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A Case of Linear Cutaneous Lupus Erythematosus in a 55-Year-Old Woman

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Study Design A
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Conflict of interest: None declared

Patient: Female, 55-year-old
Final Diagnosis: Linear cutaneous lupus erythematosus
Symptoms: Pruritic papular eruption
Medication: —
Clinical Procedure: —
Specialty: Dermatology

Objective: Rare disease
Background: Linear cutaneous lupus erythematosus (LCLE) is uncommon and occurs mainly in children and young adults. To our knowledge, only ten cases of LCLE in adults have been previously reported. A case is presented of LCLE of the left arm in a 55-year-old woman.

Case Report: A 55-year-old Caucasian woman from the Midwestern United States presented with a three-month history of a pruritic linear eruption on the left arm. She had a previous history of methicillin-resistant *Staphylococcus aureus* (MRSA) infection of the left forearm. She had previously been treated with topical triamcinolone, hydrocortisone cream, hydroxyzine, and two courses of prednisone. Physical examination showed a unilateral and linear erythematous skin lesion of the left arm that contained papules and followed the embryonal developmental epidermal lines of Blaschko. Histopathology of a 4 mm skin punch biopsy showed an interface dermatitis with keratinocyte necrosis and increased dermal mucin. Immunofluorescence of the skin biopsy, including for antinuclear antigen (ANA), was negative. Prednisone treatment reduced the symptoms of pruritis but did not resolve the rash. However, following topical treatment with betamethasone dipropionate cream for between two and three weeks, and the use of sunblock, the skin lesions resolved.

Conclusions: This rare case of LCLE in an older adult showed a similar response to treatment as other forms of cutaneous lupus erythematosus, with treatment that included topical steroids and sun protection. Also, this case supports that environmental trigger factors, such as prior infections, might provide insights into the etiology of LCLE.

MeSH Keywords: Autoimmune Diseases • Dermatology • Diagnosis • Lupus Erythematosus, Cutaneous • Treatment Outcome

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Background

Linear cutaneous lupus erythematosus (LCLE) is a rare condition that was first described by Abe et al. in 1988 [1]. LCLE mainly occurs in children and young adults. The skin lesions appear as unilateral linear erythematous inflammatory papules and plaques following the embryonal developmental epidermal lines of Blaschko, mainly on the face, neck, trunk, and extremities [2]. To our knowledge, only ten cases in adults have previously been reported, and all previously confirmed cases of LCLE followed the Blaschko lines [2–10]. A case is presented of LCLE of the left arm in a 55-year-old woman.

Case Report

A 55-year-old Caucasian woman from the Midwestern United States, who worked in a warehouse, presented with a three-month history of a pruritic linear eruption on the left arm. The pattern of the skin lesion followed the embryonal developmental epidermal lines of Blaschko. She had previously been treated with triamcinolone, hydrocortisone cream, hydroxyzine, and two courses of prednisone. Prednisone reduced the pruritis but did not resolve the rash. She denied fever, chills, or photosensitivity, and had no history of injury or increased

exposure to sunlight. She had a previous history of methicillin-resistant *Staphylococcus aureus* (MRSA) infection of the left forearm but had no previous history of autoimmune disease. She had no family history of autoimmune disease.

Physical examination showed a unilateral linear and erythematous papular rash that followed the embryonal developmental epidermal lines of Blaschko on the left upper arm, extending down the left side (Figures 1, 2). Histology of a 4 mm punch biopsy of the skin showed an interface dermatitis with keratinocyte necrosis (Figure 3) and dermal mucin deposition (Figure 4). The histology was consistent with a diagnosis of dermal lupus erythematosus. Direct immunofluorescence for IgG, IgM, IgA, and C3 performed two weeks later were negative. Antinuclear antigen (ANA) was also negative. A diagnosis of linear cutaneous lupus erythematosus (LCLE) was made.

The differential diagnosis in this patient included acquired inflammatory dermatoses with a distribution pattern along the developmental epidermal lines of Blaschko, including lichen striatus, linear lichen planus, linear morphea, linear granuloma annulare, linear psoriasis, or linear verrucous epidermal nevus. The treatment of localized LCLE includes topical steroids and sunblock, but the widespread disease may be treated with antimalarials [2]. In this case, the patient was treated

Figure 1. A 55-year-old Caucasian woman with linear cutaneous lupus erythematosus (LCLE). The appearance of the left arm shows the appearance of LCLE in the distribution of the embryonal developmental epidermal lines of Blaschko.

Figure 2. A 55-year-old Caucasian woman with linear cutaneous lupus erythematosus (LCLE). The appearance of the left arm shows the appearance of LCLE in the distribution of the embryonal developmental epidermal lines of Blaschko.

Figure 3. Photomicrograph of histology from the skin biopsy from the left arm of a 55-year-old Caucasian woman with linear cutaneous lupus erythematosus (LCLE). The histology shows an interface dermatitis with vacuolar change of the basal epidermal cells (single arrow) with keratinocyte necrosis (double arrow). Hematoxylin and eosin (H&E). Magnification, $\times 10$.

Figure 4. Photomicrograph of histology from the skin biopsy from the left arm of a 55-year-old Caucasian woman with linear cutaneous lupus erythematosus (LCLE). The histology shows dermal acidic mucin (blue) (single arrow). Colloidal iron stain. Magnification, $\times 10$.

with topical betamethasone dipropionate cream for between two to three weeks and with sunblock, which resulted in the resolution of the skin lesions. However, after this time, the patient was lost to follow-up.

Discussion

Cutaneous forms of lupus erythematosus (LE) include acute cutaneous lupus, subacute cutaneous lupus, and chronic cutaneous lupus, which is also known as discoid lupus erythematosus (DLE) [11]. Linear cutaneous lupus erythematosus (LCLE) is the linear variant of DLE. However, as LCLE is a rare condition, the prevalence of the association with serum antinuclear antibodies (ANAs) remains unknown. However, in LCLE, negative findings for autoantibodies are common, while dermal immunofluorescence results are more commonly positive [4].

Jin et al. [12] proposed that the pathogenesis of LCLE may involve the presence of mosaicism of the skin cells located along the developmental epidermal lines of Blaschko during

embryogenesis, and in their cellular, molecular, and epigenetic pathogenesis. If this theory is correct, it might be important to explore the relationship between genetic and environmental factors LCLE in the future [12]. Happle et al. [13], and Seitz et al. [14] suggested that trigger factors for LCLE might include infections, drugs, pesticides, or heavy metals.

Conclusions

This report presented a case of linear cutaneous lupus erythematosus (LCLE), a condition that is more common in children and young adults, in the left arm of a 55-year-old woman. This patient had a history that included several possible trigger factors for LCLE, including a previous history of methicillin-resistant *Staphylococcus aureus* (MRSA) infection of the left arm and worked in a warehouse work environment. This case may provide support for the possible etiological factors associated with LCLE presenting in older patients.

Conflict of interest

None.

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