A Review Article on Lichen Sclerosus, Affecting Ano-Genital Area in Females

Dr. Ahmed M. Lutfi\*

\*Medical doctor, Anatomist and Dermatologist.

MD, M.Sc. (UK)

College of Medicine, Baghdad University.

Baghdad, IRAQ.

Email/ [tesla1452@gmail.com](mailto:tesla1452@gmail.com)

Phone/ +44 (0)7482 571679 (UK)

+964 (0)7714 338199 (IRAQ)

**Abstract**

**Objective:** To collect detailed up-to-date knowledge, using systematic review of the medical literature, that is specific to the Dermatologic condition of Lichen Sclerosus (LS), which mainly affects the ano-genital area of female patients. This article will review: The disease epidemiology, pathogenesis, clinical features, complications, differential diagnosis, diagnostic methods, histopathology, psychosocial impact, and treatment options.

**Background:** LS is a chronic dermatologic condition. The condition was described at the end of the 19th century. LS incidence is higher in females than in males, affecting mainly the female ano-genital area. Diverse and crippling complications exist, including malignant transformation. Clinical features progress towards severe vulvar anatomic distortion, which results in devastating psychosocial implications. Mainstay treatments are both medical and surgical; recent randomized controlled trials promote the use of topical Calcineurin inhibitors.

**Methods:** I used a meticulous strategy across databases, led by pre-specified keywords, followed by the application of database-specific filters to scrutinize the hierarchy of available medical literature, from guidelines, systematic reviews and randomized controlled trials to medical papers with weak evidence. Some articles were requested from the British library.

**Results:** I used around 70 references; the most relevant data were extracted. This article is divided into sections of topics, starting from an introduction to a conclusion.

**Conclusion:** This article will enable the medical researcher to obtain a detailed perspective of the condition LS. Thus, a researcher can seek strongest evidence for his original research.

**Keywords:** Lichen Sclerosus, Ano-genital, females, ECM-1, Borrelia burgdorferi.

**Introduction** (Neill, Lewis, Tatnall, & Cox, 2010; Kirtschig, Baldo, Brackenbury, Lewis, & Wojnarowska, 2011; Thomas, 2009; Goodfield, Jones, & Veale, 2010; Bunker & Neill, 2010; [eMedicine](http://emedicine.com), n.d.; Hallopeau, 1887; Darier, 1892; Abdelbaky, Aluru, Keegan, & Greene, 2012; Warrington & de San Lazaro, 1996)

Lichen sclerosus (LS) is chronic dermatosis with a major predilection for the ano-genital area, mainly affecting peri-menopausal and pre-pubertal females, and extragenital sites can be involved. It was known as Lichen Sclerosus et Atrophicus, but the term “et Atrophicus” was dropped in 1976 (because not all lesions have atrophy). When it affects the male external genitals, it is known as Balanitis Xerotica Obliterans (BXO). Other synonyms of LS include Guttate morphoea, Guttate scleroderma and White-spot disease, which mainly refer to trunk and limb involvement. Hallopeau (1887) was the first to describe LS, while Darier (1892) pioneered LS histology, and both considered LS a Lichen Planus variant. Others thought LS was a localized scleroderma variant; however, LS is a separate entity.

LS in children is commonly mistaken as sexual abuse, while it causes psycho-sexual dysfunction in adults. The mainstay treatment is with super-potent topical steroids.

1. **Epidemiology** (Pinelli, D'erme, & Lotti, 2013; Smith & Fischer, 2009; Wallace & Whimster, 1951; Sherman et al., 2010; Kiss, Kiraly, Kutasy, & Merksz, 2005; Simpkin & Oakley, 2007; Meyrick & Kennedy, 1986)

Female ano-genital LS is commoner than BXO. There is no racial predilection; however, familial clustering exists. A high concordance rate is reported in mono/dizygotic twins. The bimodal age distribution in females is peri-menopausal and pre-pubertal.

