



## Depigmented plaques on vulva

The distinctive pattern of our young patient's skin changes made the diagnosis clear.

**A MOTHER BROUGHT HER 8-YEAR-OLD DAUGHTER** to our office for evaluation of vitiligo “down there” (FIGURE). The skin eruption first appeared on her vulva a year earlier and was intermittently pruritic. The lesions were initially smaller and red, but had since lightened in color, coalesced, and had begun to spread to the perianal area. The patient's mother had received a call from her daughter's teacher who observed that her daughter was scratching the area and might be masturbating in class.

The mother reported that 6 months earlier, her daughter had experienced bloody spots in her underwear accompanied by dysuria. The mother brought her to the emergency department, where she was treated with antibiotics for a urinary tract infection.

Our physical examination revealed well-circumscribed, symmetric, depigmented, confluent, crinkled, parchment-like plaques with small hemorrhagic erosions on the medial labia majora and minora. The lesions had spread to the perianal area with depigmentation superiorly and hypopigmentation inferiorly, creating a figure-8 pattern.

A review of systems was negative for pruritus, pain, dysuria, dyschezia, constipation, and vaginal discharge. The patient denied sexual activity, depression, or anxiety. Her mother denied behavioral changes in her daughter and said that her daughter hadn't had any one-on-one time alone with any adults

besides herself. Her mother was concerned that the white spots might spread to the rest of her daughter's body, which could affect her socially.

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

### FIGURE

Depigmented plaques in a figure-8 pattern



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➤ Up to 84% of cases of childhood lichen sclerosis are misdiagnosed, with an average delay of more than a year between symptom onset and diagnosis.

**Diagnosis:  
Lichen sclerosis**

Based on the history and clinical findings, including the classic figure-8 pattern, we diagnosed childhood lichen sclerosis (LS) in this patient. LS is a chronic inflammatory skin disorder that primarily affects the genital mucosa. The disorder can present at any age, but is most common among postmenopausal women, with a prevalence estimated to be as high as one in 30.<sup>1-3</sup> A second incidence peak is observed in prepubescent girls, with a prevalence of one in 900.<sup>3,4</sup> LS is less common in men and boys, with a female-to-male ratio that can reach 10:1.<sup>5</sup> The classic symptoms of LS are pruritus and pain, which may be intermittent or persistent.

■ **In girls**, initial manifestations may be constipation, dysuria, or even behavioral symptoms such as night fears, which can occur because children are less active at night and become more aware of urinary discomfort.<sup>1,2,6</sup> Typical signs of LS are thin atrophic plaques that spare the vagina and cervix. The plaques can be ivory-white, erythematous, or violaceous. Some patients have perianal lesions as well, and can display the pathognomonic figure-8 pattern of porcelain plaques around the vulva and anus.<sup>5</sup>

With more advanced disease, erosions, lichenification, and even distortion of vulvar architecture may occur.<sup>2,4,7</sup> In severe cases, labia resorption and clitoral phimosis may develop.<sup>5</sup> Complications include secondary infection, dyspareunia, and psychosexual distress. The most worrisome sequela of LS is squamous cell carcinoma of the vulva (SCCV), which occurs in 5% of female patients with LS.<sup>4</sup>

■ **In men and boys**, LS typically involves the foreskin and the glans, while sparing the perianal region.<sup>5</sup> Scarring of the foreskin can lead to phimosis, and patients may complain of painful erections and difficulty urinating. LS can also occur away from the genitalia in both males and females.

**Autoimmune mechanisms, genetics, and hormones play a role**

The exact pathogenesis of LS remains unknown, but multiple factors are likely at work.

■ **Autoimmune mechanisms.** Up to 60% of women with LS have an autoimmune disorder,

which is most commonly vitiligo, alopecia areata, or thyroid disease.<sup>5</sup> In addition, 67% of patients have autoantibodies against extracellular matrix protein 1, and 30% have them against bullous pemphigoid antigen 180.<sup>1,8</sup>

■ **Genetics.** LS is associated with certain human leukocyte antigen class II haplotypes (especially DQ7) and with polymorphisms at the interleukin-1 receptor antagonist gene locus.<sup>5,6,9</sup>

