Psoriasis: Targets and Therapy

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CASE REPORT

Successful Treatment of Linear Psoriasis With the IL-17a-Antagonist Ixekizumab: A Case Report

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Abstract: Linear psoriasis (LP) represents a rare variant of psoriasis. The clinical presentation includes erythematous plaques distributed along the Blaschko lines, reflecting the presence of embryological mosaicism. The clinical and histopathological features of this condition show many similarities with inflammatory linear verrucous epidermal nevus (ILVEN), which presents a challenge in differential diagnosis. Currently there is no disease-specific treatment guidelines causing a challenge in the therapeutic management. In this case report we describe a 28-year-old patient with LP. Clinically characterized by persistent, psoriasiform lesions that proved refractory to treatment with topical corticosteroids, vitamin D analogs, and systemic dimethyl fumarate. The histopathological findings showed psoriasiform epidermal hyperplasia with alternating ortho- and parakeratosis, subepidermal capillary dilatation, and perivascular lymphocytic infiltrates, confirming the diagnosis. Ixekizumab, a IL-17A antagonist, was administered leading to a rapid and significant reduction in disease severity within the first 16 weeks. The Psoriasis Area and Severity Index (PASI) decreased from 12.5 to 1.0 and as well as the Dermatology Life Quality Index (DLQI) improved to 1. Both scores prove significant improvement in quality of life and clinical severity. This case report shows the importance of histological confirmation in differentiating LP from clinically similar appearing diseases like ILVEN and highlights the potential therapeutic benefit of IL-17 blockade in LP. In addition, the findings emphasize the need for systematic studies to develop evidence-based treatment strategies for this rare psoriasis phenotype.

Keywords: psoriasis, linear psoriasis, biologics, genodermatosis, ILVEN, IL-17A-antagonist, ixekizumab

Introduction

Linear psoriasis (LP) is a unique variant of psoriasis that presents with lesions along the Blaschko lines, reflecting embryonic skin development patterns.[1–3] Clinically, LP features well-demarcated erythematous plaques with silvery scaling. The lesions are often unilateral and primarily affect the trunk and extremities but may also involve scalp and nails. Nail involvement is common, with changes such as pitting, onycholysis and subungual hyperkeratosis contributing to the overall disease burden.[4] The pathogenesis has been linked to genetic mosaicism, such as somatic HRAS mutation, but currently remains unclear.[5]

The clinical presentation of LP can overlap with conditions like inflammatory linear verrucous epidermal nevus (ILVEN) and other autoinflammatory keratinization diseases, complicating diagnosis. The absence of official treatment guidelines necessitates reliance on clinical experience, underscoring the importance of documenting cases and improve management strategies for patients with LP. We present the successful treatment of LP with ixekizumab.

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Case Report

In March 2023, a 28-year-old patient visited our clinic for inflammatory skin diseases presenting with eczematous and psoriasiform lesions. These first began in March 2020 as erythematous, plaque-like lesions on the right flank. The condition remained stable until January 2021, when the pre-existing lesions spontaneously increased in size and induration and spread to the right arm, hand, and knee. The patient did not have any other health issues as well as no signs of arthritis were detected.

Following the failure of various topicals with steroid-containing preparations to improve the lesions, two biopsies were obtained in 01/2021 to verify the diagnosis.

Topical therapy was conducted with vitamin A-containing preparations, vitamin D3-containing preparations and steroid-containing preparations. Furthermore, a bathing PUVA therapy (04–06/2022, 21 sessions) and a UVB therapy, both with no discernible effect, were carried out. Systemically, the patient was administered dimethyl fumarate from 10/2021 to 04/2022 at an initial dose of 30 mg (0-0-1) in week one, 30 mg (1-0-1) in week two and 30 mg (1-1-1) in week three. Subsequently, the dose was increased to 120 mg four times daily, also without response to treatment and stopped due to gastrointestinal side effects.

