Ultrapotent Topical Corticosteroid Treatment of Childhood Genital Lichen Sclerosus

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Objective: To observe the clinical effects of short-term application of ultrapotent topical corticosteroid on symptomatic genital lesions of lichen sclerosus in pediatric patients.

Design: Case series of 10 prepubertal girls with genital lichen sclerosus. Ultrapotent topical corticosteroids were applied twice daily for 6 to 8 weeks and patients were reexamined at completion of treatment. Long-term follow-up over 6 months to 3 years.

Setting: Pediatric dermatology clinic (referral center).

Patients: Ten prepubertal girls with typical clinical features of genital and/or perianal lichen sclerosus.

Intervention: Topical ultrapotent corticosteroid ointment was applied sparingly to affected areas for 6 to 8 weeks.

Main Outcome Measure: Improvement of erythema, whitening erosions, and atrophy. Subjective improvement of symptoms.

Results: All patients showed partial or total subsidence of signs and symptoms of lichen sclerosus. Frequency and severity of recurrences varied, but patients responded within a few days to reapplication of ultrapotent topical corticosteroid. No significant adverse effects were noted after the initial 6- to 8-week course of therapy or during the 6-month to 3-year follow-up period.

Conclusion: A 6- to 8-week course of ultrapotent topical corticosteroid is a safe and effective treatment for genital lichen sclerosus in pediatric patients.

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LICHEN SCLEROSUS is a chronic skin disorder of unknown cause that most commonly occurs in adult women. However, 10% to 15% of cases arise during childhood, and almost always affect the genital area. Only 6% of pediatric patients with lichen sclerosus have extragenital involvement. The disease occurs much more frequently in girls than in boys, with a reported female-to-male ratio as high as 10:1. The associated symptoms, particularly pruritus, pain, dysuria, and constipation with pain on defecation, may be chronic and are often disruptive. Because of the location of lesions, discomfort, and signs of localized purpura with bleeding and discoloration, lichen sclerosus in children may be misdiagnosed as child abuse or as a genital infection. The course in children is variable, with some experiencing spontaneous remission, while others show persistence of signs and symptoms in adulthood. However, most reports suggest that affected patients improve significantly at puberty. Earlier studies noted remission of the lichen sclerosus in 80% of patients by puberty. Others have described persistent evidence of the disorder (pallor, atrophy, or loss of tissue) in 89% to 97% of patients, but resolution of symptoms in most patients.

The increased risk of vulvar squamous cell carcinoma in association with lichen sclerosus in adult women is well known. Subsequent development of malignancy in adolescents with vulvar lichen sclerosus is a rare occurrence but has been reported. In contrast, 18% of children develop other long-term sequelae, including scarring, adhesions, and atrophy.

The treatment of lichen sclerosus in pediatric patients has been largely unsatisfactory. Mild-potency topical corticosteroids and bland emollients may reduce pruritus but generally do not alter the course of the disease. Topical testosterone has been used by some physicians; however, concerns about virilization make this an unacceptable treatment in the pediatric population. Moreover, controlled
PATIENTS AND METHODS

Patients were evaluated in the Pediatric Dermatology Clinic of Children’s Memorial Hospital in Chicago, Ill. All patients in the study had typical clinical features of lichen sclerosus at the onset of therapy. These included the presence of whitening, atrophy, erythema, erosions, and purpura in a perineal and perianal distribution. Skin biopsy was thought to be unnecessary in the presence of characteristic clinical features in this age group. Parents were counseled about the unpredictable course of lichen sclerosus, potential adverse effects associated with therapy, and proper application of the topical corticosteroid. Ultrapotent topical corticosteroid ointment with 0.05% clobetasol (Temovate), 0.05% dflorasone diacetate (Psorcon), 0.05% betamethasone dipropionate (Diprolene), or betamethasone dipropionate without propylene glycol (Maxivate) was applied sparingly twice daily to involved areas for 6 weeks. Patients were instructed to apply a “small pea-sized” amount of corticosteroid at each application. The children were reexamined after completing the initial course of treatment. The course after initial application and possible long-term adverse effects during the subsequent 1 to 3 years were monitored by reexamination and telephone follow-up (6 patients, reexamination; 4 patients, telephone follow-up).

Studies show topical testosterone is no more effective than vehicle for treatment of lichen sclerosus. Topical progesterone has been reported to be successful in a limited number of pediatric patients. Recently, topical retinoid therapy has been of some benefit for the treatment of lichen sclerosus in adult women; however, prolonged therapy is required to induce remissions. The effectiveness of this treatment and potential for irritation remains to be determined in pediatric patients.

