

REVIEW PAPER

When a gynecologist cures constipation: lichen sclerosus in prepubertal and adolescent girls

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ABSTRACT

Lichen sclerosus (LS) is a chronic inflammatory mucocutaneous disease that primarily affects the anogenital region. It is often observed in prepubertal girls. Typical symptoms include vulvar irritation and pain, itching of the vulva, difficulty urinating, bleeding due to skin fissures, painful defecation, and constipation. The diagnosis is primarily clinical, based on the patient's history and the results of a physical examination. Visual inspection of the anogenital region is an essential part of the diagnostic procedure. This is particularly important when patients complain of constipation. Typical lesions include ivory-white plaques, skin fissures, and superficial erosions. These often create a classic "figure of eight" shape when they are located in the vulva and perianal region. Initial therapy for LS involves applying high-potency topical steroids. Calcineurin inhibitors should be considered as second-line agents. Emollients are used as a supplement or in maintenance therapy for vulvar LS. Long-term follow-up is recommended.

KEY WORDS:

lichen sclerosus, prepubertal girls, constipation, anogenital dermatoses.

INTRODUCTION

Lichen sclerosus (LS) is a chronic inflammatory mucocutaneous disease of unknown etiology. About 85% of total LS cases are located in the anogenital region. The disease was known by multiple names, including LS et atrophicus and kraurosis vulvae, until the International Society for the Study of Vulvovaginal Disease in 1976 adopted the term LS [1].

Lichen sclerosus occurs in both sexes. The male-to-female ratio varies between 1 : 3 and 1 : 10 [2]. Lichen sclerosus can occur at any age, although the highest values can be observed in women aged 40–60 years old, and in pre-pubertal girls. There is a clear peak of incidence in girls aged 4–6 years old, which represents 7–15% of all LS cases [3]. It is estimated that LS can be observed in 1 : 900–1100 of premenarchal girls [1].

The initial symptoms of LS are typically non-specific, making it susceptible to misdiagnosis by doctors who are not specialized in gynecology or dermatology. In some cases, symptoms may spontaneously resolve after menarche, leading to a latent course of the disease. This contributes to the underestimation of the epidemiology of LS [4].

This article aims to emphasize the importance of examining the anogenital area of girls with constipation, and to present a narrative literature review on LS.

ETIOLOGY

The exact pathogenesis of LS remains unclear and is complex. It is generally accepted that LS is an autoimmune disorder. The condition has been associated clinically and immunologically with autoimmune thyroiditis, pernicious anemia, vitiligo, morphea, and alopecia areata [1]. Other

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TABLE 1. Proper anogenital hygiene rules

Proper anogenital hygiene
• Washing with water, possibly an emollient • Parental supervision in the toilet: – correct direction of rubbing (front to back) – the position for urination is with legs wide apart and feet on the floor or stool • Avoid irritants – bath salts, strongly scented soaps/gels, moistened toilet tissues • Laundry in hypoallergenic detergents, double rinsing, avoidance of softeners • Cotton underwear, nightdress • Clothes made of natural fibers, loose • Quick change of swimming costume • Menstruating girls – pads with natural (cotton) filling: – frequent change of pads – sanitary pads – not every day

factors contributing to the development of LS include hormonal influences, infections, trauma, and changes in connective tissue. The genetic role in the development of LS is still not well understood; approximately 10% of patients have relatives with the same condition [2].

IMMUNOLOGY

Immunological changes at the level of T- and B-cells have been described among patients diagnosed with LS. Specifically, an autoimmune phenotype has been observed in the case of vulvar LS involving increased levels of Th1-specific cytokines, dense T-cell infiltration, and enhanced BIC/miR-155 expression as well as autoantibodies against extracellular matrix protein 1 and BP180 antigen. The pathogenetic relevance of these observations is unclear. Additionally, oxidative DNA damage and TP53 mutations (tumor suppressor gene) have also been described. This could indicate an autoimmune background of LS and play a role in the slightly increased risk of vulvar carcinoma [5].

HORMONES

The higher incidence of LS in postmenopausal women and prepubertal girls suggests that the hormonal status may be of pathogenetic relevance. However, there are not enough studies to prove this theory. The increased risk of LS in Turner syndrome, despite the use of hormone replacement therapy, points to the disputable role of hormones [6].

