WHAT'S YOUR DIAGNOSIS?

PEER REVIEWED

White Vulvar Plaques in an Eight-Year-Old Girl

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An 8-year-old girl presented to the pediatric dermatology clinic with her mother with a chief concern of a pruritic rash located on the genitalia that has been worsening, on and off, for 7 years.

History. The patient's mother states that the patient has had occasional bouts of constipation but no urinary tract infections. The mother tried applying 2.5% hydrocortisone with no apparent improvement and has recently been cleansing the area with just water. Upon presentation, the patient was in no acute distress and appeared well developed, well nourished, alert, and oriented. Glistening white atrophic papules with telangiectasia, purpura, and clitoral hood edema were observed on the vulva and perianal skin. The remainder of the physical examination was unremarkable.

Diagnostic testing. No laboratory tests were obtained, and a clinical diagnosis was made.

Based on this patient's clinical presentation, what's your diagnosis?

A. Lichen Sclerosus

B. Sexual Abuse

C. Vitiligo

D. Vulvovaginal Candidiasis

Answer and discussion on the next page.

Correct answer: A. Lichen Sclerosus

Despite similarities in presentation, a close inspection of the area led to the diagnosis and exclusion of differentials. The patient was diagnosed based on distinguishable features on physical examination, including atrophic plaques and clitoral hood edema, which are clinically diagnostic of lichen sclerosus (LS). These unique clinical features include the “figure-of-eight” distribution, clitoral hood edema, telangiectasias, ecchymosis, fissure, erosions, scarring, and cutaneous atrophy.[1]

The diagnosis of vulvovaginal candidiasis was considered. This is an infection most commonly caused by the yeast *Candida albicans* and is seen in areas of the female reproductive tract.[2] This diagnosis is more common in women who are pregnant, using hormonal contraceptives, have diabetes, are immunocompromised, or with recent antibiotic use.[3] Vulvovaginal candidiasis diagnosed in a younger population is typically due to recent antibiotic use or tight clothing, creating a moist environment for the yeast to grow.[4] Though *C. albicans* is considered part of the normal flora, overgrowth of this yeast produces an inflammatory response with symptomatic consequences such as vaginal pain, pruritus, burning, and erythema.[2] Cutaneous manifestations may vary from none to a mild erythema of the mucosa.[4] Despite commonalities such as compulsive pruritus, lichen sclerosus tends to show hyperkeratotic and sclerotic skin changes and is without vaginal discharge, a feature that is otherwise characteristic of vulvovaginal candidiasis.[5] Treatments for *candida* consist of antifungal drugs (most commonly azoles), topical preparations, or vaginal suppositories.[2]

The diagnosis of vitiligo was also considered. Vitiligo is a depigmenting disorder caused by the destruction of melanocytes.[6] The physical manifestations of this condition are milky white, non-scaly macules, and patches with distinct margins.[6] Diagnosis of vitiligo is centered on sharply demarcated, depigmented areas of skin without other physical concern, whereas lichen sclerosus, in addition to ivory white or rose-colored patches, can present with areas of purpura, ecchymosis, fissures, and superficial erosions.[1] The more common distribution seen with vitiligo is that of non-segmental vitiligo, which comprises a symmetrical and bilateral distribution.[6] Its counterpart is segmental vitiligo, which is less commonly seen and manifests unilaterally.[6] Vitiligo has been seen to coexist with cases of lichen sclerosus as a more mild and superficial subtype of LS coined vitiligo lichen sclerosus.[7] This pattern of co-occurrence has been explained through the diseases’ similar pathogenetic mechanism of interface dermatitis along with other concurrent features that have strengthened the link between the two diseases.[7] Despite no definitive cure existing for vitiligo, treatments such as corticosteroids and calcineurin inhibitors are first line, and phototherapy is second line.[6] Recent studies and the FDA approval for JAK inhibitors, such as topical ruxolitinib for nonsegmental vitiligo, has proven to address melanocyte dysfunction through an immune-mediated mechanism.[8] Treatments are most successful for the facial area but overall are unsatisfactory across the body.[6]

Child abuse was also considered. Child abuse is mostly manifested cutaneously.[9] In the clinic, it is important to take a detailed history and match the history to the physical findings. If what is being

reported is not consistent with physical findings, this can indicate abuse.[9] The most common physical findings of abuse are bruises, cuts, burns, bite marks, oral trauma, and traumatic alopecia.[9] Suspicious bruising has been seen most commonly on the head and neck, in addition to areas such as the torso, ear, cheeks, and eye.[10] Other alarming signs of abuse are petechial bruising and multiple bruises in a patterned manner, such as clustered or bilateral.[10] Echymoses and bleeding in the perineal region are features of both sexual abuse and LS, but what separates the two are features in LS such as areas of hypopigmentation, atrophic skin, and telangiectasias.[11]

Thorough physical examination and consideration of differential diagnoses resulted in the correct diagnosis and treatment of our pediatric patient without a painful biopsy.

Treatment and management. The patient and mother were counseled and sent home with clobetasol 0.05% topical ointment. The patient was instructed to apply the ointment to the affected genital area once daily for a month and to follow up for a recheck in 1 month. They were told to contact the office if the lesions failed to improve despite the topical therapy.

Outcome and follow-up. At her subsequent follow-up, her condition had improved significantly with resolution of the clitoral hood edema and purpura and overall improvement of the white plaques. She was instructed to change the application of clobetasol to every other day but to increase to daily again if symptoms of any pruritus and burning returned.

Discussion. Lichen sclerosus is a chronic, inflammatory, mucocutaneous condition. It most commonly presents in the genital area of either prepubertal girls or postmenopausal women.[12] The etiology of LS remains unknown, however, a link between LS and autoimmune diseases remains a strong factor in the diagnosis and understanding of this condition.[13] The clinical characteristics of LS are typically white plaques and areas of atrophic skin in a “figure-of-eight” distribution with potential purpura or ecchymosis if there is associated hemorrhage.[5] Girls with this condition can suffer from complications such as anogenital pruritus, constipation, and painful defecation. The progression of lichen sclerosus, from an area of erythema to subsequent white hyperkeratosis and atrophic skin, can culminate in severe scarring, which may then lead to vulvovaginal stenosis.[5] This can narrow the vaginal introitus and bury the clitoris, consequently leading to dyspareunia.[5] In pediatric patients, the diagnosis is often a clinical one, but biopsy can be performed if the presentation is not clear. In older patients, if the lesions progress without signs of healing, malignancy may ensue.[5]

The first line treatment for this condition is ultrapotent topical corticosteroid ointment daily for 1 to 3 months, and then tapered to every other day as clinical improvement is noted.[14] Aggressive treatment is necessary to prevent irreversible scarring. Patients should be followed clinically every 6 months for at least 2 years, as relapses are common.[14] Calcineurin inhibitors can be used as a second line treatment, in addition to skin care to avoid irritation of the affected area.[5]

Conclusion. In the case of this 8-year-old girl presenting with a pruritic skin eruption that demonstrates glistening white atrophic papules with telangiectasia, purpura, and clitoral hood edema, lichen sclerosus should be strongly considered. Lichen sclerosus can be assessed and

diagnosed based on a thorough history and physical examination, keeping an eye out for the unique clinical features that help differentiate LS from closely related differential diagnoses.

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