Clinical Presentation

At first presentation the patient showed extensive sharp-boarded erythematous and squamous plaques on the right arm and the right side of the trunk. Some of these plaques exhibited multiple excoriations and confluent serous crusts in proximity to isolated hemorrhagic crusts. On the dorsal aspect of the right hand, from the proximal phalanx to the fourth digit linear, grouped papules on an erythematous ground was observed. The plaques were predominantly trunk focused, with a few extending up to the digits 2–4. Additionally, distal onycholysis and subungual hyperkeratosis were observed on the fingernail of the third digit. The sharp border along the anterior median line, which did not extend to the other half of the body, was notable (Figure 1a–f). The Psoriasis Area and Severity Index (PASI) score was 12.5, Body Surface Area (BSA) 14 and Dermatology Life Quality Index (DLQI) 16. No itching was reported. A skin biopsy revealed psoriasiform epidermal hyperplasia characterized by alternating ortho- and parakeratosis (Figure 2a and b). Neutrophils and lymphocytes were observed exocytosing in the epidermis. Subepidermally a dilatation of the capillaries in the papillae accompanied by a mixed perivascular infiltrate was seen. The middle and deep corium did not reveal specific findings.

Diagnosis

Based on the clinical findings and the histological report, the clinical diagnosis of a linear psoriasis (LP) was concluded.

Therapy and Course

In the light of primary non-response to systemic therapy with dimethyl fumarate in combination with various topical agents, a shared decision with the patient was made to start Ixekizumab. Currently, there are no official treatment guidelines for linear psoriasis. Therefore, it is reasonable to follow the established protocols for psoriasis vulgaris. It is recommended that the guideline-compliant therapy option with an IL-17 inhibitor be employed regarding efficacy, safety, and tolerability. In the interim period preceding the commencement of systemic therapy, local therapy was administered in the form of a fixed combination of vitamin D and glucocorticoid (calcipotriol/betamethasone 50 micrograms/g + 0.5 mg/g cream) and a calcineurin inhibitor in alternation.

Ixekizumab 2×80 mg was administered for the first time on April 19, 2023. Approximately four months after the initiation of systemic therapy, there was a significant improvement in the findings which revealed the large, reddish-brown residual macules on the right arm and hand, as well as on the right side of the trunk. Some of these macules exhibited erythematosquamous characteristics, while others displayed papular features. There were no itching sensations or adverse effects. Furthermore, it was observed that the fingernail of the third digit on the right continued to exhibit distal onycholysis. (Figure 3a–f). The PASI was 1.0, BSA 7 and DLQI 1, reflecting residual lesions but normalized health-related quality of life.

