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Vulvar Lichen Sclerosus et Atrophicus

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Abstract

Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis characterized by ivory-white plaques or patches with glistening surface commonly affecting the vulva and anus. Common symptoms are irritation, soreness, dyspareunia, dysuria, and urinary or fecal incontinence. Anogenital lichen sclerosus (LS) is characterized by porcelain-white atrophic plaques, which may become confluent extending around the vulval and perianal skin in a figure of eight configuration. Thinning and shrinkage of the genital area make coitus, urination, and defecation painful. LS is not uncommon in India and present as an itchy vulvar dermatosis which a gynecologist may mistake for candidal vulvovaginitis. There is often a delay in diagnosis of VLS due to its asymptomatic nature and lack of awareness in patients as well as physicians. Embarrassment of patients due to private nature of the disease and failure to examine the genital skin properly are the other reasons for delay in diagnosis. There is no curative treatment for LS. Various medications available only relieve the symptoms. Chronic nature of the disease affects the quality of life. Proper and regular follow-up is required as there are chances of the development of squamous cell carcinoma.

KEYWORDS: *Dermatosis*, *lichen sclerosus et atrophicus*, *vulva*

INTRODUCTION

Vulva is the most visible female genital structure, but it has received the least attention in the medical literature and has been referred to as “the forgotten pelvic organ.”[1]

Lichen sclerosus (LS) is also known as lichen sclerosus et atrophicus (LSA), balanitis xerotica obliterans in males, Csillag's disease, lichen albus, hypoplastic dystrophy, white spot disease, and kraurosis vulvae.

LS most commonly affects anogenital region with 85%–98% cases while extragenital LS can be seen in 15%–20% cases.[2]

Vulvar lichen sclerosus (VLS) is a chronic inflammatory dermatosis most commonly in women that causes substantial discomfort (intractable itching, soreness, constipation, and dyspareunia) and morbidity (introital narrowing, burying of the clitoris, and atrophy labia minora).[2]

There is a bimodal age distribution in the incidence of LS. It occurs in females with an average age of diagnosis of 7.6 years in girls and 60 years old in women while average age of diagnosis in boys is 9–11 years old.[3] Women are more commonly affected than men with 10:1 ratio, particularly during menopausal age group, but younger women or girls may also be affected.

HISTORY

In 1887, Hallopeau first described LS and used the term “lichen plan atrophique” for it which was followed as histological description by Darier who termed it “lichen plan scléreux.” Over the years, many terms were used to describe this condition such as kraurosis vulvae, hypoplastic dystrophy, weissflecken dermatose, and lichen scléreux et atrophique, which now has been accepted by the International Society for the Study of Vulvovaginal Disease.[4]

PATHOPHYSIOLOGY

Inflammation and altered fibroblast function in the papillary dermis leads to fibrosis of the upper dermis. Hypoxia, ischemia, and vascular damage are due to increased GLUT1 and decreased vascular endothelial growth factor (GF) expression in affected skin.[5] The effect of cell-mediated cytotoxicity has also been defined.[6] Although many authors have described LS and scleroderma as closely related entities or even their associations have been seen, there is no systemic involvement in LSA.

ETIOLOGY

The exact etiology of LSA is not known, but several theories have been postulated.

Genetic

LS, both genital and extragenital, has no known racial predilection. A genetic predisposition, based on family clustering, is apparent. Higher rates of LS have been reported among twins[7] and family members.[8] A significant association with human leukocyte antigen Class II antigen DQ7 has also been demonstrated.[9]

Autoimmunity

A strong association with autoimmune disorders has been reported in 21.5%–34% patients and up to 74% of patients found to have autoantibodies.[10] The common disorders associated with LS are alopecia areata, vitiligo, thyroid disease, diabetes mellitus type 1, and pernicious anemia.[11] Sherman *et al*.[8] reported that more than 40% of VLS and lichen planus patients have reactive T-cells to NC16A domain of bullous pemphigoid antigen 180; however, other studies showed that level of autoantibodies is poorly correlated to disease activity and response to treatment. Women with VLS have a higher rate of associated autoimmune disease, especially for autoimmune thyroid disease, compared with men.

