Extracutaneous lesions are represented by osteopoikilosis, characterized by small, round or oval circumscribed areas (1–10 mm) of increased bone density at the medullar epiphysis and metaphysis of long carpal and tarsal bones and the pelvis.[8] Cutaneous and skeletal lesions may occur independently.[1,5,6]

Finally, familial cutaneous collagenoma must be differentiated from two distinct types of eruptive collagenoma. Eruptive collagenoma type 1 is clinically and histologically similar to familial cutaneous collagenoma but without documented familial inheritance. Eruptive collagenoma type 2 presents cutaneous lesions smaller than those of familial cutaneous collagenoma and arranged on the extremities and lower trunk. Both types are histologically characterized by the dermal accumulation of metachromatic, mucinous material, suggesting an actual diagnosis of lichen myxedematosus.[1,9]

In summary, familial cutaneous collagenoma appears to be a rare, inherited disorder of collagen that is diagnosed by specific clinical and histologic findings, and must be differentiated from similar inherited conditions.

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Cameo

Lichen sclerosus mistaken for child sexual abuse

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A 3-year-old girl presented to the primary care clinic at King Abdulaziz Medical City (KAMC) with asymptomatic white discoloration of the vulva of 3 months' duration. Dysuria, genital itching, and vaginal discharge followed 2 months later. The discharge was yellowish-greenish in color and occasionally associated with blood-tinged staining of the underwear. The mother had noted that the genital skin changes had worsened in the last 2 weeks and that the genital itching and dysuria had become more frequent. There was no history of trauma, behavioral problems, abnormal sexual behaviour, encopresis, or any underlying disease. On questioning the mother, the possibility of sexual abuse could not be ruled out as she reported that the father was drug and alcohol dependent and behaved inappropriately when under the influence of these factors. Because of this possibility, the girl and her parents were referred to the suspected child abuse and neglect (SCAN) team at KAMC for further evaluation.

The physician, social worker, and child psychiatrist interviewed the girl, her three older brothers, and the parents separately. The girl and her siblings denied any abnormal touching by an adult. The mother reported that she and the father had been separated for 1 year; however,

the couple had been reunited for the last month and were currently living together. The mother linked the appearance of the genital changes to the time the father returned; however, no definite history of sexual abuse was obtained. The father denied any sexual or other abnormal encounter with the child. On examination, the child was well developed and well nourished, cooperative, and showed no abnormal fear of genital or anal examination. Genital examination revealed sharply demarcated, hypopigmented, atrophic plaques symmetrically surrounding the labia majora (Fig. 1). Three sharply demarcated, 0.5 cm, nonpalpable, reddish purpuric macules on the medial aspect of both the labia majora and the base of the clitoris were noted (Fig. 2). Two sharply defined healing erosions were noted on the left side distal to the introitus. The hymenal ring could not be seen, but the hymenal orifice was not dilated. The introitus and the anus appeared normal. Because of the genital findings and the mother's concerns, the possibility of sexual abuse was contemplated. Extensive laboratory testing for sexually transmitted diseases, including syphilis, chlamydia, human immunodeficiency virus, and gonorrhea, were negative. Urine analysis and culture were negative. Routine vaginal culture grew group B β-hemolytic streptococcus, and the patient was treated with amoxicillin-clavulanate orally for 10 days. Due to a lack of evidence of sexual abuse, the SCAN team decided to follow up the child and to investigate further before reporting to the protective agency.

An arranged dermatology consultation concluded that the findings were diagnostic of lichen sclerosus (LS). A skin biopsy obtained from the right labia majora showed atrophic epidermis with follicular plugging. Homogenized eosinophilic amorphous collagen replaced the papillary and upper reticular dermis. These findings were also diagnostic of LS. A 4-week course of twice-daily mometasone furoate 0.1% ointment (Elocom, Scherring, USA) was given with a dramatic resolution of all symptoms. At a 4-week follow-up visit, the signs had greatly ameliorated with the disappearance of bruising and atrophy. Only mild hypopigmentation persisted.

