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## A Rare Presentation of Vulvar Lichen Sclerosus in a Pediatric Patient: A Case Report and Review of the Literature

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### ABSTRACT

Lichen Sclerosus is a benign, chronic dermatological condition characterized by marked inflammation, epithelial thinning, lymphocytic infiltration, and symptoms of vulvar pruritus, labial adhesion, and pain. There are two peaks of onset, one in low estrogen state - among prepubertal girls and the other in peri or postmenopausal women. We present a rare case of lichen scleorsus in a pediatric patient, emphasizing the importance of recognizing this condition in the pediatric population.

Seven y/o female presented with a 6-month history of difficulty voiding, severe pruritus, and vulvar pain. Clinical examination showed white atrophic vulvar papules, partial labial fusion, and purpuric plaque in the anogenital region. Histological examination confirmed the diagnosis of lichen sclerosus. The patient was treated with topical corticosteroids and started on maintenance therapy to reduce recurrence and minimize the risk of vulvar carcinoma. Although rare in children, lichen sclerosus should be considered in diagnosing pediatric genital lesions. Given its high risk of recurrence and increased risk of vulvar carcinoma, long-term complications must be avoided by early detection and prompt management.

#### Keywords

Lichen sclerosus, Pediatric population, Vulvar pain, Pruritus, Labial adhesions, Vulvar squamous cell carcinoma (vSSC), Vulva brush, Biopsy, Histological examination, Topical corticosteroids.

#### Introduction

Pediatric lichen sclerosus is a rare inflammatory dermatologic condition that is predominantly seen in postmenopausal women, but 7-15% of cases have been reported in prepubertal females [1]. Although most cases are asymptomatic, it is estimated that 1 in 900-1100 of this age group is affected [2]. While the exact pathogenesis of LS is unknown, increasing evidence suggests that the autoimmune mechanism plays a crucial role in its onset and progression. It has been observed that in premenarchal girls, LS can be linked with various autoimmune disorders such as thyroiditis, pernicious anemia, psoriasis, vitiligo, morphea, and alopecia areata [3]. Familial cases have been reported with no clear inheritance pattern in identical and non identical twin pairs

as well as siblings [1,4]. Apart from these, several other factors are being investigated to understand the pathogenesis of LS including immunocytologic alterations, sex hormone factors, trauma, and connective tissue alterations [3,5]. Additionally, LS could also be a result of the Koebner phenomenon, which is the emergence of a lesion due to skin injury [5]. This condition frequently affects the anogenital region, with patients commonly complaining of genital symptoms such as vulvar pain and pruritus, burning sensation, vaginal bleeding, constipation, and dysuria [2,6,7]. Itching can be so intense that it leads to skin tearing and bleeding. Physical exam findings in pediatric LS often resemble those seen in adults, such as white plaques and papules, labial adhesions, and characteristic perineal lesions causing the classic "figure 8" shape [5,8]. Erosions, blisters, scars, adhesions, and bruises may also be observed. Potential long-term complications include genital scarring and atrophy, behavioural and psychological changes, and most severe vulvar squamous cell cancer (SCC) [9]. Histologically, thinning of the epidermis, hyperkeratosis, and lymphohistiocytic infiltration

characterize VLS. Therefore, the case report aims to highlight and create awareness among healthcare workers about the rising cases of LS in the pediatric population and its consideration as a differential diagnosis in patients presenting with similar genital lesions.

#### **Case Report**

A seven y/o female presented with a 6-month history of intense pruritus, vulvar pain, and difficulty voiding. No significant medical or family history was noted, and there were no allegations of trauma or abuse. On any other skin surface, there were no lesions found. Upon genital examination, the following findings were noted: a purpuric plaque in the perianal region, partial labial fusion, white atrophic papules in the upper vulva, and an intact hymen.

Observe Figure 1.



Figure 1: Genital examination findings.