Infection

LS is not contagious, but both bacterial and viral pathogens such as human papillomavirus (HPV) and hepatitis C have been implicated in its etiology and many with conflicting results. A link with Lyme disease is shown with the presence of *Borrelia burgdorferi* in biopsy tissue.[10]

Hormones

Higher incidences of VLS in postmenopausal women and prepubertal girls with a low estrogen level suggest a hormonal influence, but a protective effect from estrogen has not been shown.[10] Androgen-sensitive fibroblasts in the vulval skin are responsible for sclerosis. At menarche, there is increased metabolism of testosterone in genital skin which may be responsible for the spontaneous improvement in childhood VLS.[3] Oral contraceptives in premenopausal women have a relative risk of 2.5, showing altered hormonal axis as a possible contributory factor.[12] LS may occur in association with other inflammatory conditions such as psoriasis.[13]

Local skin changes

LS can rarely be initiated through scarring or radiation.[14] Local factors such as friction, trauma, or rubbing may cause Koebner phenomenon.[15] Oxidative stress may also be responsible for sclerosis, autoimmunity, and carcinogenesis in case of LS.[16]

Cell kinetics

An elastase-type enzyme produced by vulvar fibroblasts leads to the destruction of connective tissue in patients with VLS though there appears to be active regeneration with significant collagen synthesis.[17] Keratin differentiation markers using specific monoclonal antibodies, keratins 6 and 16 are associated with increased cell turnover in LS, consistent with a hyperproliferative state,[18] confirmed by flow cytometry analysis.[19] Vulvar skin affected by LS has a wide range of proliferative capacity.[20] The p53 and proliferating cell nuclear antigens are altered in VLS, resulting in changes of the epidermal cell proliferative capacity. Antibody formation against extracellular matrix protein 1 may contribute to disease progression[21] seen in roughly 75% of women with VLS.

The lack of correlation between the duration of symptoms and histologic appearance is consistent with an ongoing inflammatory process with the involvement of activated Langerhans cells. Increased numbers of CD1a-positive Langerhans cells have been found in the epidermis in all stages of LS. The persistently abnormal peripheral lymphocyte pattern may indicate that topical steroids improve symptoms by impeding the action of pruritic factors such as prostaglandins and leukotrienes, not by blocking the local inflammatory process.[22]

Clinical Features

VLS is one of the chronic inflammatory (lymphocyte-mediated) dermatoses with a prevalence estimated to range from 1 in 300 to 1 in 1000 of all patients referred to dermatology departments. It often remains undetected for years.

Dysuria and difficulty in voiding can occur, especially when there is a fusion of the labia minora over the urethra with advanced disease. Nine percent of cases may be asymptomatic.[10] Others present with symptoms such as intractable pruritus which is worse at night, pruritus ani, irritation, soreness, dyspareunia, dysuria, and urinary or fecal incontinence. There may also be thinning and shrinkage of the genital area that makes coitus, urination, and defecation painful. On sexual intercourse or defecation, painful skin fissures develop. Painful defecation, anal fissures, and rectal bleeding are common complaints that require extensive gastrointestinal evaluation and sometimes hemorrhoidectomy or repair of an anal fissure.

Dyspareunia is often a late symptom associated with introital stenosis, fissures, or posterior deflection of fused labial tissues. Fusion over the clitoris can also cause diminished sexual sensation or even anorgasmia. Marked dyspareunia may occur in peri- or post-menopausal women with estrogen deficiency in addition to LS.

LS commonly affects the vulva and around the anus with ivory-white plaques or patches with glistening surface. The lesions occur on the inner aspects of labia majora, labia minora, and clitoris while perianal lesions occur in 30% of cases.[3]

It usually begins as white, polygonal papules that coalesce into plaques. Evenly spaced dells or comedo-like plugs correspond to obliterated appendiceal ostia which may be easily identified with dermoscopy. With time, the plugs and dells will disappear and leave a smooth, porcelain-white plaque. The size of the plaque or plaques may vary widely and from a few millimeters resembles lichen nitidus.

Anogenital LS is characterized by shiny porcelain-white atrophic plaques, which may become confluent extending around the vulval and perianal skin in a figure of eight configuration.[3] Atrophic plaque may have a cellophane paper-like texture, wrinkled, and fragile surface which is associated with telangiectasia, purpura, erosions, fissuring, or ulceration.[23]

Involvement of labial, perineal, and perianal areas along with introital narrowing is referred to as “keyhole” or “hourglass” or “figure of eight.” Genital mucosal involvement does not occur, i.e., the vagina and cervix are always spared. However, some mucosal involvement at the edge of mucocutaneous junctions may lead to introital narrowing.[24] Atrophy can lead to loss of labia minora, burying of the clitoris, obstruction of urinary outflow, or other architectural changes.[4] VLS may progress to gradual obliteration of the labia minora and stenosis of the introitus.