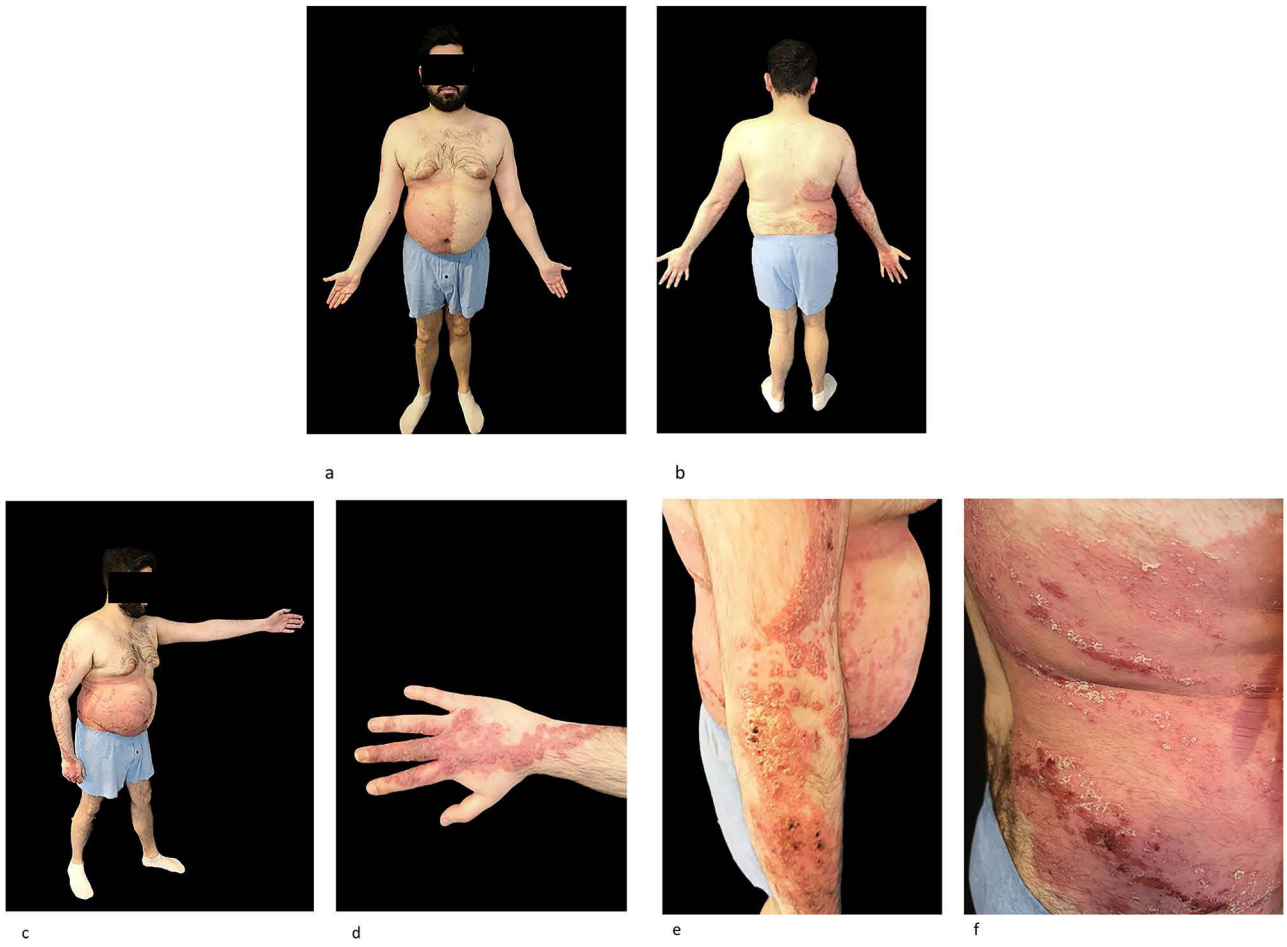

Figure 1 Photographs of the patient before treatment with ixekizumab (**a**) frontal view of the patient showing psoriasiform lesions according to Blaschko lines. The lesions are confined to the right half of the body, (**b**) posterior view of the patient showing psoriasiform lesions according to Blaschko lines. The lesions are confined to the right half of the body, (**c**) right side of the patient, (**d**) right hand of the patient, (**e**) right arm of the patient, (**f**) right upper body of the patient.