Discussion

Lichen sclerosus (LS) [also known as lichen sclerosus et atrophicus (LSA)] is an idiopathic inflammatory dermatosis of unknown etiology. Signs of LS may include genital bruises, which are very alarming to parents and healthcare workers. Frequently, this leads to sexual abuse investigations which are very stressful to all parties. We have reported one case of such an occurrence and have reviewed the current literature.

Lichen sclerosus is most commonly seen in postmenopausal women, with only 10–15% of all cases occurring in children.[1] The disease is 10 times more frequent in females.[1] The anogenital region is most commonly affected, but extragenital disease can be seen in the upper part of the trunk, forearms, neck, and face.[2] Extragenital involvement is extremely rare in children.[3] Anogenital disease tends to be more severe in adults, resulting in scarring and stenosis.[3]

The etiology of LS is unknown, but several theories have been postulated. The occasional findings of organ-specific autoantibodies in patients with LS suggest an autoimmune phenomenon.[4,5] The more frequent occurrence of autoimmune diseases in patients with LS lends support to this theory.[6] A genetic basis for the disease has also been suggested and supported by familial occurrences.[7,8] A possible role of low serum and tissue levels of androgens has been suggested in postmenopausal women.[9] The rarity of LS in very young children has been attributed to the protective effects of maternal estrogens, which last up to 2 years of age.[10]

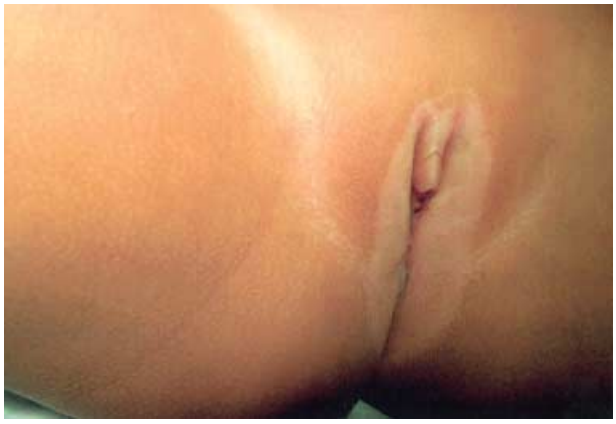

Figure 1 Photograph of the vulva showing atrophic, hypopigmented, sharply defined plaques

Itching is the main symptom of LS and the severity can be variable.[11] Other symptoms may include vulvar soreness, dysuria, chronic constipation, and recurrent ecchymoses and blood-staining of the underwear.[3,12] On examination, the initial lesions of anogenital LS are flat-topped, ivory white

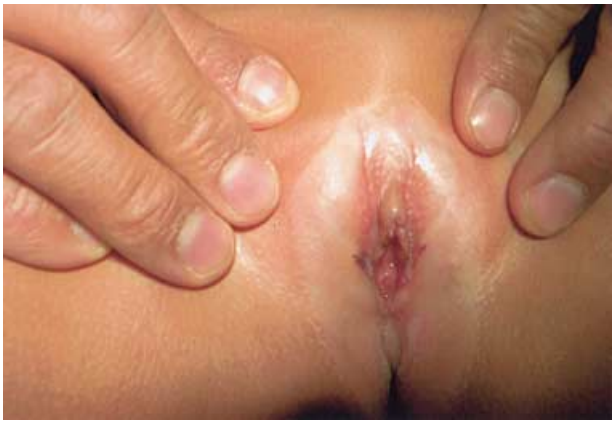

Figure 2 Photograph of the vulva showing three sharply demarcated purpuric macules on both the labia majora and the base of the clitoris

papules that coalesce to form homogeneous, hypopigmented, sharply demarcated, atrophic, and, occasionally, telangiectatic plaques, producing a "figure-of-eight" pattern encircling the vulva and anus.[1,3] The vagina and the hymen are not involved.[3] Other rare presentations of LS include labial fusion, ecchymoses, and genital atrophy.[11,13]

