations in generalized morphea. The previous studies found it as 7%-47% [2,4,6,17]. In our study, the rate of generalized morphea was 18% in total, 19% in AOLS, and 18.2% in POLS. High platelet levels in this group of patients may indicate a proclivity for

thrombosis. More research should be conducted on this subject. The ratios of neutrophils / lymphocytes (NLR), which are indicators of acute inflammation in this type, were also found to be significantly higher. This may indicate that there is a tendency of inflammation in generalized morphea, and that it may be even more progressive. Therefore, these markers can be used for treatment follow-up and progression [22].

In various studies, factors that can trigger the formation of the disease in morphea were identified as 7.3-16.6% [3,18,23]. Saracino et al determined the presence of triggering factors as 16.5% in all morphea patients, 20.9% in AOLS and 14.7% in POLS [18]. We found these rates as 6.6% in general, 6.3% in AOLS and 7.3% in POLS. These were consistent with the literature.

Some studies confirm the association of infectious agents such as Borrelia with morphea, while others do not [3,24]. Only 5 (6%) of the cases in the present study were detected to have the Borrelia antibody.

In morphea patients, extracutaneous abnormalities have also been reported in addition to skin abnormalities [14,25]. These findings range from 10.9% to 46 % [18,20,25]. There have been reports of extracutaneous involvement being more prevalent in generalized and mixed type [13,17]. We found extracutaneous involvement (arthralgia) in 10.4% of the cases. The low rate might be due to the fact that the cases could not be questioned because of the retrospective nature of the study.

Some authors have reported more frequent (43%-45%) deformities in children with linear morphea [15,16]. Saracino et al detected functional impairment in 24.8% of adult, 28% of pediatric-onset and 20.7% of adult-onset patients [18]. In our study, although linear morphea was found in 10.9% of the cases, no functional impairment was detected except for cosmetic problems.

Morphea is an immune disorder and can often be accompanied by various systemic, autoimmune, and rheumatological diseases. Diseases such as thyroid diseases and diabetes play a predisposing role. The presence of accompanying autoimmune systemic disease differs between 1%-4.9 % [6,17,18,20,25]. In some studies, the association of autoimmune diseases is up to 17 % [6,14,20,21]. Saracino et al found autoimmune disease comorbidity as 11.3% in general, 15.5% for AOLS and 8.0% for POLS [18]. In our study, the rate of comorbid diseases was similar to other studies [19,26]. Eleven patients had comorbid autoimmune skin disease. These diseases were only in the adult-onset group. Comorbid diseases increase with age [11]. Furthermore, in our cases, iron deficiency was found in 25.7% of the women. Low Hb levels may be a sign of an additional comorbid disease. In the present study, the rate of vit $B_{12}$ deficiency anemia was 9.8%, and the rate of vit D deficiency was 20.8%.

The most comprehensive multi-centred study so far was conducted by Zulian et al with 750 patients with juvenile morphea [13]. These authors found that patients with linear morphea had a high number of ocular and neurological findings [25,27]. However, unlike other studies conducted in Turkey, no neurological or ocular involvement was observed in the cases with ECDS in our study [8]. This could also be due to the fact that the study was retrospective and the records were missing.

Various laboratory changes in the serum of morphea patients have also been observed in studies [15,28-31]. Rheumatoid factor (RF), C-reactive protein (CRP), and ESR is higher in patients with extracutaneous involvement [13]. The authors reported high inflammatory parameters (ESR, CRP, RF) rather than skin manifestations, especially in patients with deep morphea and arthritis [13,14,32]. The RF rate in the patients with joint damage is up to 25%-40% [13,28,33]

Morphea is frequently associated with auto-antibody positivity [28,31,33-35]. The clinical and prognostic significance of these autoantibodies is unknown [13]. In the generalized type, it may be especially important in clinical and prognostic terms or for investigating the presence of comorbid diseases [31].

In the literature, ANA positivity was detected in 5.9%-68.7% of the cases [6,15,17,25,31]. The rate of ANA positivity has been reported to be higher in patients with generalized and mixed types, as well as those with extracutaneous involvement [6,10,11,15,31,34]. According to some studies, there is no difference in the ANA ratio among the various subtypes of morphea, but the presence of extracutaneous findings may increase these rates [28,34].