■ **Hormones.** The clear peaks of incidence during times of low estrogen, and a higher incidence in patients with Turner syndrome or kidney disease, suggest that low estrogen may play a role in the development of LS, as well.<sup>1,5,6</sup>

While it is generally accepted that trauma may trigger LS via the Koebner phenomenon (the appearance of lesions at the site of injury), there is debate as to whether microbes—especially *Borrelia burgdorferi* and human papillomavirus (HPV)—might play a role.<sup>1,5</sup>

**Diagnosis is often delayed, misdiagnosis is common**

The average delay from symptom onset to diagnosis of LS is 1.3 years, and up to 84% of childhood LS is misdiagnosed before referral.<sup>2,9</sup> The differential diagnosis includes:

■ **Sexual abuse.** In prepubertal girls presenting with genital redness, the can't-miss diagnosis is sexual abuse, which occurs in more than 25% of children in the United States.<sup>10</sup> Initial manifestations may be regression in developmental milestones, such as new-onset bedwetting, or behavioral changes such as social withdrawal or declining academic performance.<sup>11</sup>

However, physicians must be conscientious about ruling out medical etiologies before prematurely diagnosing abuse. Fourteen percent of girls with LS are incorrectly diagnosed as having been sexually abused.<sup>2</sup> A clinical pearl is that while LS may resemble abuse on exam, it rarely affects the hymenal structure.<sup>12</sup> It is also important to keep in mind that the 2 entities are not incompatible, as sexual abuse leading to LS via Koebnerization is a well-described phenomenon.<sup>12</sup>

■ **Lichen planus.** LP, which is also an immune-mediated inflammatory disorder affecting the vulva, classically presents with the

6 Ps: pruritic, polygonal, planar, purple papules and plaques.<sup>4</sup> LP is distinguished from LS by being rare in childhood, having a predilection for the flexor wrists, and involving the oral and vaginal mucosa.<sup>4</sup>

■ **Lichen simplex chronicus (LSC)** is a chronic, circumscribed, pruritic, eczematous condition that becomes lichenified with thickened skin secondary to repeated scratching.<sup>13</sup> Children with atopic dermatitis can develop LSC, but other children can also develop the scratch-itch cycle that results in the thickened plaques of LSC. Like LS, LSC can occur in areas other than the genitalia, including the neck and feet.<sup>14</sup>

■ **Allergic contact dermatitis** can occur in the genital area from diaper creams, soaps, and perfumes. Irritant contact dermatitis can occur from exposure to diarrhea, bedwetting, and other irritants. Contact dermatitis is less likely to have the classic figure-8 pattern seen in LS.

■ **Psoriasis** in the genital area can be confused with LS. However, psoriasis favors the

groin creases in what is called inverse psoriasis. In addition, psoriasis tends to involve multiple areas, including the extensor surfaces of the elbows and knees, the nails, and the scalp.

■ **Vitiligo** can present on the genitals as circumscribed hypopigmented and depigmented patches that are flat. Vitiligo is asymptomatic, and the only pathology is the change in skin color. With LS, there is lichenification, atrophy, and sclerosis.<sup>4</sup> Vitiligo often occurs with bilateral symmetric involvement in areas of trauma including the face, neck, scalp, elbows, wrists, hands, knees, ankles, and feet.

#### **Treatment aims to improve symptoms**

LS is usually diagnosed clinically (especially in children, as a biopsy is a great challenge to perform). However, when the clinical presentation is unclear, a skin biopsy will demonstrate the diagnostic findings of thinning of the epidermis, loss of rete pegs, hyperkerato-

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sis, and dermal fibrosis with a T-lymphocyte-dominant inflammatory infiltrate.<sup>1,2,4,5</sup>

LS is a remitting and relapsing condition with no cure. The goals of treatment are to provide symptom relief and minimize scarring and atrophy,<sup>2</sup> but it is unknown whether treatment reduces the risk of malignancy.<sup>9</sup>

First-line treatment for both genders and all ages is ultrapotent topical corticosteroids; clobetasol propionate 0.05% is most commonly used.<sup>1,6</sup> Regimens vary, but the vast majority of patients improve within 3 months of once-daily treatment.<sup>4</sup>

For refractory LS, calcineurin inhibitors such as tacrolimus may be used. Although it has a black box warning regarding a potential cancer risk, long-term studies of children using tacrolimus for atopic dermatitis have not demonstrated an increased risk of malignancy.<sup>6,9</sup> Because of a considerable adverse effect profile, oral retinoids are limited to refractory cases in adults.<sup>6</sup> Surgery is reserved for scarring and adhesions.<sup>4</sup>

**Follow-up plays an important role in management**

Historically, it was believed that pediatric LS had an excellent prognosis, with patients achieving complete resolution after puberty.<sup>1,4</sup> Recent findings have shown mixed results, with LS persisting in many patients beyond puberty.<sup>2,4</sup> Therefore, regular follow-up is recommended every 6 to 12 months.