The most common variation occurs when the inflammation is intense enough to cause separation of a large area of epidermis, creating blisters, or large, occasionally hemorrhagic, bullae. As this occurs more often in genital cases, it may be confused with the trauma of sexual abuse or other genital ulcerative disease.

Advanced disease severely affects the quality of life (QoL) and is associated with increased risk of vulvar squamous cell carcinoma (SCC) with 4%–5% risk. Extragenital lesions may occur in 10% of cases. Common extragenital sites are trunk, sites of pressure, upper back, wrists, buttocks, and thighs.[24]

Differential Diagnosis and Workup

Naswa and Marfatia[25] have administered a physician clinical score on the basis of clinical symptoms of erosions, hyperkeratosis, fissures, agglutination, stenosis, and atrophy. They graded them as Grade 1 - no changes or normal, Grade 2 - moderate, and Grade 3 - severe. The proper and timely usage of clinical score helps in early diagnosis and prompt treatment of the disease and will help alleviate symptoms, prevent architectural damage, and reverse histologic changes. It is a handy tool to diagnose and evaluate the progression of the disease. According to Naswa and Marfatia, it cannot replace confirmation of VLS by histopathology, so biopsy is still mandatory in doubtful cases.[25]

An autoimmune workup such as antinuclear antibodies, Vitamin B12 level, thyroid profile, and *Borrelia* antibodies is as such not recommended but should be done in selected cases. Imaging studies can be done to rule out any urinary obstruction, but noninvasive intravenous pyelogram can be done. Biopsy is required to rule out any malignancy in cases with nonhealing ulcer or mass.

Clinicians must also remain astute to the possibility of coexisting bacterial or fungal infections. Investigative studies (e.g., microscopy or culture) to diagnose infection should be performed if signs suggestive of vulvar or vaginal infection are present. *Candida* infection is not uncommon in VLS patients who are taking estrogen and progestin preparations (e.g., contraception and replacement therapy) or who are diabetic. Anogenital LSA closely resembles vitiligo, lichen simplex chronicus,

genital lichen planus, cicatricial pemphigoid, vulvar intraepithelial neoplasia, anal fissures or hemorrhoids, and extramammary Paget's disease. It needs to be differentiated clinically, but biopsy is required to confirm the diagnosis.

Premature ovarian failure should be considered in a woman with hot flashes and oligomenorrhea or amenorrhea before age 45. Estrogen deficiency can lead to a thinned epidermis, labial adhesion, and dyspareunia. These changes should respond to topical estrogen within 2 weeks of treatment. Failure of a response should prompt a vulvar biopsy to exclude LS.

Characteristic histopathological changes show lichenoid infiltrate in the dermal-epidermal junction compact hyperkeratosis with follicular plugging, atrophy of stratum malpighii with hydropic degeneration of basal cell, flattening of rete ridges, pronounced edema, and homogenization of collagen in upper dermis with lymphocytic infiltrate in mid-dermis. Remarkable edema in the papillary dermis is replaced by a dense, homogenous fibrosis as the lesion matures. Extensive and deeper biopsies may show areas more consistent with scleroderma than classic LS.[26]

Epidermal hyperplasia or dysplasia associated with LS on vulvar biopsy specimens is associated with an increased risk of malignant transformation. Overexpression of wild-type p53 is also associated with increased cancer risk as it is a HPV-associated increase in p16INK4A.[27]

Dermatoscopy shows white chrysalis-like structure which correlates with homogenization of collagen in the dermis seen only in late lesions. Comedo-like opening is predominant in early lesions, indicating follicular plugging. White structure-less areas indicate hyperkeratosis and epidermal atrophy; telangiectasia and dotted vessels represent atrophic epidermis with dilated blood vessels.[26]

TREATMENT

There is no definitive cure for LS.[28] Behavior change, such as good hygiene and minimizing scratching, is an important part of treatment,[29] so more realistic goal is to control pruritus rather than resolution of the lesion. Various treatment modalities have been tried with varied results.

TOPICAL TREATMENT

Hormonal therapy

- Estrogen is an effective treatment for postmenopausal vulvovaginal atrophy and should be considered in women with dyspareunia, labial fusion, or epidermal thinning due to estrogen deficiency but not as a primary therapy of LS. Moisturizers and estrogens help in dryness and atrophy
- Topical testosterone (2%) and progesterone (2%) were mainstay of treatment for decades and were reported to induce remission of LS. It acts by reducing inflammation and helps relieving symptoms and in some cases, resolves the lesion, but androgenic side effects such as clitoral enlargement, hirsutism, acne vulgaris, and amenorrhea are common.