1. **Pathogenesis** (Neill, Lewis, Tatnall, & Cox, 2010; Kirtschig, Baldo, Brackenbury, Lewis, & Wojnarowska, 2011; Thomas, 2009; Goodfield, Jones, & Veale, 2010; Darier, 1892; Cox, Mitchell, & Morley, 1986; Günthert, Faber, Knappe, Hellriegel, & Emons, 2008; Taylor, Guzail, & Al-azzawi, 2008; Powell & Wojnarowska, 2001; Oyama et al., 2003; Howard, Dean, Cooper, Kirtshig, & Wojnarowska, 2004; Aberer, Kollegger, Kristoferitsch, & Stanek, 1988; Ansink et al., 1994; Drut, Gómez, Drut, & Lojo, 1998; Nasca, Innocenzi, & Micali, 2006)

LS etiology is unclear, with autoimmune/hormonal/genetics/infectious and other causes incriminated, and the predilection for the ano-genital area is poorly understood (Pinelli et al., 2013). Female predominance with a bimodal age pattern and girls’ remission at puberty/menarche indicate a hormonal influence. Moreover, the oral contraceptive pills-mediated disturbance of androgen-controlled vulvar skin growth might trigger early LS, and estrogen receptor expression is critical (Cox et al., 1986; Goodfield et al., 2010; Günthert et al., 2008; Thomas, 2009).

The presence of a family history of autoimmune diseases/organ-specific antibodies supports autoimmunity (highest at 41–60 years) (Goodfield et al., 2010; Taylor et al., 2008; Thomas, 2009). Two-thirds of LS patients had serum IgG-autoantibodies towards extracellular matrix protein-1 (*ECM-1).* One-third of LS patients had circulating anti-*BMZ* antibodies (*BP180 and BP230*, involved in immuno-bullous dermatoses) (Goodfield et al., 2010; Oyama et al., 2003; Powell & Wojnarowska, 2001; Thomas, 2009). The geographical distribution in Germany/Austria/Japan signifies the role of *Borrelia burgdorferi* (especially in early LS). A prior history of vaginitis/chronic balanitis was also implicated. Human papilloma virus (*HPV*) (types 6, 16, 18, 33, 45 and 51) were reported, especially in penile LS (Aberer et al., 1988; Ansink et al., 1994; Drut et al., 1998; Goodfield et al., 2010; Howard et al., 2004; Kirtschig et al., 2011). Koebnerization occurs in LS (more with extragenital lesions) at the site of mechanical trauma. Penile LS also occurs in hypospadias/hypospadias repair and penile grafts (Darier, 1892; Goodfield et al., 2010; Kirtschig et al., 2011). Chronic ano-genital skin’s exposure to urine and occlusion can be a factor in urine-dribbling males and peristomal LS (mainly urostomies). LS histological changes in flexural-occluded skin tags support the occlusion of flaccid skin’s role (Goodfield et al., 2010; Nasca et al., 2006; Thomas, 2009).

* 1. **Immunology** (Goodfield et al., 2010; Thomas, 2009; Pinelli et al., 2013; Al-niaimi & Lyon, 2013; Regauer, Reich, & Beham-Schmid, 2002; Farrell, Dean, Millard, Charnock, & Wojnarowska, 2006; Strittmatter, Hengge, & Blecken, 2006)

Subepithelial dense lymphocytic infiltrate, with immuno-regulatory and activated *T*-cells with monoclonal *γ-TCR*-rearrangement (Al-niaimi & Lyon, 2013). Changes in other immunological indices also occur (Table 1):

Table 1

*Immunological Changes in LS*

|  |
| --- |
| IgG/IgM/IgA, complement & fibrin are present. |
| IgG-autoantibodies to *ECM*-1 and Bullous pemphigoid antigens (*BPAG1*& *BPAG2*). |
| Skin basement membrane damage with over-expression of *laminin*, *collagen* 4 & 7. |
| Significant lipid peroxidation (mainly in stratum basale) confirmed *ECM-1* role. |
| Reduced manganese superoxide-dismutase makes cells vulnerable to oxidative stress. |
| Cytokine response mimics that of Lichen Planus (LP) & chronic wounds. |
| Increased *TNF-α*, *IL-1*, *interferon-γ*, *TGF-ß*, *CD25*, *CD11α* & *ICAM-1*. |
| Dysfunctional fibroblast & collagen over-production driven by *TGF-ß* exist. |