For uncomplicated LS, specialist follow-up is not indicated. Female patients should regularly conduct self-examinations and, at a minimum, undergo annual examinations by their primary care physician. Those who require specialist follow-up include patients with difficult-to-control symptoms, hypertrophic lesions, a history of SCCV or differentiated vulvar intraepithelial neoplasia (dVIN), or pathology showing possible dVIN.<sup>15</sup>

■ **Our patient.** We prescribed clobetasol propionate 0.05% ointment to be used once

daily for 8 weeks. We stressed the importance of genital self-examinations using a mirror to monitor for any concerning changes such as skin thickening. We showed the patient and her mother photos of normal female genitalia to help normalize the genital exam, and taught the patient how to find her plaques in the mirror. We set expectations by emphasizing the chronic nature of LS and the likelihood of recurrence. We also encouraged HPV vaccination in the upcoming years to prevent both cervical cancer and HPV-related SCCV. **JFP**

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**References**

- Murphy R. Lichen sclerosis. *Dermatol Clin.* 2010;28:707-715.
- Dendrinis ML, Quint EH. Lichen sclerosis in children and adolescents. *Curr Opin Obstet Gynecol.* 2013;25:370-374.
- Eva LJ. Screening and follow up of vulval skin disorders. *Best Pract Res Clin Obstet Gynaecol.* 2012;26:175-188.
- Focseneanu MA, Gupta M, Squires KC, et al. The course of lichen sclerosis diagnosed prior to puberty. *J Pediatr Adolesc Gynecol.* 2013;26:153-155.
- Funaro D. Lichen sclerosis: a review and practical approach. *Dermatol Ther.* 2004;17:28-37.
- Heymann WR. Lichen sclerosis. *J Am Acad Dermatol.* 2007;56:683-684.
- Tong LX, Sun GS, Teng JM. Pediatric lichen sclerosis: a review of the epidemiology and treatment options. *Pediatr Dermatol.* 2015;32:593-599.
- Lagerstedt M, Karvinen K, Joki-Erkkilä M, et al. Childhood lichen sclerosis—a challenge for clinicians. *Pediatr Dermatol.* 2013;30:444-450.
- Keith PJ, Wolz MM, Peters MS. Eosinophils in lichen sclerosis et atrophicus. *J Cutan Pathol.* 2015;42:693-698.
- National Sexual Violence Resource Center. Child sexual abuse prevention. 2011. Available at: [https://www.nsvrc.org/sites/default/files/Publications\\_NSVRC\\_Overview\\_Child-sexual-abuse-prevention\\_0.pdf](https://www.nsvrc.org/sites/default/files/Publications_NSVRC_Overview_Child-sexual-abuse-prevention_0.pdf). Accessed February 8, 2018.
- Dubowitz H, Lane WG. Abused and neglected children. In: Kliegman RM, Stanton BF, St. Geme JW, et al, eds. *Nelson Textbook of Pediatrics.* 20th ed. Philadelphia, PA: Elsevier; 2016:236-249.
- Powell J, Wojnarowska F. Childhood vulvar lichen sclerosis: an increasingly common problem. *J Am Acad Dermatol.* 2001;44:803-806.
- Reamy BV, Bunt CW, Fletcher S. A diagnostic approach to pruritus. *Am Fam Physician.* 2011;84:195-202.
- Warshaw E, Hook K. Dermatitis. In: Soutor C, Hordinsky MK, eds. *Clinical Dermatology.* 1st ed. New York, NY: McGraw-Hill; 2013.
- Jones RW, Scurry J, Neill S, et al. Guidelines for the follow-up of women with vulvar lichen sclerosis in specialist clinics. *Am J Obstet Gynecol.* 2008;198:496.e1-e3.

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