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Exclusive extragenital lichen sclerosis in a child presenting in a lichen planus distribution

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Abstract

Lichen sclerosis (LS) is a chronic inflammatory dermatosis primarily affecting the genitalia, commonly characterized by pearly-white papules and plaques. Although predominantly affecting females, LS can manifest across all age groups, with a bimodal distribution observed in prepubescent girls and postmenopausal women. This case report presents an unusual instance of exclusive extragenital LS in a 10-year-old girl, showcasing hyperpigmented patches and wrinkled plaques resembling lichen planus on her forearms and lower legs. Histopathological analysis confirmed LS, revealing distinctive epidermal changes and lymphocytic infiltrates. The absence of mucosal involvement and unique clinical presentation differentiated this case from typical LS manifestations. Treatment with topical clobetasol propionate demonstrated significant improvement in pruritus. Extragenital LS is infrequent, particularly among children, and its diagnosis necessitates a comprehensive clinicopathological correlation. The reported case contributes valuable insights into this uncommon variant, emphasizing the importance of accurate diagnosis and tailored treatment strategies. Additionally, it highlights the efficacy of high-potency topical corticosteroids in managing this condition.

Keywords: corticosteroid, hyperpigmented patches, lichen planus, pediatric dermatology, sclerosis, topical

Introduction

Lichen sclerosis (LS) is a long-standing inflammatory skin condition that presents with whitish plaques distributed most commonly in the genital area [1-3]. Although the exact cause remains unclear, hereditary and autoimmune factors have been suggested as possible contributors [1-3]. Lichen sclerosis can affect individuals of any age, with females being 10 times more likely than males to experience it [1-4]. The condition shows a bimodal distribution, with the highest prevalence observed in prepubescent girls and postmenopausal women [1]. Lichen sclerosis is considered one of the most prevalent conditions treated in dermatology clinics, with a documented frequency ranging from one in 300, to one in 1000 women, and one in 900 girls [5, 6]. However, it is important to note that LS may be under-reported and under-recognized [5,6]. Among all LS cases, only 5-15% are estimated to occur in children, with postmenopausal women accounting for a large portion of cases [6-9]. Extragenital involvement of LS is observed in only about 6% of these patients, and in pediatric patients, it invariably affects the genital region [10]. Girls are affected by this condition more frequently than boys, with a female-to-male ratio as high as 10:1 [10,11].

Case Synopsis

A 10-year-old girl visited the dermatology clinic with complaints of multiple brownish wrinkled plaques

on her forearms and lower legs that had been bothering her for the past three years. The plaques were severely itchy and had gradually increased in size. They initially appeared on the volar aspect of her distal forearms bilaterally and then spread to the medial and lateral aspects of her lower legs bilaterally (**Figure 1**). The patient had no personal or family history of dermatological or autoimmune diseases and no mucosal involvement; she had not used any new medications. She had only used low-potency topical corticosteroids which did not improve her symptoms.

During the physical examination, multiple brownish wrinkled plaques with minimal scaling and central ivory areas were observed on the volar aspect of her distal forearms and the medial and lateral aspects of her lower legs bilaterally. There were no lesions on the mucosal surfaces or any other skin abnormalities.

A 4mm skin punch biopsy was taken from one of the plaques. H&E histopathology showed orthokeratotic hyperkeratosis and epidermal thinning (**Figure 2A**). The usual ridge pattern of the skin was lost and there was reduced cellularity in the papillary dermis, along with increased collagen density. Lymphocytic infiltration was visible in certain areas of the dermoepidermal junction and around blood vessels. Further observations include follicular plugging, papillary dermal edema, and homogenization of collagen bundles. Elastin van Gieson stain showed marked elastic fiber diminishment in the papillary dermis (**Figure 2B**).

Based on the clinical and histopathological findings, the patient was diagnosed with exclusive extragenital lichen sclerosis in a lichen planus-like distribution. Topical clobetasol propionate was used twice daily for two weeks followed by frequency



Figure 1. Multiple hyperpigmented wrinkled plaques with minimal scaling and central ivory sclerotic over the volar aspect of the forearms bilaterally, and over medial and lateral aspects of the lower legs bilaterally.

tapering. On a follow-up visit, the patient reported significant improvement in pruritus.

Case Discussion

Lichen sclerosis is a chronic inflammatory skin condition primarily affecting the anogenital region [1-3]. In children, LS almost always involves the anogenital area and rarely extends extragenitally [1-3]. A systematic review of 4516 cases of children with LS found that 97.2% of female patients had some degree of anogenital involvement, whereas only 2.8% had extragenital manifestations [12].

Lichen sclerosis can occur in individuals of all genders and age groups, but it most commonly affects women in their fifth and sixth decades. Children make up 7% to 15% of LS cases [13]. The bimodal distribution of LS incidence in prepubertal girls and postmenopausal women is believed to be associated with low estrogen physiological conditions. However, the exact role of sex hormones in the onset of LS remains unknown [14,15].