Due to the similarity in clinical presentation between vulvovaginal candidiasis and lichen sclerosus, lichen sclerosus was suspected when the patient's symptoms persisted despite therapy. However, additional differential diagnoses, such as dermatitis, mucous membrane pemphigoid, lichen planus, lichen simplex chronic, and estrogen insufficiency, which tend to mimic LS, should be ruled out. The European Academy of Dermatology and Venerology's evidence-based guidelines suggest that a diagnosis can be made based on characteristic clinical features, with biopsy reserved for children with atypical manifestations. Due to the uncertainty surrounding the diagnosis and the possible cancer risk linked to LS, a vulvar biopsy was performed to look for atypical cells. To avoid invasive procedures, we used the less invasive Vulva Brush (Rovers Medical Devices BV) diagnostic tool. Although the cytological smear analysis revealed the presence of a few atypical cells, the probability of malignancy was uncertain based on the Cytological Smear Classification as shown in Table 1. Therefore, we proceeded with a punch biopsy since, in a comparatively short amount of time, an asymptomatic recurrence can induce scarring and architectural damage. However, a biopsy is not required if no aberrant cells are discovered [10].

Table 1: Classification	of Cytologic Smears.
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Suspicious for (pre)malignancy			
Normal	Uncertain	Favour uVIN	Favour dVIN
No atypical cells or dysplastic cells	Some atypical cells	Evident Dyskaryotic cells and cell groups. Increased N/C ratio, irregular coarse chromatin, irregular nuclear membrane, Koilocytes.	Large atypical cells often isolated , Eccentric nuclei, Prominent nucleoli, Absence of koilocytes.

Abbreviations - dVIN: Differentiated vulvar intraepithelial neoplasia, N/C ratio: Nucleo-cytoplasmic ratio, uVIN: Usual vulvar intraepithelial neoplasia.

Cytology of the vulva: Feasibility and preliminary results of a new brush - Scientific Figure on ResearchGate. Available from: https://www.researchgate.net/figure/Classification-of-cytological-smears-Suspicious-for-premalignancy\_tbl1\_51846822.

Epidermal atrophy, hyperkeratosis, and dermal lymphohistiocytic infiltrate were observed on Vulvar punch biopsy, confirming the diagnosis of lichen sclerosus. Observe Figure 2.



Figure 2: Vulva punch biopsy.

Epidermal atrophy, hyperkeratosis, and dermal lymphohistiocytic infiltrate. (H&E-stained, x20)

#### Discussion

Lichen Sclerosus recurrence rate in prepubertal girls after medical therapy is reported to range from 44%-82 % [11,12]. Remission was achieved in 83% of cases after high potency topical steroid, while relapse occurred in 44%. While it was previously believed that LS in childhood would naturally resolve at puberty, recent findings suggest that many children may continue to experience

symptoms even after puberty, making early treatment highly advisable whenever possible [13].

Topical corticosteroids are commonly used as first-line agents. In this case, the patient was prescribed clobetasol propionate 0.05% ointment following a specific regimen. The treatment started with applying the ointment every night for four weeks, followed by every other night for four weeks, and then twice a week for four weeks. The entire treatment period required a 30g tube of ointment, with a maximum of 10g applied per month. The patient was revaluated after the sixth week of therapy, and we noticed that the patient experienced relief from pruritus, dysuria, and pain. After the treatment was completed, no symptoms or vulvar lesions were present. To minimize the chances of symptoms recurring, the patient was prescribed a maintenance regimen of clobetasol ointment, to be applied twice a week, which was found to be effective in maintaining standard skin color and texture and reducing the incidence of vulvar intraepithelial neoplasia and vulvar cancer. The patient was revaluated after 12 weeks, and no symptoms or lesions were present. The patient was offered longterm follow-up and examination at least once a year to exclude long-term complications.

#### Conclusion

The case report highlights the challenges faced in diagnosing lichen sclerosus in the pediatric population due to its rarity and non-specific symptoms that overlap with other skin conditions. Its potential association with autoimmune conditions as well as its long-term risk of malignancy warrants early recognition and management to minimize complications and improve their overall prognosis.

#### Additional Information Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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