Sources: Farrell et al., 2006; Goodfield et al., 2010; Oyama et al., 2003; Regauer et al., 2002; Sander, Ali, Dean, Thiele, & Wojanrowaska, 2004; Strittmatter et al., 2006; Thomas, 2009

* 1. **Genetics** (Thomas, 2009;Goodfield et al., 2010;Taylor et al., 2008;Sander et al., 2004; Senturk et al., 2004; Scurry & Cohen, 1998; Prowse, Ktori, Chandrasekaran, Prapa, & Baithun, 2008)

Genetics is highly implicated: Familial clustering/high concordance in monozygotic and dizygotic twins/autoimmune diseases-coexistence and *HLA* (Human Leukocyte Antigen) association (Table 2) (Goodfield et al., 2010; Thomas, 2009). *HLA-DQ7* has predominant LS association (Goodfield et al., 2010; Sander et al., 2004; Taylor et al., 2008; Thomas, 2009).

Table 2

*HLA Association with LS.*

|  |  |
| --- | --- |
| **HLA-type/subtype** | **Notes** |
| HLA-DQ7, 8 or 9 | Found in 78% of LS. |
| HLA-DRB112 | Enhances possibility of vulvar disease. |
| HLA-DRB10301 | Confers disease protection. |
| HLA-DR11, DR12 & DQ7 (in men) | Associated with disease development. |
| HLA-B08 & B18 | Lack of data, interestingly in report on 4 siblings with LS, they had unaffected sister; surprisingly, she lacked these alleles. |

Sources: Goodfield et al., 2010; Sander et al., 2004; Taylor et al., 2008; Thomas, 2009

High *Ki67* expression is found in thickened LS lesions (due to squamous cell hyperplasia or Lichen Simplex chronicus) (Senturk et al., 2004). High *p16INK4A* expression with coexisting *HPV-16* in penile LS confirms *HPV*’s role in interfering with the retinoblastoma pathway (Scurry & Cohen, 1998). Genomic silencing (via hyper-methylation) of the *MGMT* and *RASSF2A* genes was found in vulvar SCC and LS coexisting with SCC, but was absent in isolated LS (Prowse et al., 2008).

1. **Clinical Features** (Neill et al., 2010; Kirtschig et al., 2011; Thomas, 2009; Goodfield et al., 2010;Farrell et al., 2006;Guerrero et al., 2011; Pugliese, Morey, & Peterson, 2007; Ridley, 1987; Neill, Tatnall, & Cox, 2002; Hassanein, Mrstik, Hardt, Morgan, & Wilkinson, 2004; Bussen, 2009; Christman, Chen, & Holmes, 2009; Farrell, Marren, & Wojnarowska, 2000; Bonin, Gubertini, & Trevisan, 2011; Meyrick-Thomas, Ridley, McGibbon, & Black, 1988)

Spontaneous remission usually occurs in pre-pubertal girls. Peri-menopausal women are the prime LS target. Early lesions have erythema with/without hypopigmentation, then form typical porcelain-white ivory macules/papules and plaques. The genito-crural folds/inter-labial sulci/labia minora/labia majora-inner aspect/clitoris/clitoral hood and perineal body are mainly affected. In contrast to Lichen Planus (LP), mucosal involvement does not occur (sparing the cervix and vagina). LS lesions surround the anus and vulva in a figure-of-eight pattern, forming ivory atrophic papules/plaques with follicular hyperkeratosis/plugging; however, LS may resemble macerated intertrigo with a flat and glistening appearance. LS involves scarring (Table 3), vulvar shrinkage and narrowing of the introitus (Figure 1) down to one centimeter.

Table 3

*Factors Contributing to Vulvar Scarring and Shrinkage in LS*

|  |
| --- |
| Atrophy of labia minora (labial resorption). |
| Atrophy of clitoris, clitoral hood with clitoral burial. |
| Fusion of labia minora & labia majora. |
| Narrowed vaginal introitus (due to anterior & posterior labial fusion). |



*Figure 1.* Gross vulvar shrinkage, right image shows typical ivory figure-of-eight pattern (Farrell et al., 2006; Goodfield et al., 2010).

