Our Dermatology Online

Brief Report

Vulvar lichen sclerosus in female children

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ABSTRACT

Background: Vulvar lichen sclerosus (VLS) is an uncommon inflammatory dermatosis affecting the anogenital region. There are two peaks of incidence: one in prepubertal girls and the other in postmenopausal women. Its etiopathogenesis remains unknown. **Objective:** The objective was to specify the clinical characteristics of VLS in the female pediatric population in order to plan early treatment. **Results:** A total of 12 pediatric female patients were identified in the study. The mean age of onset was six years. Numerous potential etiological factors for VLS development were identified. Pruritus was the main symptom in a significant proportion of the patients. All patients had a good response to topical therapy with no significant side effects. **Conclusion:** VLS should be suspected in the presence of chronic vulvar pruritus in order to avoid tragic repercussions in adulthood. Appropriate treatment for each patient is essential as is the prevention of the side effects of applied treatments.

Key words: Vulvar Lichen Sclerosis; pediatric; adolescent

INTRODUCTION

Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis with a predilection for the anogenital region, yet it may affect any area, with an estimated prevalence of 1 in 900 girls. [1]. It has two peaks of onset: in the prepuberty and post-menopause [1].

In the pediatric population, vulvar pruritus is the most common symptom. It is most often treated wrongly by general practitioners as recurrent pinworm infection or considered a hygiene defect, which leads to a delay in diagnosis. Pain, dysuria, and a burning sensation along the perineal region may also be symptoms of this condition. Anorectal lesions may cause additional symptoms such as constipation or painful defecation, without any gastrointestinal problems in the patient's medical history [2]. The pathogenesis of VLS is unknown, yet a number of potential etiological factors have been identified.

An association with other autoimmune diseases has been reported [3]. Its treatment is effective as long as it is done early. However, we are faced with the constraints of the location of the lesions and the side effects of the treatments applied.

MATERIALS AND METHODS

Data was collected from case notes of the female patients below 18 years of age with a diagnosis of VLS attending our dermatology department over a five-year period. Case notes were reviewed for clinical presentation, predisposing factors, comorbidities, therapy, and outcome. The diagnosis of VLS was based on clinical criteria, and histology was reserved for doubtful cases.

RESULT

A total of twelve pediatric female patients were identified. The median age of symptom onset was six

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years (range: 3–11 years). The average delay between the onset of symptoms and the diagnosis was 2.6 years. The majority of the patients were referred from the primary health care system or by pediatricians. In most cases, the diagnosis was not suspected prior to referral.

The reason for consultation was mainly vulvar pruritus in seven cases, a burning sensation in three cases, and achromia giving rise to suspicion of vitiligo in two cases. An association with urinary incontinence was found in three cases. In our patients, VLS was associated in two cases with vitiligo, in three cases with alopecia, and in one case with Down syndrome. On clinical examination, skin sclerosis was noted in five cases, clitoral hooding in three cases, disappearance of labia minora in two cases, and vulvar edema in two cases. A skin biopsy was performed on a single patient to exclude Crohn's disease, who presented with severe vulvar edema associated with rhagades and the transit disorder (Fig. 1). For the remaining, the diagnosis was clinical.

All our patients were put on ultra-high-potency topical corticosteroids at a rate of five days per week, associated with an antimycotic at a rate of two times per week for a period of three months with a regression pattern (Fig. 2). Ten patients had a satisfactory outcome; for the rest, we opted for the prolongation of the treatment up to 4–6 months. Patients with secondary enuresis were referred to pediatrics for further treatment. A maintenance treatment was prescribed based on dermocorticoids or topical tacrolimus, depending on availability, and the total duration of treatment varied from nine to eighteen months. In our patients, no recurrence of the disease was noted over a period of three years, and due to the sequential application of dermocorticoids and their association with antimycotics, no side effects have been observed.

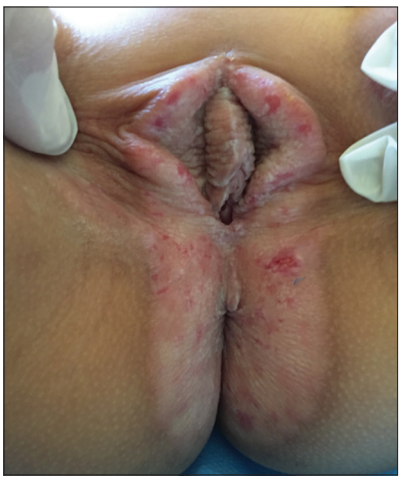

Figure 1: Associated severe vulvar edema with erosions and fissures.

DISCUSSION

The vulvar lichen sclerosus course is highly heterogeneous; therefore, it causes diagnostic difficulties. It may occur at any age and in both sexes, with two peaks of onset: in the prepuberty and post-menopause [4]. In premenarchal girls, LSV may be observed at 1 in 900 [1].

VLS is rarely asymptomatic. Itching and a burning sensation were found to be the commonest presenting symptoms in our cases. Vulvar edema, burning sensation, perineal pain, bleeding, dysuria, and reflex constipation have also been reported [5,2].

Its presentation is heterogeneous, which leads to a delay in diagnosis [6]. VLS presents clinically with classic figure 8 [7] (Fig. 3). Color changes may also be seen: white pearly scales, post-inflammatory pigmentation, and ecchymosis, which may condemn a sexual assault, although these two situations may coexist [8,9]. Pallor, sclerosis, edema, and fissuring were the most frequent signs found in our cases. This was similar to findings in other studies [1,10].

There was a delay between the first symptoms and diagnosis in most patients. Scarring symptoms were present at the time of consultation, highlighting the importance of early detection and treatment of this condition.