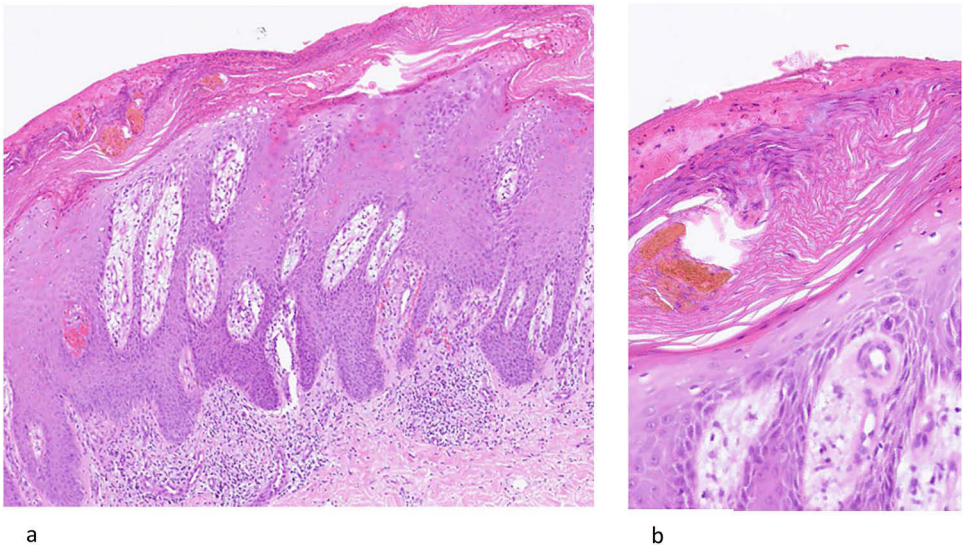

Figure 2 Histopathology of skin lesions before treatment. (**a**, **b**) psoriasiform epidermal hyperplasia characterized by alternating ortho- and parakeratosis with neutrophils.

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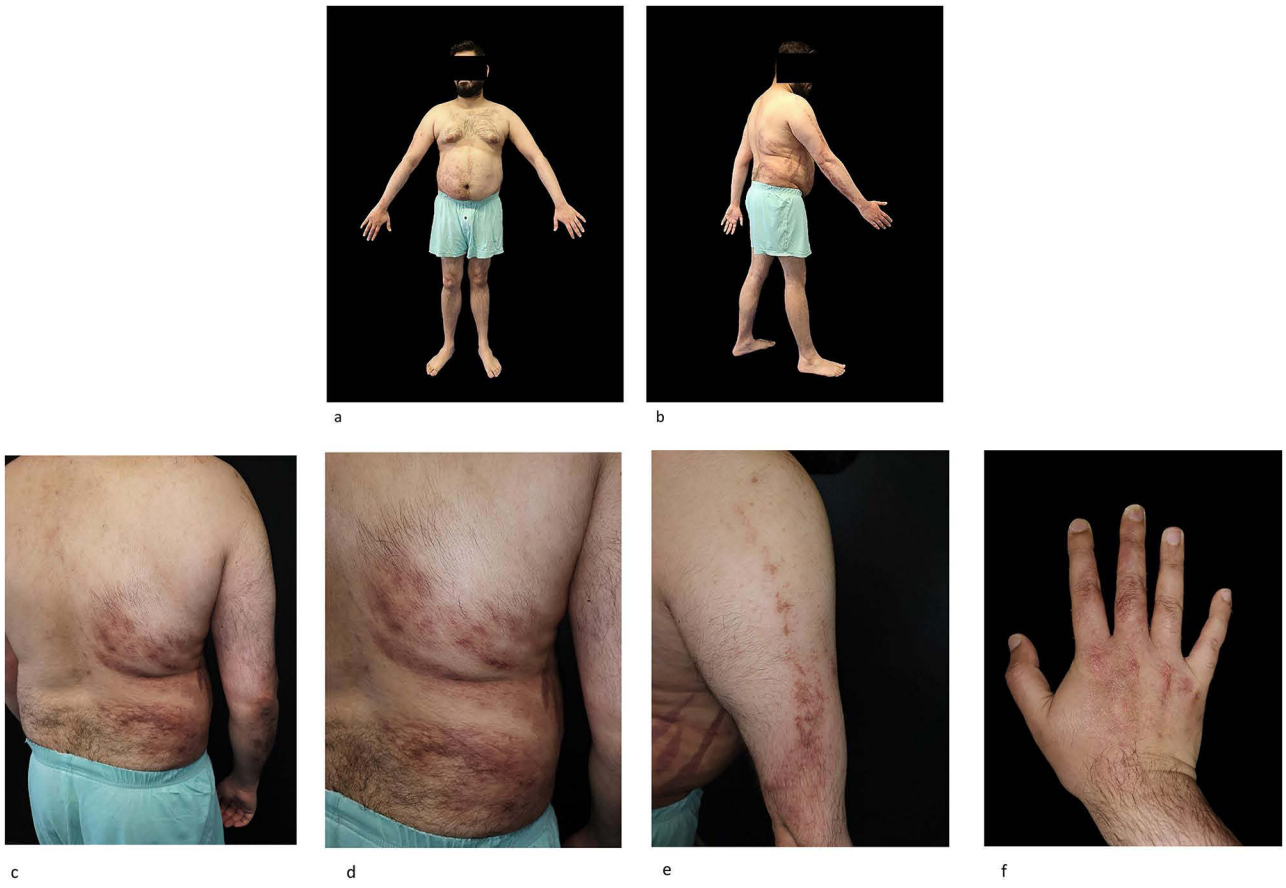

