Vulvar Lesions in a 5-Year-old Girl

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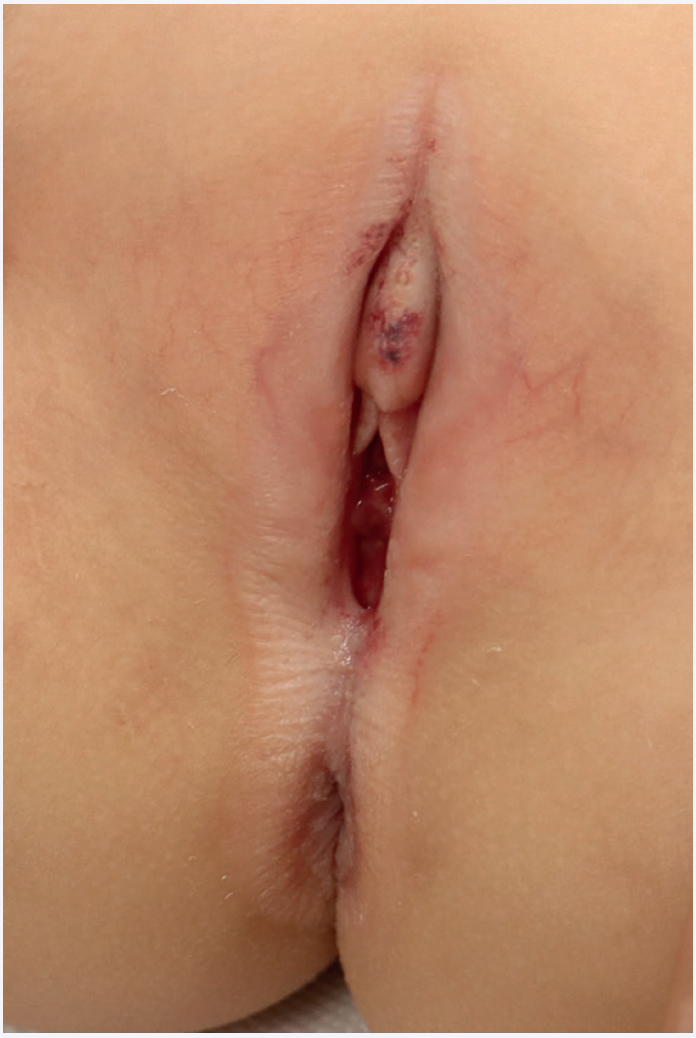

Case report

A 5-year-old girl presented to the Sexual Assault Center of Antwerp (SAC). A SAC is a center that provides multidisciplinary care to victims of sexual violence and advice and help to their support circle. This care is offered in one place by a team, consisting of specially trained nurses, physicians and psychologists. The girl was referred by a physician because of genital injuries suspected for sexual abuse. According to the mother, a single parent, there were no known suspicious contacts. The mother could not provide an explanation for her daughter's genital injuries, although she described an incident in which the child was accidentally kicked in the vulvar region when she was three years old. According to the mother there were no problems with the child's behavior, either at home or at school. The girl's development was normal, sleeping behavior was normal for her age. Physical examination revealed small hematomas around the preputium and labia, with lacerations at the fourchette and around the anus. There was hypopigmentation of the skin around the vagina and anus (Figure). The hymen was normal. What is the diagnosis?

Diagnosis

The vulvar lesions in this girl are caused by lichen sclerosus (LS), a chronic autoinflammatory disease. It can present in a number ways and is often initially misdiagnosed with an average diagnostic delay of 1 to 2 years. Because of the genital lesions, sexual abuse is suspected in 14% - 70% of children with LS (1).

LS is more prevalent in females than males with a bimodal age distribution affecting both premenarchal and postmenopausal women. It is most commonly diagnosed in postmenopausal women. However, approximately 5–15% of cases occur in childhood, typically from ages 4 to 6 (1). The prevalence is estimated at 1:900 in premenarchal girls, but because of underdiagnosis of this condition the true prevalence numbers are unknown (2).

Clinical presentation

Typical for LS are ivory white and rose-colored patches forming a hypopigmented area around the vulva. This area can spread to the perianal region forming a classic "figure of eight" or "key-hole" shape. The patches may be atrophic and shiny or thickened due to hyperkeratosis caused by repeated excoriations from scratching. Repeated excoriations from scratching can lead to ecchymoses, subepithelial hemorrhage, superficial erosions, fissures and telangiectasias. Subsequently, these skin changes may progress to genital scarring which can result in adhesion of labia and clitoral hood, narrowing of the vaginal introitus and loss or attenuation of labia minora (2). These architectural changes are more frequently present in adolescents (3). Also, melanotic macules and nevi can emerge as a result of melanocytic proliferations (2,3).