Most of our patients had a history of autoimmune disease. Several case reports and studies have shown a potential link with autoimmune diseases such as thyroiditis, vitiligo, alopecia areata, type 1 diabetes mellitus, and celiac disease [11]. As a result, patients are screened for autoimmune diseases, although there are no specific antibodies that could be markers of VLS [12,13].

Incontinence is not uncommon in the pediatric population. Many patients in our case series reported secondary enuresis at the time of diagnosis.

The role of urine in the pathogenesis of VLS is increasingly recognized due to its high permeability to irritants and abrasion sensitivity [14]. Not to mention the role of skin occlusion [14]. In addition, it adds the washing-off effect of urine on topical treatment [14].

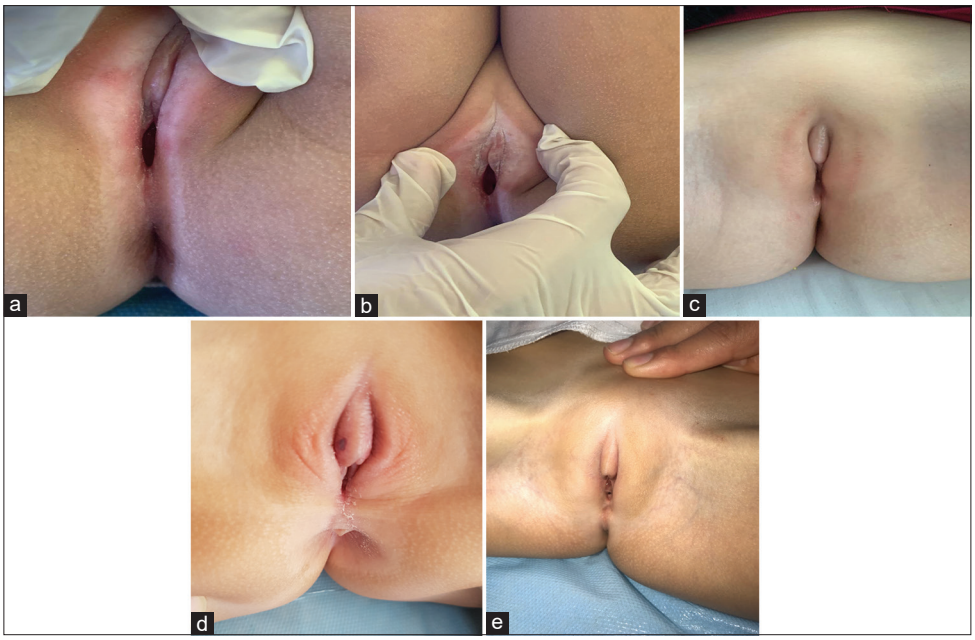

Figure 2: Disappearance of achromia, erosions, and ecchymosis: a) patient 1 after treatment, b) patient 1 before treatment, c) patient 2 before treatment, d) patient 2 before treatment, e) patient 2 after treatment.

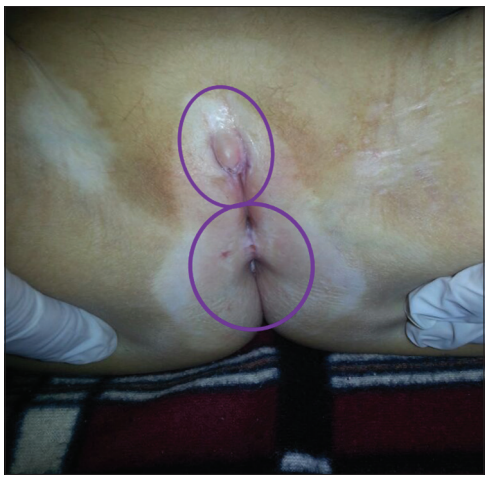

Figure 3: Classic figure 8 pattern involving the labia minora, clitoral hood, and perianal region.

If not treated early on, complications may arise, in particular, the modification of the relief, repeated infections, vulvodynia, etc. [8,15]. However, the risk of transformation into squamous cell carcinoma by chronic inflammation in pediatric patients is lower than in adults [15]. Vulvar biopsies are generally not performed on children, as they may be highly painful. It should be reserved only for cases with an uncertain diagnosis, as in the case of our patient, who was suspected of having Crohn's disease due to the presence of edema with rhagades and transit disorders, and for those who fail to respond to treatment [16].

Treatment with ultra-high-potency topical corticosteroids is the gold standard, with a progressive degression in order to assess the efficacy and safety of the treatment and the occurrence of complications. It is recommended that patients are monitored after four weeks of treatment [8,16,17]. Maintenance therapy for VLS is still debated among clinicians and requires an individualized approach for each patient. To establish long-term effective maintenance strategies to prevent disease recurrence and the remote effects of VLS scarring, adhesions, or dyspareunia, further research is needed in this age group of patients [18]. Calcineurin receptor inhibitors may be offered as maintenance therapy or as an effective alternative [8].

Other treatments have been reported, such as topical retinoids, phototherapy, cyclosporine, and vitamin D, A, and E therapy [8,18].

In our patients, sequential treatment with dermocorticoids for five days a week combined with topical antimycotics on the next two days resulted in optimal treatment while avoiding the side effects of dermocorticoids.

CONCLUSION

Vulvar lichen sclerosis is an inflammatory pathology whose diagnosis is based on clinical findings. In

children, if treated early, it evolves without sequelae, yet it must be suspected in the presence of chronic vulvar pruritus in order to prevent tragic repercussions in adulthood. In our cases, treatment adapted to each patient enabled optimal therapy and the prevention of the side effects of the treatments applied.

Statement of Human and Animal Rights

All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

Statement of Informed Consent

Informed consent for participation in this study was obtained from all patients.

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