Topical steroids

VLS responds to ultrapotent topical corticosteroids, i.e., clobetasol propionate or mometasone furoate, though the clinical appearance does not reverse, but patient gets symptomatic relief and it prevents scarring. Clobetasol or halobetasol propionate 0.05% ointment is a Class I superpotent topical steroid which suppresses mitosis, increases the synthesis of proteins, decreases inflammation and cause vasoconstriction. It is given daily at night for 6–12 weeks and then one to three times per week for maintenance. Treatment is usually prolonged and short-term treatments lead to suboptimal control of

symptoms. Potent steroids in pulse dosing can be given, i.e., 2 consecutive days/week with mild steroids or simple emollients in between to reduce the side effects. Overuse of superpotent topical corticosteroids may induce atrophy, telangiectasia, and striae as early as 2–3 weeks following daily application. Long-term follow-up of VLS patients has not generally demonstrated steroid-induced changes as the modified mucous membranes of the vulva, labia, clitoris, and thickened or lichenified skin are somewhat steroid resistant but the groin fold creases, hair-bearing skin of the labia majora, and perianal skin need to be monitored carefully for steroid-related problems.[30]

Females should be evaluated regularly to see for skin atrophy or any malignant change. Prepubertal LS in girls may resolve spontaneously although some of them may suffer from various types of vulvodynia in adulthood.

Topical calcineurin inhibitors

Tacrolimus (0.1%) and pimecrolimus (1%) have a role as maintenance therapy but not as effective as potent topical corticosteroids and may be useful as alternative treatment options. It reduces itching and inflammation by suppressing the release of cytokines from T-cells and inhibits transcription for genes that encode interleukin (IL)-3, IL-4, IL-5, granulocyte-macrophage colony-stimulating factor, and tumor necrosis factor-alpha, which are involved in the early stages of T-cell activation. It also inhibits the release of preformed mediators from skin mast cells and basophils and downregulate expression of FCeRI on Langerhans cells. Pimecrolimus is derived from ascomycin, a natural substance produced by *Streptomyces hygroscopicus* var. ascomyceticus which selectively inhibits the production and release of inflammatory cytokines from activated T-cells by binding to cytosolic immunophilin receptor macrophilin-12. The resulting complex inhibits phosphatase calcineurin, thus blocking T-cell activation and cytokine release.[31]

In a 12-week randomized trial of 38 patients with VLS that compared pimecrolimus 1% cream to clobetasol 0.05% cream, both significantly reduced patient symptoms and histopathologic inflammation.[32] Patients with VLS, who are refractory to topical corticosteroid have exhibited improvement with pimecrolimus[33] or tacrolimus.[34] Tacrolimus causes burning sensation so is often discontinued by patients. Although the US Food and Drug Administration has issued a “black box” warning for topical calcineurin inhibitors due to risk for internal malignancy, but definitive relationship has not been established, i.e., the impact of topical calcineurin inhibitor therapy on risk for vulvar SCC in patients with LS is unknown.

Topical retinoids

Topical tretinoin (0.025%) and tazarotene (0.01%) have off-label indication in LS. They act by downregulation of fibroblast function. Especially in genital areas, short-contact therapy is used in which the gel is initially applied for 15 min and washed off. Every 2–3 weeks, application time is increased by 15 min until therapeutic effects are noted.

In an observational series, topical use of tretinoin improved the symptoms, gross appearance, and histopathologic features of LS with minimal side effects.[35]

Vitamin D analogs

Calcitriol, calcipotriene, and calcipotriene plus betamethasone helped some patients with localized sclerotic diseases. They can be irritating over genitals which require alternate-day therapy and should be used with caution in patients having compromised renal function.[28]

Topical TRPM8

There is a single case report of the use of topical icilin in LS with relief in pruritus. Icilin is a TRPM8 receptor antagonist similar to menthol but with a higher affinity to the TRPM8 receptor.[36,37]

Topical avocado and soybean extracts

Borghi *et al*.[38] suggest topical avocado and soybean extracts as alternative treatments for mild-to-moderate LS in patients wishing to avoid corticosteroids.

Results in their study provide evidence that the topical and dietary supplements exert anti-inflammatory, antifibrotic, emollient, and lenitive actions and are effective alternatives in the treatment of mild-to-moderate VLS.