TRAUMA

The Koebner response is recognized as one of the most notable manifestations of LS. In the event of a physical injury to the skin, lesions will manifest along the trajectory of the trauma [6]. Consequently, it is hypothesized that recurrent irritation may be a contributing factor to

the development of the disease. Another potential causal factor may be radiation [7].

INFECTION

There is no compelling indication of an infectious trigger of LS. Although there was a considerable interest in *Borrelia burgdorferi* and human papillomavirus (HPV), these theories were later disproven [2]. However, some studies indicate that the bacterial environment of the skin and intestines may contribute to the development of LS. Chattopadhyay *et al.* [8] suggest that there may be a relation between intestinal and cutaneous dysbiosis and the disease in children, although this needs further investigation.

HYGIENE

Improper vulval hygiene can also contribute to the development of the disease. Nonspecific inflammation of the vulva and anogenital area, which is common among girls, favors the development of LS. Therefore, proper hygiene and correct urination (with legs wide apart) can help to reduce the recurrence of lichen (Table 1).

REASONS FOR COUNSELLING

Children with LS may present with a variety of symptoms. Typical complaints include vulvar irritation and pain, vulvar pruritus, dysuria, bleeding due to skin fissures, painful defecation, and constipation. Physicians should pay particular attention to girls who complain of constipation. In such cases, the external genitalia and anal region should always be examined. Lichen sclerosus is quite often the underlying cause of constipation in prepubertal girls. Once treatment has been applied to the skin and mucosa of these regions, the constipation resolves. Dermatoses affecting the vulva and anal area can also lead to pain during defecation, causing children to avoid passing stool. Constipation occurs in 50–67% of girls with LS [9, 10]. If the lesions are more severe around the anterior labia majora, this can result in withholding urine.

The extent of anogenital lesions does not correlate with the intensity of symptoms. Compulsive scratching of the genitalia and grabbing at clothing due to itching may be disturbing to parents and confused with masturbation. The vulvar irritation is often more significant at night. Some parents report that their daughters wake up at night due to vulvar pain, and this is the main reason for the consultation. Older adolescents may present with dyspareunia and lacerations at the base of the posterior fourchette from trauma during intercourse. Furthermore, the anogenital lesions may also be detected incidentally by a parent or physician when the patient is asymptomatic. However, LS is rarely asymptomatic.

Extragenital LS is less common – data vary between 6% and 20% – and can be found often on the torso, e.g., in the submammary area. Extragenital LS is usually asymptomatic and is not associated with an increased risk of malignant transformation [2].

The symptoms of LS can mimic other conditions. That is why it is often misdiagnosed, and the average delay from onset to diagnosis is 1–2 years [1]. Most patients have been treated for vulvovaginal candidiasis and urinary tract infections by the time of diagnosis.

CLINICAL FEATURES

The characteristic clinical appearance of LS is of ivory white- or rose-colored plaques. The plaques can be atrophic with a shiny or crinkled “cigarette paper” appearance or can be thickened due to hyperkeratosis as a result of repeated excoriations. These can lead to ecchymoses, bleeding, and superficial erosions. The superficial erosions can be painful and are at risk for secondary superimposed infection. In the anogenital area, at first, only a slight redness may be seen. White hyperkeratosis, atrophic skin, lacerations, and ecchymosis typically occur later in the course of the disease (Figures 1, 2). The border of the hypopigmented area is often indistinct, and the affected lesion can spread to the perineal and perianal regions, causing a classic “figure of eight” shape. The excessive genital rubbing can lead to tearing the delicate skin, which can result in bleeding. With skin changes, genital scarring may occur, which can result in labial adhesions, a buried clitoris, and narrowing of the vaginal introitus. Lichen sclerosis in the anogenital region is rarely asymptomatic [1].

DIAGNOSTIC PROCEDURES

The diagnosis is mainly clinical, made on the basis of history and physical exam findings. The need for a biopsy is extremely rare. In this case, histology shows thinning of the epidermis, hyperkeratosis, degeneration of the basal layer cells, and dermis fibrosis with perivascular inflammation. It may be helpful to advise testing for other autoimmune pathologies (especially antithyroid antibodies and celiac disease tests) [11]. In some cases, the lesions persist, albeit in an attenuated form, after the menarche and can make it difficult to use tampons or provoke lacerations of the posterior commissure during sexual activity.