Some studies have also reported that ANA positivity varies between patients with AOLS and POLS [6,10,11,19,31]. In our study, the ANA positivity rate was found 38.3% for AOLS and 40 % for POLS, which was similar in both groups.

In addition to ANA, Scl-70, anti-dsDNA, anti-histone antibody positivity can be seen in patients. Some researchers found anti-dsDNA positivity in 4%-8% of morphea patients with AOLS and POLS [25]. These rates were particularly higher in those with the linear type and with joint involvement [28,29]. In our study, the anti-dsDNA antibody was found positive only for AOLS (13.7%, N = 13), but not for POLS.

Sato et al found that anti-histone antibodies were related to the number of morphea lesions and the skin localization of the lesions, and suggested that they may be serological indicators in cases with generalized morphea [29]. We detected anti-histone antibodies in only one patient. The presence of autoantibodies may indicate that morphea is a connective tissue disease with systemic involvement rather than a skin disease [31,35].

Due to the incomplete understanding of the cause of morphea, there is no specific effective treatment. The purpose of

the treatment is to stop the disease from progressing at an early stage in order to avoid functional and cosmetic complications. Treatment options were assessed according to the subtype of the disease, the extent of involvement, the level of activation and the presence of complications [27,36].

While some studies report that topical treatment was administered at a rate of 45.9 %, in this study, topical agents were preferred as the first line in 65.6% (N = 120) of both groups [18].

In our study, patients with limited plaque-type lesions were treated with topical corticosteroids, calcipotriol, calcipotriol + steroid combination, tacrolimus and pimecrolimus. This rate was found to be 65.6% (N = 120) in general, 62% in AOLS, and 75.6% in POLS.

Even if topical medications alone are effective in treating plaques, they are unable to suppress disease activity. Therefore, they are not advised to be used alone in the treatment of severe, functionally disruptive, and progressive diseases [36]. MTX, phototherapy, systemic steroids, and additional systemic immunosuppressants should be added to prevent future complications in linear, generalized, and mixed morphea types [7,27,36,37]. Generalized-, mixed-, and linear-type patients in this study, who represented about half of the cases for comparable reasons, were treated systemically. MTX (54.6%) was the most frequently used systemic treatment in both groups and systemic steroids were preferred only in AOLS.

Majority of authors suggest aggressive treatments for morphea types such as linear, generalized morphea which cause deformity [36]. In various studies, it has been demonstrated that MTX and MMF are effective treatment options in resistant cases [15,36]. It has been reported in various studies that children were administered MTX at rates of 17.8%–42 % [24,37]. In this study, MTX treatment was given at a rate of 27.3% (N = 50) in general, 27.5% in AOLS and 26.8% in POLS. Litaiem et al administered a combination of MTX and steroids to 28% of patients. In this study, this combined treatment rate was 8.5% [38].

The authors of some studies reported positive outcomes for patients who received colchicine [39,40]. In this study, colchicine was generally given at a rate of 16.9% (N = 31).

According to some researchers, treatment of plaque-type morphea is unnecessary because lesions will naturally regress in three to five years [25,27,36]. Zulian et al showed that 17% (N = 128) of the patients did not receive any treatment [25]. Fifteen percent of the patients in this study did not require treatment.

Treatments administered provided partial recovery in the majority of the patients. Full recovery rate was 25.4% for the AOLS patients and 29.3% for the POLS patients.

Adult and pediatric onset morphea have different disease characteristics, different systemic associations and laboratory anomalies. That why patients with adult and pediatric onset morphea require different management and treatment strategies. Additionally, clinicians should be aware of the extracutaneous disease and anomalies that may accompany morphea and patients should be examined accordingly.

The retrospective nature, inability to create a morphea severity scale, and the lack of standardization in the treatment administered for different types of morphea are the limitations of our study. Multi-centered, more comprehensive and prospective studies are required to obtain more accurate data.

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