Studies performed in adult women with vulvar lichen sclerosus have demonstrated that twice daily application for 6 to 12 weeks of 0.05% clobetasol propionate ointment, an ultrapotent topical corticosteroid, is a safe and effective treatment. In these studies, patients were noted to have both clinical and histological improvement of their skin lesions without the occurrence of significant adverse effects from the topical corticosteroids. These patients were subsequently able to control their symptoms with the intermittent use of a lower-potency topical corticosteroid that was previously ineffective, or occasional use of the ultrapotent topical corticosteroid. Ultrapotent topical corticosteroid application was more effective and resulted in fewer adverse effects than topical testosterone or topical progesterone therapy. The use of ultrapotent topical corticosteroids for children with genital lichen sclerosus has rarely been described, and concerns about the risk of associated cutaneous adverse effects have limited its routine use.

We investigated the efficacy and safety of treatment of genital lichen sclerosus with a 6-week course of twice daily application of ultrapotent topical corticosteroid.

REPORT OF CASES

CASE 1

A 6-year-old girl with a history of perianal fissuring presented with vaginal erythema, erosions, and discharge. Physical examination revealed vulvar and perianal erythema with superficial erosions and purulent discharge. Bacterial cultures yielded group A β-hemolytic streptococci. She was treated with a combination antibiotic of amoxicillin-clavulanic acid and topical gentamicin with some improvement. Follow-up examination revealed whitening, purpura, and erosions consistent with lichen sclerosus. She was treated with 0.05% betamethasone dipropionate ointment (Diprolene) applied twice daily for 8 weeks with subsidence of symptoms and improvement of her physical examination. During a 2-year follow-up period she has developed intermittent recurrences, but a 2-day course of treatment with 0.05% betamethasone ointment clears the lesions. No evidence of steroid-induced atrophy or telangiectasias has developed.

CASE 2

An 11-year-old girl with a history of vulvar and perianal lichen sclerosus presented to the clinic for treatment. Physical examination showed whitening, atrophy, erythema, and erosions of the labia majora and introitus. The perianal area was whitened and fissured. She was treated with 0.05% dflorasone diacetate ointment, the use of which was discontinued after several days of treatment because of increased erythema and discomfort. A change in therapy to 0.05% bethamethasone ointment resulted in significant improvement. After 6 weeks of treatment her symptoms had completely resolved. Physical examination continued to show whitening and focal purpura; however, erosions, fissuring, and erythema had resolved. Over a 21-month follow-up period she had 4 mild flares of lichen sclerosus that were treated for 3 to 5 days with betamethasone ointment. No evidence of steroid-induced adverse effects has been noted.

CASE 3

A 9-year-old girl was referred to our clinic with a history of vulvar lichen sclerosus that did not improve with topical progesterone treatment or topical hydrocortisone. Examination showed that she had whitening and erosions of the vulvar and perianal area. She also had numerous atrophic white macules with follicular plugging on her upper chest and back. We initiated 0.05% betamethasone ointment for treatment of the lesions. The patient complained of pain associated with application of the ointment and was switched to another preparation of 0.05% betamethasone ointment that did not contain propylene glycol. This was well tolerated and on follow-up at 6 weeks her discomfort had resolved. Physical examination revealed improvement, but mild whitening and atrophy persisted. The truncal lesions remained unchanged with treatment. She experienced 1 flare during the subsequent 20 months, which responded to ap-
Ten children with lichen sclerosus were enrolled in the study, ranging in age from 2.5 to 11.5 years (Table). Ten girls with genital and/or perianal lichen sclerosus were treated with an initial 6-week course of topical ultrapotent corticosteroid ointment. For most patients, the correct diagnosis was made in the dermatology clinic. The duration of symptoms of lichen sclerosis before initiating treatment ranged from several weeks to more than 7 years. Pruritus, chronic discomfort, dysuria, and pain with defecation were common chief complaints in our patients. Previous application of nonfluorinated corticosteroids (eg, hydrocortisone) or other topical medicaments (eg, antibacterial ointments, antifungal creams, or bland emollients) had resulted in no significant or persistent improvement. Most patients had perianal and vulvar involvement in the classic "figure eight" or "keyhole" distribution. One of the 10 children (patient 3) had extragenital involvement with truncal lesions.

All children demonstrated clinical improvement of their symptoms and skin lesions on follow-up examination. Four of the 10 girls showed improvement but were not clear after 6 weeks, and they showed further improvement after treatment with corticosteroid for an additional 2 weeks.

Four girls complained of irritation and burning and overall initial worsening with application of ultrapotent topical steroid. One patient who experienced burning from use of topical corticosteroid continued application of the clobetasol that was initially prescribed (patient 6). She was examined 5 weeks after initiation of the treatment, and application of the topical corticosteroid was discontinued because the signs of lichen sclerosus had almost cleared despite the burning sensation. The ultrapotent topical corticosteroid preparation was changed in the other 3 girls, and in 2 of them (patients 2 and 8) the second corticosteroid preparation was tolerated well. In the third girl (patient 3), burning was experienced with the applications of both betamethasone and diflorasone, but she tolerated the betamethasone ointment without propylene glycol, suggesting the occurrence of either reaction to the propylene glycol or perhaps spontaneous subsidence of the inflammatory response after 2 weeks of therapy.