Figure 3 Full body and close-up images of the patient 16 weeks after initial treatment with Ixekizumab. (**a**) Frontal view of the patient after treatment showing postinflammatory hyperpigmentation, (**b**) right side of the patient after treatment showing postinflammatory hyperpigmentation, (**c–e**) close up images of the right side trunk and arm of the patient, (**f**) right hand of the patient.

The patient was last seen in April 2024 and reported satisfaction with the therapy. The DLQI was 1. Only residual erythema without infiltration was seen. The pictures of the patient can be seen in Figure 4.

Linear psoriasis (LP) is a rare psoriasis subtype that is poorly documented and frequently misdiagnosed as ILVEN due to the clinical and histological similarities between the two diagnoses.[6] Lacking documented and published cases make a proper estimation of prevalence for LP difficult, and the overlap with ILVEN is likely to result in underreporting and underdiagnoses. In LP Ixekizumab has shown effectiveness in a few cases.[7–9]

Due to the polymorphic nature of the skin findings, a visual diagnosis is challenging and may be subject to misinterpretation. The histological examination provided corroboration of the diagnosis of psoriasis vulgaris and enabled the exact diagnosis to be confirmed. Following the administration of an IL-17 blockade, there was a notable improvement in the patient's skin findings and quality of life within a relatively short period of time. In mosaic patients with psoriasis, a diligent physical examination and careful histological work-up are essential. The various differential diagnoses, each accompanied by a range of treatment options, and the absence of a treatment guideline for LP, present difficulties in determining the most appropriate course of treatment. LP exhibits certain parallels to genodermatoses. Consequently, it is recommended that collaborations with dermatologists in specialist centers are established. Furthermore, immunohistochemical examinations can support the selection of therapy, improve defining the mechanisms of disease, and allow a better understanding of the underlying mechanisms of action.