The presence of purpura/ecchymosis/telangiectasia is characteristic due to skin atrophy vs. irritation, humidity and area occlusion. Edema/blistering/hemorrhagic bullae/erosions and ulcerations occur too. The main symptoms are soreness and itching (this is worse at night and disturbs sleeping), pain and dyspareunia (due to erosions/fissuring/narrowed introitus), urinary symptoms and constipation (due to irritation/ano-genital-anatomical distortion). LS can be asymptomatic in pre-pubertal girls; such cases are commonly mistaken as sexual abuse (while sexual abuse can trigger LS via Koebnerization). Extragenital lesions with severe/distressing itching co-exist in 10% of cases, with predominant truncal distribution at pressure sites (possible Koebnerization), and scalp involvement is rare (Figure 2).



*Figure 2.* LS of the scalp with extensive cicatricial alopecia (Thomas, 2009).

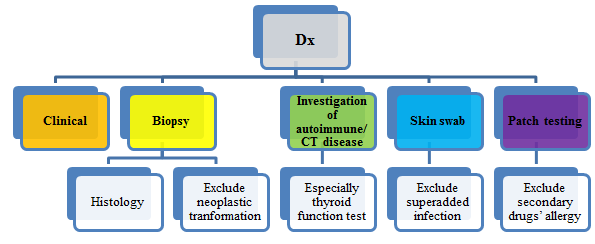
Pre-malignant/malignant transformations occur in LS, and there is a definite link between LS and vulvar Squamous cell carcinoma (SCC), which co-exist in 5% of cases, usually arising in the vulvar-anterior part on top of the longstanding sclerotic area. Oncogenic *HPV* seems to have carcinogenic potential in pre-pubertal females. It is unknown if LS-management reduces carcinogenesis. Other superimposed pre-malignant/malignant variants include: Stratum basale dysplasia/carcinoma in situ/verrucous carcinoma/basal cell carcinoma, while melanoma occurrence is rare.

Coexisting vulvar melanoma in pre-puberty is extremely rare. Hassanein et al. (2004) reported a case in a Caucasian girl in which partial vulvectomy was performed. Bussen et al. (2009) reported junctional melanocytic nevus of the labia minora in a pre-pubertal girl.

1. **Complications** (Farrell et al., 2006; Goodfield et al., 2010;Thomas, 2009;Ridley, 1987;Bussen, 2009):

* Malignant transformations.
* Narrowed introitus.
* Superadded infections.
* Digestive tract and urinary system problems.
* Pseudocyst of the clitoris.
* Psychosexual problems, attributed to Dysesthesia, vestibulodynia and vulvodynia.

1. **Diagnosis (Dx):** (Neill et al., 2010; Kirtschig et al., 2011; Thomas, 2009; Goodfield et al., 2010;Bonin, Gubertini, & Trevisan, 2011;Meyrick-Thomas, Ridley, McGibbon, & Black, 1988)



*Figure 3.* Diagnostic tools in LS.

**Histopathology** (Neill et al., 2010;Thomas, 2009;Goodfield et al., 2010;Regauer & Reich, 2005; Mann & Cowan, 1973; Stanley, Vinay, & Ramzi, 2010**)**

Histopathology is diagnostic (except in the early stages), there is peculiar peri-vascular dense dermal mononuclear/lymphocytic infiltrate (which is initially scanty and focal) hugging the epidermis, the infiltrate becomes deeper, forming a band with the overlying thick sub-epidermal sclerotic/hyaline zone with unique changes in the dermal collagen/elastin (under EM). The sub-epidermis appears edematous/structureless/with dilated capillaries and sparse cells. Initially, there is variable epidermal thickening/hyperkeratosis/follicular plugging. Later, there is epidermal thinning/absent rete pegs/superficial hyperkeratosis/stratum basale-vascular degeneration potentiated with dermo-epidermal clefting. Marked dermal edema (Figure 4) can precede/coexist with sclerotic changes (Goodfield et al., 2010; Kirtschig et al., 2011; Neill et al., 2010; Regauer & Reich, 2005; Stanley, Vinay, & Ramzi, 2010; Thomas, 2009).