Lesions in the course of LS may imitate the clinical symptoms of many dermatoses. The differential diagnosis includes lichen planus, vitiligo, psoriasis, and contact dermatitis (Table 2). Vitiligo is most often misdiagnosed due to hypopigmentation. However, patients with this condition are asymptomatic. Lichen planus is rare in children. Lichen sclerosis is not associated with vaginal mucosa involvement and rarely affects the oral mucosa, whereas



FIGURE 1. Lichen sclerosis – typical features, Patient 1



FIGURE 2. Lichen sclerosis – typical features, Patient 2

lichen planus typically involves the oral and vaginal mucosa. Furthermore, it is crucial to ascertain whether any alterations observed in the anogenital region are indicative of child abuse. The structures that are damaged most often during penetrative sexual abuse are the hymenal membrane, the fossa navicularis, and the fork. Traumatic anogenital injuries heal rapidly, often leaving no trace. In prepubertal girls, hymenal tissue can be completely absent in the suburethral area, approximately from 10–11 o'clock to 1–2 o'clock [12]. Physical signs of sexual abuse can also

TABLE 2. Lichen sclerosus – differential diagnosis

Disorder	Distinguishing features
Vitiligo	Asymptomatic loss of vulvar pigmentation
Lichen planus	Usually involves the oral and vaginal mucosa
Atopic dermatitis	History of allergen exposure, skin sensitivity in another area
Contact dermatitis	Erythema usually in the distribution with pads or diapers
Psoriasis	Often involves knees and elbows; intense erythema with discrete borders and silvery scale
Candidiasis	Very uncommon in hypoestrogenic, immunocompetent patients; erythema with satellite lesions

be bruises, hematomas, abrasions, and grazes. Kellog's review [13] on 36 pregnant adolescents is very interesting: only 2 of 36 girls presented hymenal complete transection of the posterior half. The explanation is that penetration does not always cause visible tissue damage and/or that acute injuries can heal without leaving any sign.

The most common complaints from patients are related to pruritus and discomfort. The presence of skin fissures, whitish discoloration, and constipation in cases of perianal involvement constitutes a core set of diagnostic features. A biopsy of vulvar lesions in children is indicated in three situations: firstly, in the absence of a response to treatment, secondly, in cases where there is a significant uncertainty regarding the diagnosis, and thirdly, if there is any suspicion of a neoplastic lesion [1, 11].

TREATMENT

The goal of the treatment is to alleviate symptoms and resolve signs of atrophy and scarring. The mainstay of therapy includes the use of high-potency topical corticosteroids and topical immune modulators. Vulvar irritation can also be treated with hypoallergenic emollients. Treatment for LS may complement the standard management of constipation, which includes dietary and lifestyle changes, bowel training, and the use of lactulose or macrogols [14].

HIGH-POTENCY CORTICOSTEROIDS

Clobetasol propionate and betamethasone valerate are the most commonly used topical corticosteroids in the treatment of LS. Prolonged use of these medications can result in thinning of the dermis, secondary superimposed infections and, rarely, hypothalamic-pituitary-adrenal axis suppression. Ointments are generally recommended as they are more easily absorbed and cause less contact dermatitis. In rare cases where a girl develops contact dermatitis due to sensitivity to the vehicle in which the steroid is suspended, a physician may consider referring her to a compounding pharmacy to create the steroid with a hypoallergenic vehicle [1]. The initial stage of treatment is depends upon the severity of the condition. The ointment should be applied twice daily for a period of 14 days. After this time, the response

should be assessed and adjustments made if necessary. Once improvement has been achieved, a maintenance treatment plan should be implemented. For this purpose, a high-potency steroid can be applied less frequently, or a medium-potency preparation such as mometasone furoate can be used. The number of applications must be progressively reduced to one per day and then every other day to cover another 2–4 weeks of treatment. Some authors recommend maintenance once a week for a few months to prevent recurrence [11]. Any superinfection should be treated (topical mupirocin). The severity of the primary lesions, the response to the first line of treatment, and its possible side effects all determine the individual treatment plan required for each patient.