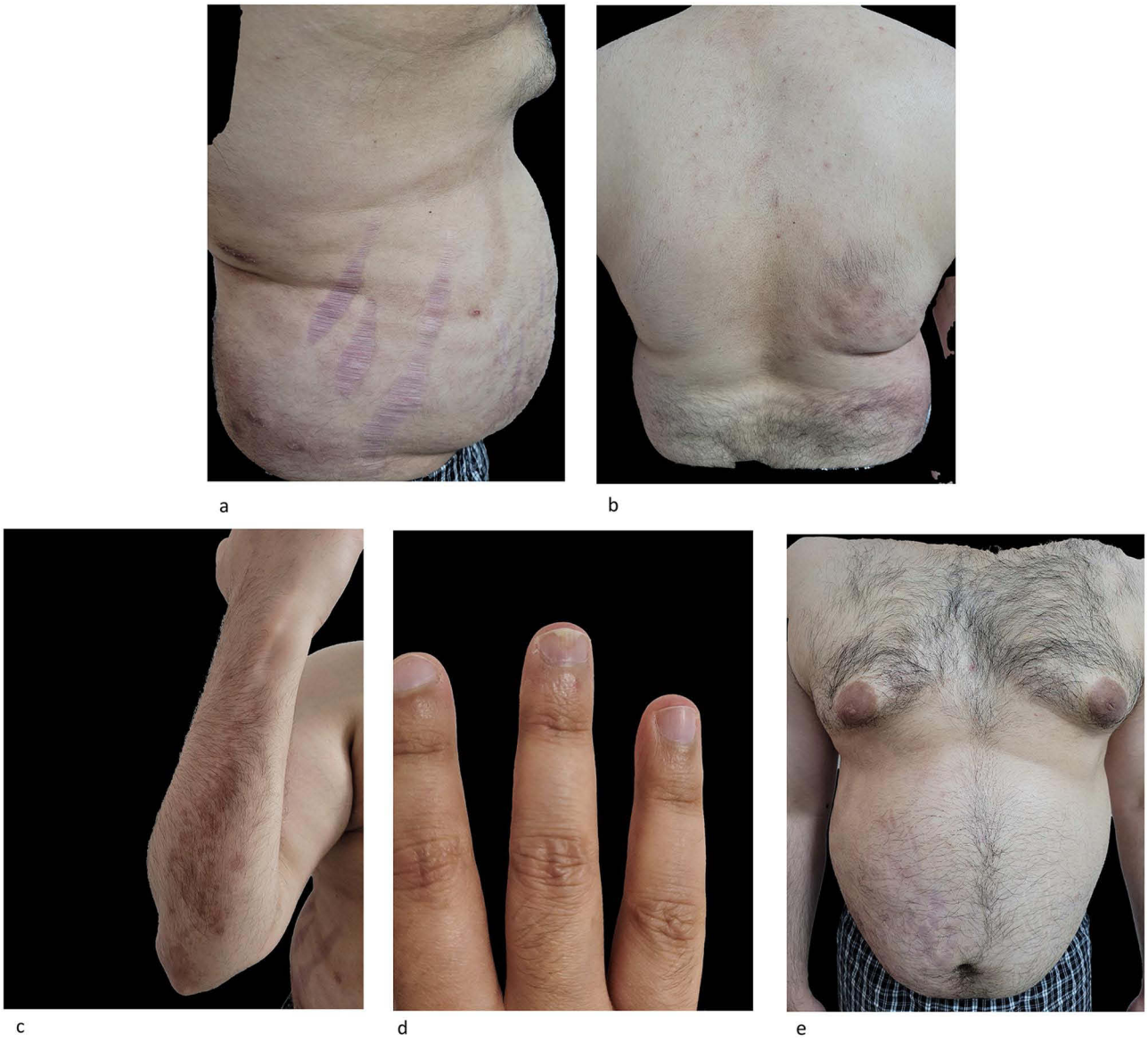

Figure 4 Full body and close-up images of the patient under treatment with Ixekizumab. (**a**, **b**) Posterior view and right side of the patient showing discrete postinflammatory hyperpigmentation, (**c**) right forearm of the patient showing discrete postinflammatory hyperpigmentation, (**d**) close up image of the right hand showing discrete oil-drop sign on nail, (**e**) frontal view of the patient without signs of psoriasis.

Abbreviations

BSA, Body Surface Area; DLQI, Dermatology Life Quality Index; EASI, Eczema Area and Severity Index; ILVEN, Inflammatory Linear Verrucous Epidermal Nevus; LP, Linear Psoriasis; PASI, Psoriasis Area and Severity Index.

Ethical Statement

Complete written informed consent was obtained from the patient for the publication of this case report and accompanying images. Patient confidentiality has been maintained throughout the study.

Patient Consent

Complete written informed consent was obtained from the patient for the publication of this study and accompanying images.

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Disclosure

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