When LS affects the vulva of prepubertal girls, the vulvar skin becomes thin, fissured, and easily traumatized by minimal pressure and friction, resulting in bruising or bleeding.[1] This appearance may be mistaken for trauma and, in particular, for sexual abuse, which may lead to false accusation and unnecessary stressful investigations.[1] In contrast with sexual abuse trauma, the hymen is not involved in LS.[1,3] Although it is important to recognize LS when it occurs in children (in order to avoid the misdiagnosis of sexual abuse), it must be understood that the two conditions are not mutually exclusive diagnoses.[14–18] As LS is one of the diseases that tends to occur at sites of trauma (also referred to as koebnerization), some authors have speculated that sexual abuse-related trauma and infections might act as a trigger to induce or aggravate LS.[14,15] This has been emphasized by many authors.[16–18] In one retrospective review of 42 children with an established diagnosis of LS, evidence of sexual abuse was found in 12.[10] When indicated, full laboratory and multidisciplinary assessment to rule out sexual abuse is mandatory. The presence of hymenal trauma is an important marker for sexual abuse, regardless of the presence of LS.[19] In our patient, the genital findings, marital conflict, and father's addiction induced the mother to suspect sexual abuse by the father. Because the primary care physician could not explain the findings, the assistance of the suspected child abuse and neglect (SCAN) team was rightly requested. Work-up did not lead to any findings supportive of sexual abuse and the diagnosis of LS was confirmed; therefore, the patient was not reported to the legal authorities. Alleged sexual abuse cases are more frequently seen amongst couples who are in the process of separation or divorce. These cases are generally more difficult and time-consuming for the pediatrician and the child protective service system. This is due to the fact that the diagnosis of sexual abuse depends largely on the history as, in most instances, physical signs and symptoms of abuse are lacking.[20]

The differential diagnosis of LS is limited because of the distinct constellation of signs and symptoms. It includes vulvovaginitis, psoriasis, contact dermatitis, and sexual abuse trauma. The clinical appearance is usually sufficiently characteristic to make skin biopsy unnecessary in most children;[3] however, skin biopsy can be performed to confirm the diagnosis of LS, when necessary.

Symptomatic treatment with emollients and antipruritic medications, such as oral sedating antihistamines, may be used to control pruritus.[21] Many specific treatment modalities have been used for LS. Early and aggressive treatment with potent topical steroids seems to be the most favorable and effective.[22,23] This treatment usually alleviates symptoms rapidly and effectively.[22] The British Association of Dermatologists (BAD) guidelines for the management of LS recommend the ultrapotent topical corticosteroid ointment clobetasol propionate.[24] The recommended regimen is initially once a night for 4 weeks, then on alternate nights for 4 weeks, and for the final third month, twice weekly.[24] Potential local side-effects of this treatment include atrophy and telangiectasia, but these are surprisingly uncommon in children when used in short courses.[23] Children treated with this modality should be closely followed on a weekly or biweekly basis.[25] In general, extragenital lesions are not as responsive as genital disease to potent topical steroids.[26]

Although used in adult LS, the use of topical testosterone is not encouraged, especially in young girls, because of its virilizing adverse effects.[1,27] Moreover, its beneficial effect has not been well documented and, in one study, its effect was no better than emollient.[28] Topical progesterone has been reported to be effective in a limited number of pediatric cases.[29]

Topical retinoids have been reported to be effective in adult women, but their irritation potential probably limits their use in children.[27,30] Bracco *et al.*[31] compared topical testosterone 2%, progesterone 2%, and clobetasol propionate, and found that topical steroid was the treatment of choice. The superiority of topical clobetasol propionate over topical testosterone has been documented by other authors.[32]

The course of LS in children has been described as "self-limiting," with the disappearance of signs and symptoms at puberty when a high estrogen state is reached.[22] Helm *et al.*[33] reported the resolution of LS in 44% of their patients. This favorable prognosis of LS, however, has been challenged by many authors.[22,25] Powell and Wojnarowska[22] noted the persistence of the signs and symptoms of LS in a majority of children when followed until puberty. If left untreated, LS may

lead to the loss of the vulvar architecture.[3] The malignant potential of genital LS in adults is well established and estimated to be 5%; however, the cancer risk in children is probably insignificant.[1]

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