

# A Rare Case of Exclusively Extragenital Lichen Sclerosus in a Child

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## Abstract

**Background.** Lichen sclerosus (LS) is a chronic, inflammatory skin condition of unknown etiology that shows a predilection for the anogenital region. Although LS primarily affects postmenopausal women, some cases do occur in children. However, in the pediatric setting, LS almost always affects the anogenital region with exclusively extragenital involvement very rarely diagnosed in childhood. Here, we present a 6-year-old girl with a 2-year history of thin, pink macules and plaques on her chest and back, which biopsy revealed to be extragenital LS. A review of this case and the literature describes the epidemiology, clinical features, pathophysiology, and treatment options for pediatric extragenital LS.

**Key words:** Lichen sclerosus, pediatric dermatology, skin biopsy, dermatopathology

Lichen sclerosus (LS) is a poorly understood, chronic, inflammatory skin condition that primarily affects the anogenital region.<sup>1,2</sup> While LS has been reported in all age groups and both sexes, it primarily affects women in the fifth to sixth decades of life with only 7% to 15% of cases occurring in children.<sup>1,3</sup> In the pediatric setting, LS almost always affects the anogenital region with exclusive extragenital involve-

ment very rarely diagnosed in childhood.<sup>4</sup> Here, we present a case of extragenital LS without anogenital involvement in a pediatric patient in order to raise awareness of this rare disease subtype.

### Case Presentation

A 6-year-old white girl with a history of mild eczema presented to the dermatology clinic because of a 2-year history of a

nonpruritic, nonpainful rash on her back.

### History

The patient had a family history of allergies and asthma in both her mother and father but no other family history of skin conditions, nail abnormalities, or autoimmune disease. She had previously been treated with topical hydrocortisone and over-the-counter antifungal cream without relief. The rash did resolve with use of triamcinolone 0.1% ointment but recurred whenever use of the ointment was discontinued.

### Physical examination

Physical examination was notable for very thin, pink to whitish macules to minute plaques with micaceous overlying scale and slight atrophy on the patient's chest and back (**Figure 1**). A solitary pit was also observed on the right thumbnail. The patient's scalp, ears, genitals, and perianal area did not display signs of rash.

### Differential diagnosis

The initial differential diagnosis includ-

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### DISCLOSURES:

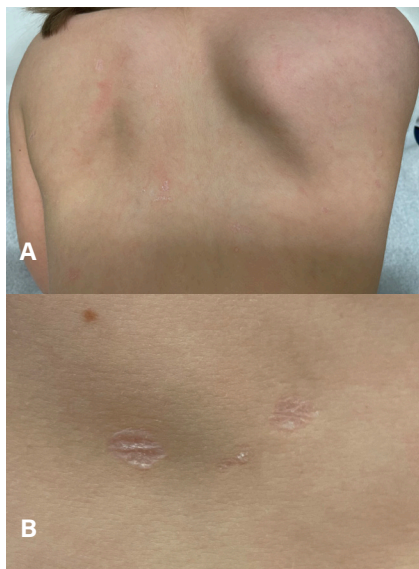
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The authors report no relevant disclaimers.

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**Figure 1.** Thin, pink to whitish macules to minute plaques with micaceous overlying scale and slight atrophy scattered along the patient's (A) back and (B) chest.

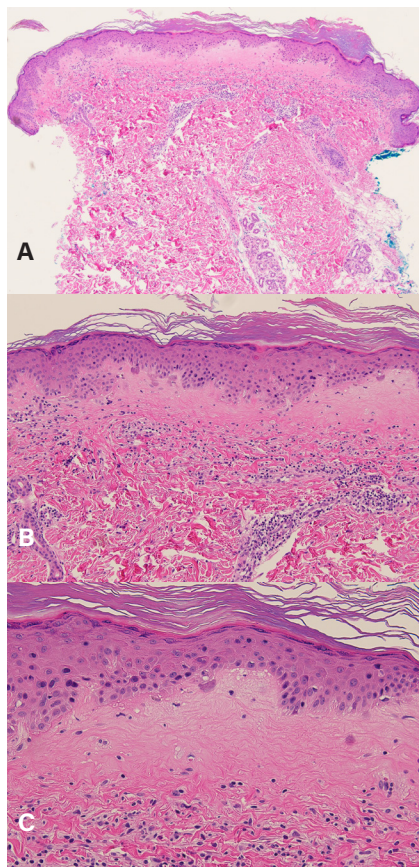
ed extragenital lichen sclerosis (LS), atrophic lichen planus, unusual psoriasis, and pityriasis lichenoides chronica. Discoid lupus erythematosus and Degos disease were also considered, although these were lower on the differential.