IMMUNE MODULATORS

The topical calcineurin inhibitors, pimecrolimus and tacrolimus, have also been used to treat LS. Theoretical advantages include decreased systemic immunosuppression and a reduced risk of atrophic changes to the local skin. However, both medications carry an the Food and Drug Administration black box warning regarding a potential link between long-term use of topical calcineurin inhibitors and an increased risk of skin cancer and lymphoma. Therefore, it is sensible to consider pimecrolimus and tacrolimus as second-line agents. Neither is recommended for use in children under the age of two [1].

SURGICAL TREATMENT

Surgery for LS is usually only considered if there are complications due to adhesions and scarring. Atrophy of the labia minora, scarring of the clitoral hood, and labia/clitoral hood adhesions can lead to urinary tract outflow obstruction, pseudocysts, and dyspareunia. Patients should undergo medical management to the maximum extent possible before surgery, with plans to continue therapy during the recovery phase [1].

EMOLLIENTS

Emollients increase the water content of the skin and improve the skin barrier function, protecting it from attack by noxious substances [15]. Daily use of emol-

lients accelerates the normalization of damaged skin and has been shown to effectively improve mild subclinical inflammation in experiments. Consequently, emollients are used as a supplement and in the maintenance therapy of LS [16].

COURSE OF DISEASE AND PROGNOSIS

RECURRENCE

Lichen sclerosis should be thought of as a chronic condition with possible recurrence of symptoms after appropriate treatment. The recurrence rate of prepubertal LS after medical therapy has been reported to be 44–82% [1]. It was previously thought that the condition would resolve with puberty. Still, recent studies have shown that the symptoms and signs of the disease persist after puberty in the majority (75%) of patients [1].

PSYCHOSOCIAL EFFECTS

Lichen sclerosis can have a significant impact on quality of life, particularly in cases with a high recurrence rate [2]. In daily life, anogenital itching is distressing. Patients' social activities are limited since their clothing must be suitably selected (it must not be too tight). Some sports (horseback riding, bicycling) are only feasible to a limited extent because they strain the genital area too much. Some patients fear developing carcinoma [2].

RISK OF MALIGNANCY

Vulvar squamous cell carcinoma mainly affects women aged 60–80. It is believed to develop *via* two independent pathogenic pathways. The most known pathway is associated with the HPV and termed HPV-associated usual vulvar intraepithelial neoplasia (VIN). The second one, the HPV-independent differentiated VIN, is thought to have LS as a precursor. Postmenopausal women with LS are known to have an increased risk of squamous cell cancer of the vulva, but the carcinogenic process is not understood. The mechanism of development of genital malignancies in LS is unclear, but they may involve oncogenic HPV infection, altered expression of p53 oncogene, chronic inflammation of LS, and oxidative stress [17]. It is also currently unknown if children and adolescents diagnosed with LS are at the same risk. This is why long-term follow-up with visits every 6–12 months is recommended. All adolescent patients should be encouraged to complete the HPV vaccination series to prevent HPV-related squamous cell carcinoma of the vulva and other HPV-dependent conditions [1].

For the reasons mentioned above, long-term follow-up of patients is essential. Ideally, this should be car-

ried out by the doctor who made the diagnosis. Patients under 18 years of age can be treated and monitored by a pediatrician, dermatologist, or gynecologist.

RECOMMENDATIONS

Lichen sclerosis should be suspected in patients presenting with any urogenital complaint, i.e., constipation, dysuria, dyschezia.

Examining the external genitalia is crucial for making an accurate diagnosis.

Lichen sclerosis can be diagnosed in the pediatric/adolescent population without biopsy.

Clobetasol propionate and betamethasone valerate are the first-line treatment of LS.

All adolescent patients should be encouraged to complete the HPV vaccination.

Long-term follow-up is recommended.

CONCLUSIONS

Lichen sclerosis is a chronic condition that commonly affects prepubertal girls and is associated with vulvar symptoms. It should be suspected in patients presenting with urogenital symptoms and constipation. This is why an examination of the anogenital region is mandatory among girls complaining of constipation. The initial therapy for LS should involve the application of high-potency topical steroids. It is recommended that LS patients undergo long-term monitoring.

DISCLOSURES

1. Institutional review board statement: Not applicable
2. Assistance with the article: None.
3. Financial support and sponsorship: None.
4. Conflicts of interest: None.

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