Simultaneous Occurence of Vulvar and Mammalian Lichen Sclerosus: A Case Report

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Simultaneous occurence of vulvar and mammalian lichen sclerosus was described. Typically present with symptoms of itching and soreness in the vulvar area at vulvar evaluation reveals a specific appearance. In this case report we encountered an unusual initial presentation of this disease.
Any skin site may be affected but lichen sclerosus is most common in the anogenital area, where it causes intractable itching and soreness. There is increased risk of developing vulvar cancers. Patients should be kept under long term review. The underlying cause is unknown, but there seems to be a genetic susceptibility and a link with autoimmune mechanisms.
Lichen sclerosus et atrophicus confined to the vulva and mammalia is uncommon. We report the case of a 38-year old primer infertile women with a 10 year history of a white patch on her vulva and bilateral mammalia.
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Key Words: Lichen sclerosus, Vulva, Mammalia

Lichen sclerosus is a chronic inflammatory skin disease that causes substantial discomfort and morbidity, most commonly in adult women.The first report of lichen sclerosus was by Hallopeau in 1887. Many terms have been used for the disorder but the International Society for the Study of Vulvovaginal Disease favours the term lichen sclerosus.[1]

The mean age of onset is the fifth to sixth decade in women.[2,3] In women,the disorder is most common when endogenous estrogen production is low, which led to attempts to confirm a protective effect of estrogen.[4] The exact prevalence of lichen sclerosus is unknown. Affected patients may have no symptoms and those with symptoms may be too frightened or embarrassed to seek help.

Lichen sclerosus most commonly affects the anogenital area (85-98% of cases), with extragenital lesions in 15-20% of patients.[5] Genital lesions may appear as a figure-of-eight pattern around the vulva and anus. Patients report intractable pruritis and soreness of the vulvar and perianal areas. Observed skin changes include areas of pallor, which may be small polygonal patches or large plaques. Hyperkeratosis and areas of sclerosis may occur.

Studies of large groups of women with lichen sclerosus[3,6] have shown that the risk of squamous-cell carcinoma (SCC)of the vulva is 4-5%. The reported incidence of carcinoma may depend on the throughness of the histological examination. The degree of risk of carcinoma in diagnosed and treated lichen sclerosus is unknown.

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The extent of extragenital lesions varies, from single small, well-defined areas to widespread scattered lesions. Areas most commonly affected are the inner thighs, the submammary area.

Familial lichen sclerosus has been reported has been reported in identical twins and non-identical twins, sisters, mothers and daughters.[7] Also the association between lichen sclerosus and autoimmunity has been shown in several studies.[8,9] Several infective agents have been linked with lichen sclerosus,including pleiomorphic and variably acid-fast bacilli,[10] although that finding was not substantiated by later studies.[11]

The patient should be aware that as yet there is no cure for lichen sclerosus, but treatment can be offered to relieve symptoms. Long-term follow up is advisable because of the small association with malignant disorders. Potent topical steroids g ve relief from symptoms.The steroid may be combined with atopical antibiotic in the short term if infection is suspected or proven on culture. Androgens may alter the clinical and microscopic appearance of the disorder. Vulvectomy is no longer indicated if there is no tumour.

Case Report

We report the case of a 45 year old postmenopausal woman with a 10 year history of a white patch on her submammalian area and vulva. Her main concern was its cosmetic appearance and the possibility of further spread.

She denied any long term history of vulvar itching or irritation Her menses were normal with no complaints of dysmenorrhea.Onset of menarche and pubertal development were also normal. On her reproductive history primer infertility was the main concern. She was diagnosed with labial adhesions and itching and mammalian soreness.Her first course of treatment included topical steroid therapy for 8 weeks. Her second course of therapy included topical tes-

tosterone for 6 weeks without any improvement or side effects.Punch biopsy was consistent with diagnosis of lichen sclerosus.

We performed the biopsy on both mammalian and vulvar lesions with punch biopsy device under local anesthesia.Routine tissue processing was performed after punch biopsy material was fixed in 10% formalin solution for 24 hours. The sections in 5 micrometers thickness were prepared and the sections were stained with haematoxylen and eosine. Prepared sections were evaluated on the light microscope.

Clinical Features

The patient was medically fit and not taking any medication. There was no family history of similar lesions. Examination revealed a macular white lesion affecting submammaliar area (Figure 1). Clinically it may be difficult to differentiate the lesion from telengiectasia, sexual abuse and infection. Patient was have dysuria, dyspareunia and pain on defecation. There was hyperkeratosis and areas of sclerosis on the vulva (Figure 2) and polygonal atrophic plaques on the submammaliar area.