Complaints associated with LS in children are mostly vulvar irritation and pruritus. Other significant symptoms include pain, bleeding due to skin fissures, constipation and dysuria (2,3). Vulvar irritation and pain may be more present at night (2,4). Adolescents can report dyspareunia and are less likely to complain of pruritus (3). Extragenital LS occurs in 18% of cases and emerges mostly on the trunk and extremities, less frequently in the mouth (2).

Diagnosis is based on history and clinical examination. In some cases, the lesions remain asymptomatic and may be discovered incidentally. Performing a biopsy is rarely necessary for the diagnosis in the pediatric population, although it can be warranted in case of treatment failure or when the lesions are atypical and the diagnosis is unclear. In postmenopausal women vulvar biopsies are more frequently performed because of the increased risk of vulvar squamous cell carcinoma (2).

Differential diagnosis

Lichen sclerosus presents in a variety ways and may mimic other conditions, such as trauma or sexual abuse. Children who have been victims of sexual abuse will generally show no or subtle injuries on examination (5). If there are lesions in girls due to sexual abuse, lacerations of the labia, perineum, posterior fourchette or vagina can be seen in the acute phase. Also, bruising, petechiae and lacerations or abrasions of the hymen can be seen. Although early recognition of LS is important to counter unnecessary worries about sexual abuse and to initiate appropriate treatment of the condition, it should be noted that LS and sexual abuse can of course coexist. LS is diagnosed in 4% of cases of confirmed sexual abuse (1). If sexual abuse is suspected, a trained healthcare professional should evaluate the child. In Belgium, such care is available at ten SACs spread throughout the country. In the future 3 more SACs will be opened, which means that each province of Belgium will have its own SAC. SACs provide comprehensive multidisciplinary support to individuals affected by possible sexual violence and their families. This includes medical and psychological care, but also forensic evaluation and assistance with initiating legal proceedings, including the possible filing of a police report (6).

Other differential diagnoses to consider include vitiligo, lichen planus, psoriasis, atopic dermatitis, eczema or candidiasis. Although vitiligo is also characterized by hypopigmentation, it is sharply demarcated with a patchy distribution. It is asymptomatic and is not associated with vulvar atrophy or structural changes. Lichen planus in prepubertal girls is extremely rare; when it presents in the vulvar region, mucosal involvement is possible, whereas this is not seen in LS (2).

Pathogenesis

The pathogenesis remains uncertain but LS is considered an autoimmune disorder. In children it is related to other autoimmune disorders like thyroiditis, pernicious anemia, psoriasis, vitiligo, morphea and alopecia areata. An unspecified genetic contribution is suspected, as 25% of girls with LS have a first-degree relative with autoimmune disorder. In addition, there is a higher prevalence of HLA class II antigen DQ7 in children with LS. Patients with Turner syndrome, who are at risk for autoimmune disorders, have a higher prevalence of LS. The role of other factors such as connective tissue alterations, sex hormone factors and trauma, remains unclear (1,2).

Treatment

The primary goal of treatment is not only to relieve symptoms but also resolve atrophic changes and prevent scarring. First-line treatment is with high-potency topical corticosteroids (class IV). Although there can be considerable side effects, such as thinning of the dermis, these outweigh the risk of untreated or undertreated LS. Topical calcineurin inhibitors (tacrolimus and pimecrolimus) are considered second-line treatment. These immunomodulators are as efficient as corticosteroids, have fewer systemic effects and a lower risk of atrophic changes. However, they are not the first choice treatment given the long-term safety is not yet warranted. After initial treatment with high-potency corticosteroids for at least 4 weeks, maintenance therapy with low/medium-potency corticosteroids is warranted to prevent relapse. Supportive measures, such as good vulvar hygiene, cool compresses and hypoallergenic topical emollients are encouraged to relieve discomfort. In addition, vulvar pruritus may be treated with antihistamines (2).

LS is a chronic condition. Continued monitoring into puberty and adulthood is recommended due to considerable risk of relapse. Winfrey et al. reported that childhood disease persists into adolescence in approximately 40% of patients (3). Consequently, periodic follow-up is important to prevent long-term sequelae including anatomic changes that can significantly impact quality of life and sexual function (2–4).

Conclusion

Lichen sclerosus typically presents with genital injuries that may raise concerns of sexual abuse. Recognition of LS is important to initiate timely and adequate treatment, resolve symptoms and prevent long-term sequelae. Diagnosis may also prevent unnecessary concerns of sexual abuse. However, LS and sexual abuse can coexist and referral to a trained health care professional is warranted in case of concern of sexual abuse. In Belgium, 10 different sexual assault centers are easily accessible for specialized care.

A written permission for the publication of the figure has been given by the parents.

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