Topical oxatomide

It helps relieve pruritus through its antihistamine effects, but the course of disease is not affected.[39]

Intralesional therapy

Injection of triamcinolone acetonide 20 mg directly into the thickened hypertrophic plaques of VLS once per month for 3 months[40] after topical anesthetic help to minimize patient discomfort. Intralesional injection of adalimumab has also found to be beneficial.[28]

Lights and Lasers

Phototherapy

Narrow-band ultraviolet B and psoralen plus ultraviolet A A single study of ultraviolet A1 (UVA1) in seven women with VLS that had not been controlled by topical steroids[41] reported initial improvement in five patients although two relapsed and the others required ongoing treatment with topical steroids. Studies have compared ultrapotent topical corticosteroids with the calcineurin inhibitors showing more efficacy of clobetasol propionate which works better compared with UVA1 phototherapy.

Photodynamic therapy using a photosensitizer Successful treatment of VLS with photodynamic therapy (PDT) has been reported.[42] In an open study of ten patients treated with two sessions of PDT, all patients reported some improvement in symptoms of VLS (itching, burning, and pain).[43]

In an open study of PDT for VLS (topical 20% 5-aminolevulinic acid, argon laser light, and one to three treatments), 10 of 12 patients derived significant improvement.[44]

It caused significant burning although itching improved in 8 of 12 women. Another study demonstrated good symptomatic benefit in six of ten patients treated with aminolevulinic acid-PDT using a bioadhesive patch.[45]

Cryotherapy

Cryotherapy of affected genital lesions after one or a series of treatments shows improvement. In one small study of 12 patients with VLS and severe intractable itch, 75% obtained symptom relief with cryotherapy.[46]

Laser

Tissue-vaporizing carbon dioxide lasers, nonablative lasers such as the pulsed dye, and erbium-doped yttrium aluminum garnet lasers have been reported to benefit LS symptomatically but did not stop the disease recurring. There is a report showing benefit in 17 out of 31 cases of untreated LS using frequencies of 5–8 MHz with focused ultrasound.[47]

Systemic Therapy

Retinoids

Retinoids appear to reduce connective tissue degeneration in LS. However, the use of these agents is limited by significant and potentially harmful side effects including cheilitis, xerosis, teratogenicity, elevated liver enzymes, hypertriglyceridemia, abdominal pain, and alopecia.

Isotretinoin decreases sebaceous gland size and sebum production. It may inhibit sebaceous gland differentiation and abnormal keratinization.

Oral acitretin (20–30 mg/day for 16 weeks) was effective in one randomized trial.[48] Doses of 8–30 mg daily for 4 months have been used which gave benefits both in symptoms and also induce resolution of lesions. Mechanism of action of systemic retinoids in LS particularly in genital LS is not clear; mostly, it acts by downregulation of fibroblast function.

Hydroxychloroquine

It is reported to be effective in widespread genital and extragenital LSA, with conflicting results.[28]

Hydroxycarbamide (hydroxyurea)

Hydroxycarbamide is an antineoplastic drug used in myeloproliferative disorders. It inhibits T-lymphocyte proliferation and gamma interferon production and has antiretroviral properties. It is used in LS in dose of 1 g daily reducing pruritus and soreness in 6 months.[49]

Cycloferon

Cycloferon is a low molecular weight interferon-inducing substance which has antiviral, immunomodulating, and anti-inflammatory effects. A prospective randomized study involved sixty patients with chronic dystrophic diseases of the vulva (45–65 years); cycloferon given intramuscular on days 1, 2, 4, 6, 8, 10, 12, 16, 20, and 23 was reported to induce rapid remission, improvement of QoL, and psychosocial function.[50]

Cyclosporine

Cyclosporine has been used in only ten patients with VLS.[51] Three patients had a significant improvement, five had some response, and two had no response.

Potassium Para-aminobenzoate

One report of five patients with LS at various sites and resistant to numerous other therapies documented good improvement with potassium para-aminobenzoate (4–24 g daily, in divided doses) in all five.[52]

A small number of women initially have a partial response to medical treatment but have ongoing burning, irritation, and pain. In these cases, obtain cultures to exclude superinfection by *Staphylococcus, Streptococcus*, or *Candida*. The patient may have bacterial cellulitis, vulvar candidiasis, or vaginal candidiasis, which requires treatment with appropriate antibiotics or antimycotic drugs.