### Diagnostic studies

Because of the uncertainty of the diagnosis, a punch biopsy was performed on one of the lesions on the right upper back. The biopsy revealed a slightly atrophic epidermis with vacuolar basal keratinocytes and rare dyskeratosis (**Figure 2**). The superficial dermis was notable for a hyalinized stroma and band-like lymphocytic infiltrate below the fibrosis, prototypical features seen in LS (**Figure 2, panels B and C**). There was no increase of dermal mucin, periadnexal lymphocytic infiltrate, or basement membrane thickening seen with hematoxylin and eosin staining. The histopathology in conjunction with the physical examination findings confirmed a diagnosis of extragenital LS.

### Patient outcome and follow-up

Topical clobetasol 0.05% ointment daily was initiated. Significant improvement was seen after 6 weeks, after which the



**Figure 2.** Hematoxylin and eosin histologic staining of a punch biopsy taken from a lesion on the upper right side of the patient's back, shown at (A) 4x magnification, (B) 10x magnification, and (C) 20x magnification, and demonstrating a slightly atrophic epidermis with prominent interface changes, including vacuolar basal keratinocytes and rare dyskeratosis. The superficial dermis demonstrated a hyalinized stroma and a band-like lymphocytic infiltrate below the fibrosis, prototypical features seen in lichen sclerosis.

clobetasol was reduced to weekends only. The patient's mother was instructed to watch the patient's genital area very carefully and to report any new genital involvement.

### Discussion

LS is a chronic, inflammatory skin condition that primarily affects the anogenital region. The true prevalence of LS is unknown because patients may be asymptomatic, and they often present to a variety of specialists, including dermatologists, pediatricians, gynecologists, and

urologists.<sup>5,6</sup> However, the prevalence of LS has been estimated to range from 1 case in 70 to 1 case in 1000 person-years in women and 1 case in 900 person-years in prepubertal girls.<sup>7,8</sup> While LS has been reported in all genders and age groups, it primarily affects women in the fifth to sixth decades of life, with 7% to 15% of cases occurring in children.<sup>3</sup> The bimodal distribution of incidence in prepubertal girls and postmenopausal women is thought to be correlated with low estrogen physiological states; however, the role of sex hormones in the development of LS remains unclear.<sup>5,9</sup> In the pediatric setting, LS almost always affects the anogenital region with exclusive extragenital involvement very rarely diagnosed in childhood. A systematic review of 4516 identified cases of childhood LS found 97.2% of female patients had some degree of anogenital involvement while only 2.8% had exclusively extragenital disease.<sup>4</sup>

While the pathogenesis of LS is poorly understood, it is postulated that LS may result from an autoimmune etiology in conjunction with genetic predisposition.<sup>10</sup> Circulating autoantibodies to extracellular matrix protein 1 have been reported in up to 80% of LS patients, and there is a known association between LS and other autoimmune diseases, such as alopecia areata, autoimmune thyroid disease, and pernicious anemia.<sup>11,12</sup> Interestingly, a study of 532 patients with LS showed that female patients present with autoimmune disease and increased autoantibody levels significantly more often than their male counterparts.<sup>12</sup>

The presentation of extragenital LS differs significantly from anogenital disease. In children, anogenital involvement classically presents with painful and intensely pruritic white or red patches that can lead to secondary dysuria, constipation, and behavioral problems.<sup>2,5</sup> Extragenital lesions, on the other hand, are usually asymptomatic. While these lesions can occur anywhere on the body, extragenital LS usually involves the trunk, neck, shoulders, and wrists. The typical lesions of extragenital LS are pink to ivory white

macules or papules that can coalesce into shiny, atrophic plaques over time, as seen in this case.<sup>6,9</sup> Oral mucosal and nail involvement have also been reported but are much more rare.<sup>6,8</sup> Other diagnoses that may mimic extragenital LS include irritant contact dermatitis, atrophic lichen planus, vitiligo, psoriasis, and morphea.

The treatment of extragenital LS is the same as that for the anogenital form. Because LS is a chronic, relapsing-remitting disease, the goal of treatment is to relieve symptoms and minimize any scarring or cosmetic sequelae. Mild cases of extragenital LS are generally responsive to high-potency topical corticosteroids, which is the first-line treatment for all forms of LS. As demonstrated in this case, lower dose corticosteroids, such as hydrocortisone, may not be sufficient. For high-dose steroid-resistant cases, other treatment options include topical calcineurin inhibitors, such as tacrolimus or pimecrolimus cream, tangential excision, carbon dioxide laser therapy, and various forms of phototherapy.<sup>13,14</sup>

### Limitations

This review is a retrospective case report reflecting the clinical presentation and treatment of a single patient. It is therefore inherently limited in its generalizability; however, our findings are in line with previous reports of extragenital LS.<sup>4,6,10</sup>

### Conclusion

Extragenital LS in the absence of anogenital involvement is rare, especially in the pediatric population. A biopsy and examination of the histopathologic findings may be necessary to make the diagnosis. Treatment of extragenital LS is the same as that of the anogenital subtype, mostly relying on high-potency topical steroids. This case of exclusive extragenital LS in a pediatric patient is presented to raise awareness of the diagnostic and therapeutic approach to this rare disease subtype.

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