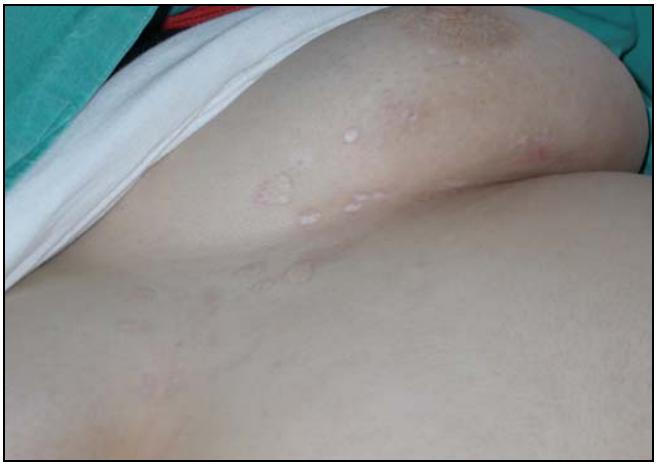

Figure 1. Macular white lesion affecting submammaliar area

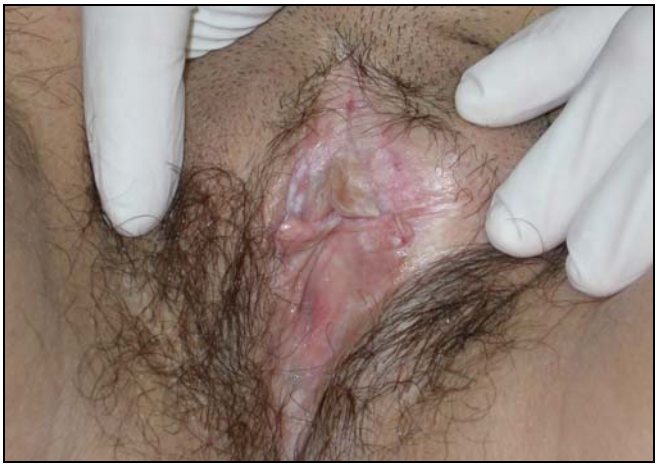

Figure 2. Hyperkeratosis and areas of sclerosus on the vulva

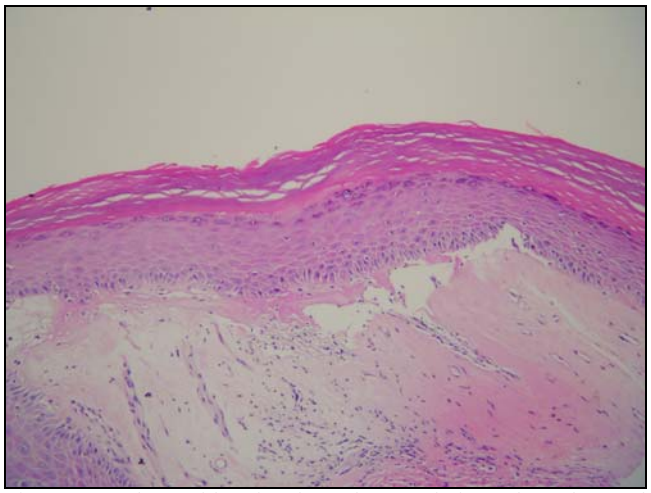

Figure 3. The epidermis that shows obvious hyperkeratosis becomes thinner in vulvar skin and it must be noted that dermis that shows homogenisation, edema and infiltration of focal inflammatory cells detached from the epidermi

Histologic Features

The classic histological features are hydropic degeneration of the basal cells and a pale-staining homogeneous zone in the upper dermis, below which is a bond of inflammatory cells that are mainly monocytic (Figure 3). Squamous epithelium of vulva was relatively thinner than adjacent squamous epithelium. Dermal areas showed the stratification finding consisted of edema in upper dermal layer, a homogenious appearance in the middle dermal areas below the edematous areas and chronic lymphoplasmosytic inflammatory in deep dermal areas. Inflammatory changes involve all zones of the skin. The presence of increased number of mast cells could explain the inflammation.

Discussion

Onset of lichen sclerosus has been reported at all ages, although it is not common under 2 years old.Lichen sclerosus is more common in women than in men.[2,3,6] No association with age at menarche or menopause, pregnancy, hysterectomy or use of oral contraceptives or hormone replacement therapy was found.

Affected patients may have no symptoms and those with symptoms may be too frightened or embarrassed to seek help.Patients refered to hospital may be seen by specialist in dermatology, gynecology, urology and genitourinary medicine. In a comparison of women with and without the disorder, Sideri and colleagues[12] showed no difference in sexual habits, smoking history, education or dietary habits though the unaffected group had a higher dietary carotenoid intake.

The association between lichen sclerosus and squamous cell carcinoma has been known for decades but whether lichen sclerosus is actualy premalign is not certain. However most clinicians believe that the risk is high enough to war-

rant long-term follow-up of such patients[13] including biopsy of any clinically suspicious lesions.

The patient should be aware that as yet there is no cure for lichen sclerosus, treatment can be offered to relieve symptoms. Long term follow up is advisable because of the small association with malignant disorders. Potent topical steroids give relief from symptoms. Androgens may alter the clinical and microscopic appearance of the disorder. Vulvectomy is no longer indicated if there is no tumour.

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