The duration of further follow-up varied from 4 to 36 months after completing the initial course of therapy. All reported overall significant subjective improvement; however, 6 girls were noted to have recurrences that were described as mild, and responded to brief courses (2 days to 1 week) of ultrapotent topical steroid application. No steroid-induced atrophy or telangiectasias were noted on follow-up examinations.

Lichen sclerosus is an inflammatory condition of unknown cause that most commonly affects the genital skin of prepubertal girls and postmenopausal women, although extragenital sites may also be involved. In children, symptoms usually begin between 3 and 7 years of life. Dysuria, pruritus, constipation, and pain are common presenting complaints. As shown in the selected cases described, the recognition of lichen sclerosis in children is poor among primary care physicians and even among specialists.

Most commonly, affected children are erroneously thought to have bacterial or, less commonly, yeast vaginitis, and are treated with several topical and systemic antibiotics before referral is made. It should be realized, however, that secondary bacterial or yeast infection may occur, as seen in case 1. In a British study, bacterial cultures from the genital area of children with lichen sclerosus yielded organisms in 56% of 42 affected girls. Streptococcus was cultured from the area in 21% of the patients, while Escherichia coli and Staphylococcus were found in 10% and 7% of patients, respectively. Candida albicans was detected in 1 patient. Secondary bacterial infections may show intense erythema, increased discomfort, and purulent drainage; candidal infections tend to be associated with increased erythema. If infection is suspected, cultures should be performed to verify the presence of bacterial or candidal organisms, and appropriate antibiotic therapy should be initiated before administration of topical corticosteroid therapy for the underlying lichen sclerosis.

Patients with lichen sclerosis may present with severe pain with defecation and/or urination that leads to gastroenterology or urology referral, respectively, with generally ineffective management unless the underlying cause is recognized. The other frequent misdiagnosis for patients with lichen sclerosus is child abuse, with parents accused on the basis of the symptoms of pain and pruritus, and the clinical signs of bleeding, erosion, and discoloration of the genital area. On the other hand, a diagnosis of lichen sclerosus does not rule out the possibility of concomitant sexual abuse.

The lesions of lichen sclerosis in children are often associated with considerable burning pain requires
intervention. Treatment of childhood lichen sclerosus has been difficult. Application of bland emollients and mild-potency topical corticosteroids has been the mainstay of therapy. These treatments are often only partially effective. Local application of hormones has been reported to be effective in some patients. Progesterone may be effective in some patients. Its use has been limited in children because of concerns about potential adverse effects associated with hormonal therapy. Topical testosterone has been found to be of no more effective than petrolatum for the treatment of this disorder.10 Ultrapotent topical corticosteroid application has been the most effective and well-tolerated therapy for adults with this disorder.

We reviewed our experience using ultrapotent topical corticosteroid ointments to treat vulvar and perianal lichen sclerosus. All 10 children experienced rapid subjective reduction of symptoms and improvement in the clinical appearance of lesions after treatment. Improvement was defined as a resolution of purpura, erosions, and reduction of erythema and whitening.

Atrophy and telangiectasia, cutaneous adverse effects of potent topical steroid application, were not observed in any of our patients following treatment, although Fischer and Rogers17 have recently described labial telangiectasia in 18% of 11 pediatric patients with lichen sclerosus treated with clobetasol. Four of our patients complained of burning with application of initially applied ultrapotent corticosteroid ointment, associated with increased erythema and edema at sites of application. In 3 of these patients, therapy was changed after 1 to 2 weeks to an alternate ultrapotent topical corticosteroid ointment. The frequency of this reaction, even though the particular potent corticosteroid preparation varied, made us consider the possibility that the mechanism of corticosteroid action involved a burst of released mediators of inflammation, which ultimately resulted in improvement and sometimes resolution of the lichen sclerosus. In the long-term follow-up study by Dalziel and Wojnarowska14 of 12 adult women treated with 0.05% topical clobetasol cream, 2 patients experienced burning and discomfort with application. One patient developed complaints after initially tolerating treatment but was found to have a contact sensitivity to the preparation on patch testing. No contact sensitivity could be determined to the topical corticosteroid or to the vehicle in the second patient. Our patients did not undergo patch testing, but contact dermatitis to propylene glycol may have caused the reaction in patient 3. Further observation of children receiving this therapy needs to be undertaken to determine the frequency and cause of the early inflammatory response to the potent corticosteroids.

In conclusion, we have found a 6- to 8-week course of ultrapotent topical corticosteroid applied twice daily to affected areas to be a safe and effective treatment for vulvar and perianal lichen sclerosus in children. Continuing observation every 6 to 12 months is important to monitor for recurrences. Further study is needed to assess the safety of long-term therapy that may be necessary to prevent scarring that is associated with recurrence.

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REFERENCES