If there are adjacent invasive carcinomas, LS frequently transitions from classic atrophy to epithelial hyperplasia and squamous cell atypia (Stanley et al., 2010).



*Figure 4.*Evident dermal edema with lymphocytic infiltrate (Stanley et al., 2010).

1. **Differential Diagnosis (DDx)** (Christman et al., 2009;Farrell et al., 2000;Neill et al., 2010;Thomas, 2009;Goodfield et al., 2010)

Vitiligo/morphoea/LP and mucous membrane pemphigoid may present with clinical similarities or co-exist with LS. Clinical-histological overlap with morphoea/LP/scleroderma (Table 4) may represent the possible disease spectrum. Scalp LS may resemble “en coup de sabre” of localized Scleroderma (Christman et al., 2009;Farrell et al., 2000; Goodfield et al., 2010; Thomas, 2009).

Table 4

*Medical Entities Associated with LS*

|  |
| --- |
| Morphoea |
| Vitiligo |
| Alopecia areata (in males) |
| Pernicious anemia (in females) |
| Limited cutaneous systemic sclerosis |
| Systemic lupus erythematosus |
| Lichen planus |

Source: Thomas, 2009

1. **Psycho-social Impact** (Neill et al., 2010; Kirtschig et al., 2011; Warrington & de San Lazaro, 1996; Mills, 2009; AAFP, n.d.; Shasi, Chapman, Evans, & Jaleel, 2010; Brown, McKenna, Siddhi, McGrouther, & Bayat, 2008; Wikipedia, n.d.; Wehbe-alamah, Kornblau, Haderer, & Erickson, 2012; Evers et al., 2008; Hong, Koo, & Koo, 2008; Koblenzer, 2005)

A psycho-social burden occurs in LS due to: Chronicity, physical-genital disfigurement, sexual implications/dysfunction, associated autoimmune disease/connective tissue diseases and neoplastic transformation risk. Patients have to be comforted regarding the rarity of malignancy and impossibility of infectivity to others (AAFP, n.d.; Mills, 2009).

Emotional wellbeing will be disturbed due to factors related to: Physical comfort and functioning, acceptability to the self and others and confidence in the nature and management of the condition (Shasi et al., 2010).

LS patients can experience isolation/hopelessness/anger/low self-image/depression/anxiety/limited physical activities/work problems and sexual dysfunction. Frustration can emerge from healthcare givers’ defective knowledge, resulting in wrongful diagnosis and prolonged life disruption (Brown et al., 2008; Wikipedia, n.d.). Dalziel was the first to study LS-associated sexual dysfunction. Females reported dyspareunia/sex-avoidance/loss of sexual interest. Van et al. (2010) studied factors associated with sexual dysfunction through the Quality of Life Index (*QoL*), Dermatology Quality of Life Index (*DLQI*), Female Sexual Function Index (*FSFI*) and Female Sexual Distress Scale (*FSDS*). Patients scored low for sexual desire/arousal/lubrication/orgasm/satisfaction. Those with low *QoL* scores experienced more sexual difficulties (age-independent). Another study reported that patients’ worries about sexually transmitting the condition and cosmetic appearance negatively influenced libido (AAFP, n.d.; Koblenzer, 2005; Warrington & de San Lazaro, 1996).

1. **Treatment** (Neill et al., 2010; Kirtschig et al., 2011; Thomas, 2009; Goodfield et al., 2010; Warrington & de San Lazaro, 1996; Kiss et al., 2005; Taylor et al., 2008; Ridley, 1987; Farrell et al., 2006; Brown et al., 2008; Van de Nieuwenhof et al., 2010; Hagedorn, Buxmeyer, Schmitt, & Bauknecht, 2002; Bousema et al., 1994; Carli, Cattaneo, Taddei, & Gianotti, 1992; Gurumurthy, Morah, Gioffre, & Cruickshank, 2012; Fischer & Rogers, 1997; Garzon & Paller, 1999; Lascano, Montes, & Mazzini, 1964; Penneys, 1984; Shelley, Shelley, & Amurao, 2006; Prowse, Ktori, Chandrasekaran, Prapa, & Baithun, 2008; August & Milward, 1980; Hillemans et al., 1999; Kartamaa & Reitamo, 1997)

