

# Plaque Psoriasis Formed on Linear Morphea During Topical PUVA Therapy: Case Report

## Topikal PUVA Tedavisi Sırasında Lineer Morfea Üzerinde Oluşan Plak Psoriazis

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**ABSTRACT** Linear morphea is a type of morphea that is commonly seen in childhood. It frequently starts as an erythematous inflammatory plaque and evolves into an atrophic band, causing limitation of movement at the affected extremity. Psoriasis is an inflammatory, scaly erythematous disease, which rarely follows a segmental fashion. The presence of morphea and psoriasis in the same patient has been rarely reported in the literature. Exposure to ultraviolet (UV) radiation is usually beneficial in psoriasis. However, it is recognized that psoriasis worsens in a significant minority of cases in response to UV treatment. We report a case of an 11-year-old female who developed newly diagnosed psoriasis plaques on a linear morphea lesion during PUVA treatment. The main features of psoriasis caused by phototherapy and the association of psoriasis with linear morphea are discussed.

**Key Words:** Scleroderma, localized; psoriasis; phototherapy

**ÖZET** Lineer morfea sıklıkla çocukluk çağında görülen bir morfea alt tipidir. Genellikle, eritemli, inflamatuvar lineer bir plak şeklinde başlar ve etkilenen ekstremitede hareket kısıtlılığına neden olan atrofik bant şeklinde görülür. Psöriazis ise nadiren segmental olarak görülen eritemli skuamli inflamatuvar bir hastalıktır. Psöriazis ve morfeanın aynı hastada görüldüğünü bildiren az sayıda yayın vardır. Ultraviyole maruziyeti psöriazis tedavisinde başarılı bir şekilde uygulanmaktadır. Fakat az sayıda olguda UV tedavisinin psöriazisi şiddetlendirdiği bildirilmiştir. Burada lineer morfea tanısı ile lokal PUVA tedavisi aldığı sırada psöriazis lezyonu ortaya çıkan 11 yaşında kız çocuğu bildirilmektedir. Bu yazıda psöriazis ve morfea birlikteliği ile fototerapinin tetiklediği psöriazis olasılığı tartışılmıştır.

**Anahtar Kelimeler:** Skleroderma, lokalize; psöriazis; fototerapi

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Scleroderma is a chronic disease of unknown etiology characterized by skin fibrosis. It is divided into two clinical entities: localized scleroderma (LS) and systemic sclerosis.<sup>1</sup> T cell mediated cytokines are thought to play a role in the etiology of morphea like psoriasis and other autoimmune diseases.<sup>2</sup> LS has an estimated annual incidence rate of 0.4-2.7 per 100 000 individuals and usually appears in school-aged children.<sup>3,4</sup> LS has been classified into the following three types: plaque morphea, linear morphea and generalized morphea. This classification has been widely accepted. Children are more likely than adults to develop linear morphea.<sup>5</sup> Hypopigmented lesions occur in all of the subsets.<sup>6</sup>

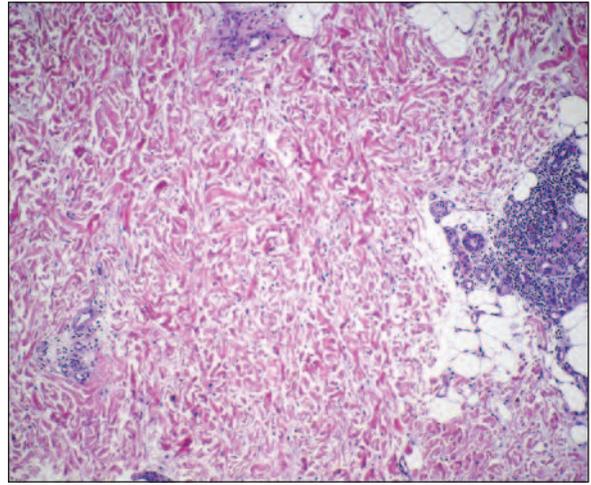
## CASE REPORT

An 11-year-old girl presented to our dermatology department with an asymptomatic hypopigmented linear patch extending from the left knee to the dorsum of the left foot (Figure 1). The patient had neither systemic complaints nor a history of medication. The lesion had developed a few months earlier. A skin biopsy from a hypopigmented plaque on the left limb revealed epidermal atrophy, hyperkeratosis, increased dermal collagen bundles and perivascular mononuclear infiltration (Figure 2). The results of the initial investigations, including routine analyses of hematological parameters, full biochemical profiles, erythrocyte sedimentation rate were normal. Antinuclear antibody, rheumatoid factor, anti-Scl-70 and anti-double stranded DNA antibodies were negative. On the basis of these findings, the patient was diagnosed as linear morphea and topical PUVA with 8-methoxypsoralen therapy three times a week was initiated. An erythematous scaly lesion was noticed on the morphea plaque at the 27. session of the treatment (Figure 3). The phenomenon of wax stain and auspitz sign were positive on the lesion clinically. A skin biopsy from the erythematous scaly plaque on the left ankle revealed parakeratosis, hyperkeratosis, papillamatosi and hypogranu-



**FIGURE 1:** An asymptomatic hypopigmented linear patch extending from the left knee to the dorsum of the left foot

(See color figure at <http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html>)



**FIGURE 2:** Increase of dermal collagen bundles and perivascular mononuclear infiltration (hematoxylin and eosin staining X 20 magnification).