Little is currently understood about the precise pathophysiology of LS, but it is hypothesized that an autoimmune mechanism combined with a hereditary predisposition may be responsible for its development [16]. Research has shown that up to 80% of LS patients have circulating autoantibodies against extracellular matrix protein one [17,18]. Additionally, there is a documented association between LS and other autoimmune disorders like

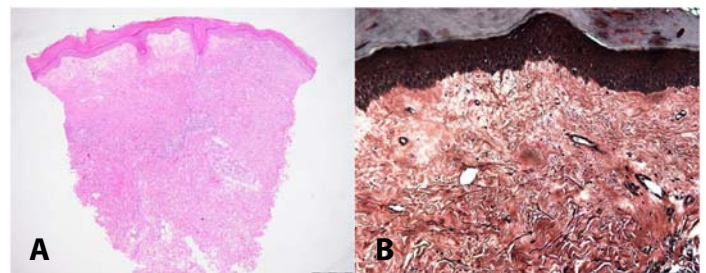


Figure 2. A) A low power view showing orthokeratotic hyperkeratosis, epidermal atrophy with loss of the normal rete ridge pattern, and hypocellular papillary dermis with condensation and homogenization of collagen bundles. Additionally, a lymphocytic infiltration is seen at the dermoepidermal junction and in perivascular distribution. H&E, 40x. B) Elastin van Gieson stain of the papillary dermis shows marked diminishment of elastic fibers, 200x.

alopecia areata, autoimmune thyroid disease, and pernicious anemia [17,18].

An intriguing finding from a study involving 532 LS patients is that female patients tend to have a higher prevalence of autoimmune conditions and elevated antibody levels compared to male patients [17]. This suggests a possible sex-related difference in the expression or susceptibility to autoimmune responses in LS. However, further investigation is required to fully understand the complex interplay between autoimmune factors and LS development [17].

In this study, we present a 10-year-old girl who presented with several brownish wrinkled plaques on the volar aspect of the distal forearms bilaterally and on the medial and lateral aspects of the lower thighs bilaterally. These plaques were covered with minor scaling. Unlike the findings in two other reports, one involving a 6-year-old girl with very thin, pink-to-white macules and minute plaques with micaceous overlying scale and slight atrophy on the chest and back, and the other study involving a nine-year-old girl with asymptomatic, vaguely defined erythematous lesions on her thighs and trunk, our patient showed no involvement of mucosal surfaces and had no other skin lesions over a period of three years [19,20].

Extragenital LS exhibits distinct characteristics compared to anogenital involvement. In children, anogenital LS typically presents painful, intensely itchy, white or red patches, which can lead to secondary issues such as dysuria, constipation, and behavioral problems [6,15]. Conversely, extragenital lesions are usually asymptomatic. Although extragenital LS can appear anywhere on the body, it commonly affects areas like the trunk, neck, shoulders, and wrists. These lesions often start as pink-to-ivory white macules or papules that may eventually merge to form glossy, atrophic plaques over time [14,21].

In our patient's case, she had multiple wrinkled, brownish plaques, gradually increasing in number, that had been present for three years, causing extreme itching. Although less common, extragenital LS can also involve the oral mucosa and

nails [21,22]. It is important to note that other conditions, such as irritating contact dermatitis, atrophic lichen planus, vitiligo, psoriasis, and morphea, may be misdiagnosed as extragenital LS [20]. Therefore, accurate diagnosis and differentiation are crucial for appropriate management and treatment.

In our patient's case, the treatment involved using topical clobetasol propionate for two weeks. On a follow-up visit, she reported a marked improvement in pruritus. She was maintained on the same management plan and will be monitored in the clinic for further evaluation. This treatment approach differs from a previous report by Stavrianeas et al. in which initially a potent topical corticosteroid cream was used without improvement at four weeks. As a result, the topical corticosteroid therapy was discontinued and treatment was changed to topical pimecrolimus twice daily for 6 months. One month after starting pimecrolimus treatment, the skin lesions showed significant regression of the erythematous and sclerotic components [19]. It is essential to tailor the treatment plan based on individual patient responses, as the effectiveness of different medications can vary from person-to-person. Regular follow-up and careful evaluation of the patient's response to treatment are crucial for determining the most appropriate course of action. It is worth noting that our findings align with previously reported cases of extragenital LS in the existing literature [12,16,22].

Conclusion

Extragenital LS without involving the anogenital region is a rare entity, especially in children. Diagnosing this condition requires clinicopathological correlation to ensure accurate identification. Treatment for extragenital LS is similar to the anogenital subtype and high-potency topical corticosteroids are commonly used. The presentation of this exclusive extragenital LS case in a pediatric patient contributes to a better understanding of diagnostic and treatment approaches for this uncommon variant of the disease.

Potential conflicts of interest

The authors declare no conflicts of interest.

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