Diet

Twenty-three patients also received dietary supplements with Vitamin E and para-aminobenzoic acid showed improvement.[36]

Surgery

VLS can be surgically excised, but mutilating gynecologic surgery is usually not recommended. The rationale behind surgical therapy is primarily to treat those patients who did not respond or responded poorly to medical treatment, secondarily to postinflammatory sequelae and prevent the development of invasive carcinoma of the vulva.

Surgical intervention in LS is done to release a buried clitoris, to separate fused labia, or to widen a narrowed introitus in the case of pain or sexual dysfunction. V-Y advancement flap is an effective method for the reconstruction of perineal region. This technique will allow the expansion of vaginal orifice with good cosmetic results and rapid healing after surgery.

Introital stenosis, posterior fissuring, and scarring at the fourchette can be treated by vulvoperineoplasty.[53] Since vaginal tissue is not affected by LS, part of the vaginal wall is used in the repair to prevent recurrent adhesions and fissuring at the introitus. Adhesions at the fourchette which cause dyspareunia can be extirpated. Adhesions burying the clitoris can result in formation of painful pseudocysts. Clitoral adhesions are released with delicate knife strokes. Reformation of adhesions can be prevented by resection of a fragment of the clitoral hood in the shape of a tricorn.[54]

Other points which need to be considered treating VLS are as follows:

- Menopausal women may have symptoms related to atrophy and dryness, which will respond to topical estrogen cream and moisturizers
- A diagnosis of superimposed vulvodynia should be considered if pain persists despite resolution of pruritus and dermal changes. It is likely that vulvodynia represents neuropathic pain, which is pain arising from abnormal neural activity secondary to disease, irritation, or injury of the nervous system that persists in the absence of ongoing disease or acute injury
- An allergy to the topical medication may be present. A topical steroid with a different base or consultation with an allergy specialist should be considered.

Dermasilk

A controlled, randomized, double-blind study showed that patients undergoing treatment for VLS have fewer symptoms when wearing silk than cotton briefs.[55]

Future Modality of Treatment

Recent studies indicate that the injection of platelet-rich plasma (PRP) and stem cells used along with autologous fat transfer deserves a special mention as a novel technique in the management of LS of vulva. PRP is an effective concentration of multiple fundamental GFs by virtue of platelets alone (stored as α-granules in platelets) as enumerated in and plasma proteins, namely, fibrin, fibronectin, and vitronectin. This cocktail of GFs is pivotal in the modulation of tissue repair and regeneration,[56] whereas the plasma proteins act as a scaffold for the bone, connective tissue, and epithelial migration.

Counseling

According to the National Vulvodynia Association, women with VLS or any vulvovaginal condition experience feelings of isolation, hopelessness, depression, anxiety, anger, and low self-image. Some women are unable to continue working, any physical activity, or sexual relations.

Psychological counseling is very much needed in patients of VLS. Education relating to sexual dysfunction and dyspareunia may be required. Patients should be educated on what changes (e.g., ulceration) might indicate malignant transformation so that immediate consultation can be done.

Counseling involves self-care to be taken by patient themselves such as:

- Avoid washing with soap or to use an emollient soap
- Carefully dry the area after passing urine to reduce the contact of urine or using a moisturizer or soft paraffin as a barrier cream to protect skin from exposure to urine
- If sexual intercourse is painful because of tightening at the entrance to the vagina, use of lubricants and vaginal dilators if required
- Keep an eye on your skin. Regular self-examination is very important. If any skin change develops which does not respond to steroid creams, or there is any skin thickening, soreness, or ulceration lasting more than 2 weeks, consult without delay and get a biopsy done to rule out skin cancer.

COMPLICATIONS

Complications include dyspareunia, urinary obstruction, secondary infection from chronic ulceration, and skin atrophy due to chronic steroid use. As high as 5% for the lifetime risk of vulvar SCC in patients with LS is reported[57] where risk factors include older age, longer duration of LS, evidence of hyperplastic/early vulvar carcinoma *in situ* changes, and coexistent chlamydia infection.

PROGNOSIS

VLS has no associated increased mortality except malignancy at the site and progressive scarring. QoL is affected as it results in severe sexual dysfunction.

CONCLUSION

There is often a delay in diagnosis of VLS due to lack of awareness in patients as well as physicians; it is recommended to examine the female genital properly in cases with any vulvar dermatosis or pruritus at that site in menopausal women. It not only affects the QoL but may also lead to sexual dysfunction and avoidance of sexual intimacy.

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