(See color figure at <http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html>)



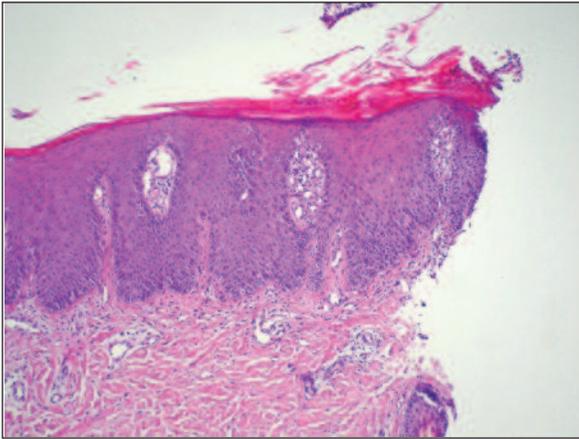
**FIGURE 3:** An erythematous scaly lesion on the plaque of morphea .

(See color figure at <http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html>)

losis (Figure 4). The patient did not have any skin lesions elsewhere on the body. On the basis of these findings, the patient was diagnosed as psoriasis and topical steroid and calcipotriol combination therapy was started. At the 47. session, the psoriasis plaques had regressed and the plaque on the morphea lesion showed distinct remission.

## DISCUSSION

Autoimmune diseases with morphea have been well documented in the last two decades and re-



**FIGURE 4:** Parakeratosis, hyperkeratosis, papillomatosis and hypogranulosis (hematoxylin and eosin staining X 10 magnification)

(See color figure at <http://www.turkiyeklinikleri.com/journal/turkiye-klinikleri-journal-of-case-reports/1300-0284/tr-index.html>)

cently this disease has been generally considered as having an autoimmune background.<sup>4</sup> The coexistence of morphea with other autoimmune diseases, including systemic lupus erythematosus, vitiligo, primary biliary cirrhosis, autoimmune hepatitis, Hashimoto's thyroiditis, bullous pemphigoid and myasthenia gravis has been shown.<sup>2,7</sup>

The association of psoriasis with morphea has rarely been reported.<sup>5</sup> Leitenberger et al. reported that 18% of adults with morphea have a concomitant autoimmune disease, including psoriasis.<sup>4</sup> Five of the 245 morphea patients in their study had psoriasis.<sup>4</sup> Walls et al. reported eight patients who had both morphea and psoriasis at the same time according to data from two tertiary care hospitals covering an 11-year period. Four of the eight patients had circumscribed morphea, whereas the other four had generalized morphea. In six patients, morphea occurred first. The first patient was using methotrexate the second was using hydroxychloroquine and the third was taking hydroxychloroquine plus mycophenolate mofetil. Three of the patients were not undergoing treatment. Two of the patients had psoriasis before the morphea occurred. One of the psoriasis patients was using methotrexate when the morphea occurred. There was no information available about the psoriasis clinic.<sup>5</sup> In another study Zulian et al. reported that 21 of 750 patients with localized juvenile sclero-

derma had psoriasis.<sup>1</sup> However, in this study there were no data about the date of start of the psoriasis lesions or the previous treatment.<sup>1</sup> Psoriasis associated with localized plaque morphea and with superficial and bullous morphea have been reported.<sup>8-11</sup> One case involved a 24 years old woman who had plaque morphea since 20 years. The plaque psoriasis developed on her knees, elbows and ankles.<sup>9</sup> Another case reported by Bilen et al. involved a 62 years old woman who developed generalized plaque psoriasis and plaque morphea simultaneously.<sup>8</sup> In the current case the psoriasis was localized solely on the plaque morphea. There was no family history of psoriasis or morphea in our patient. To our knowledge, this is the first report associating linear morphea with psoriasis vulgaris at the same localization, during topical PUVA.

The factors linking morphea and psoriasis remain poorly understood. Mechanical trauma and genetic basis have been suggested. Morphea is pathologically defined as an increase in collagen bundles and extracellular matrix deposition. The role of the increase the expression of vascular cell adhesion molecule-1 (VCAM-1) and intercellular adhesion molecule-1 (ICAM-1) in the pathogenesis of morphea is well known.<sup>12</sup> The role of ICAM-1 and VCAM-1 in the pathogenesis of psoriasis has also been discussed.<sup>13</sup> These adhesion molecules may be involved in the pathogenesis of both diseases in patients who are genetically predisposed.

Phototherapy (PUVA) is a widely used and effective treatment in psoriasis. It has been shown to reduce cutaneous T-cell activity and to have a demonstrable suppressive effect on Langerhans cells, cutaneous macrophages and the function of keratinocytes.<sup>14</sup> In addition, PUVA has been shown to induce matrix metalloproteinase, leading to an increase in collagen breakdown.<sup>15</sup> Several case reports and small studies using both oral and topical psoralen have documented the benefit of PUVA in the treatment of morphea.<sup>16,17</sup>

Exogenous factors, such as trauma, infections, alcohol, tobacco, drugs, stress, occupation and cold and foggy weather, may be responsible for the etiopathogenesis of psoriasis, relapses and deterior-

ration observed in current practise.<sup>18</sup> Exposure to UV radiation is usually beneficial in psoriasis, but the condition worsens in a significant minority of cases.<sup>19</sup> Severely photosensitive psoriasis shows a pronounced female predominance, develops at an early age, and is associated with a positive family history in the majority.<sup>19</sup>

Psoriasis lesions appeared solely on the area where topical PUVA was applied in the current case. No other psoriatic plaque developed during follow up. The lesions decreased with topical steroids and calcipotriol although PUVA was con-

tinued. Due to the absence of new lesions, despite the continuation of the phototherapy, we conclude there is a relationship between psoriasis and morphea on the basis of autoimmunity. In conclusion, further studies of patients with psoriasis and morphea are required to understand the relative disease courses and potential common pathophysiologic mechanisms.

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