LS can be a permanent relapsing condition. Spontaneous resolution can occur at puberty and before the age of 30 in extragenital LS (Kiss et al., 2005; Taylor et al., 2008). Patients must receive written information about LS. A multi-disciplinary team approach by a dermatologist/gynecologist/urologist/psychiatrist and vulvar clinic is required (Farrell et al., 2006). Neoplastic risk must be explained, to be encouraged for periodic follow-ups (Brown et al., 2008; Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987).

* 1. **Rx in adult females**

In newly diagnosed cases, super-potent topical corticosteroids, such as *Clobetasol propionate* (*Dermovate)*, are the mainstay treatment, and they are initially applied once-daily at night for 4 weeks, then every other day for the next 4 weeks and 2 times/week for the last 4 weeks (a 30-gm tube is sufficient for 12 weeks). Some argue that Dermovate should be used for 6–8 weeks only (due to side effects, including steroid-induced atrophy). If there is a secondary fungal/bacterial infection, *Dermovate-NN* or *Dermovate* and *Nystaform* are used. Some patients achieve full remission; however, if LS symptoms recur, patients are instructed to use *Dermovate* at the previous effective frequency and when needed (30–60 gm is sufficient for 1 year). Topical steroids may fail in LS (Table 5). Hagedorn et al. reported the use of intralesional steroid injection (Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987; Van de Nieuwenhof et al., 2010).

Table 5

*Possible Causes of Failure of Super-Potent Topical Steroids in LS*

|  |  |
| --- | --- |
| **Causes** | **Notes** |
| Patients’ non-compliance | In elderly/disabled patients. |
| Due to pharmaceutical company warning against use of high-potency steroids in ano-genital area, patients stop Rx. |
| Incorrect diagnosis |  |
| Added/coexisting problem | Medications induced contact allergies. |
| Secondary infection as candidiasis. |
| Intraepithelial neoplasia. |
| Malignancy. |
| Psoriasis. |
| Mucus membrane pemphigoid. |
| LS is treated, but is still symptomatic | Due to secondary sensory problem of dysaesthetic vulvodynia. |
| Sexual dysfunction problems. |
| Mechanical problems/complications | Necessitates surgery. |

Source: Van de Nieuwenhof et al., 2010

Topical testosterone has been reported as effective in some cases, but they are less efficient than *Dermovate* and no more efficient than emollients. One possible explanation for the conflicting data is the alteration in androgen receptors’ expression during disease progression. Topical androgens are less commonly used due to their high cost and possible clitoral hypertrophy/virilization. Debate also exists regarding topical progesterone (Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987). Topical *Calcineurin* inhibitors are promising, but there are few well-structured randomized controlled trials to use it as trustable evidence. Topical retinoids are not effective in uncomplicated cases; moreover, they are irritants and best reserved for complicated cases unresponsive to super-potent steroids. Topical Cyclosporin was reported as ineffective in one study. Regarding systemic therapeutics, Bousema et al. (1994) reported the effective use of Acitretin (oral retinoid) at 20–30 mg/day in severe vulvar LS (administered by a specialist, retinoids are to be used with caution at childbearing age due to teratogenic potential). Hydroxychloroquine can be useful at 125–150 mg/day up to a few months, but ocular S/E is dangerous (Bousema et al., 1994; Hagedorn et al., 2002; Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987). Surgical intervention (vulvectomy and Fenton’s median perineotomy) are reserved for complicated LS cases. However, LS with dysplastic foci are conservatively treated with regular follow-up. Even with radical surgery, 80% of LS recurs around the surgical site (Carli et al., 1992; Goodfield et al., 2010; Ridley, 1987; Thomas, 2009).

* 1. **Rx of sexual problems**

Collaboration with a psychiatrist and sexologist is needed. In selected cases, emollients or estrogen creams are used before/after intercourse; this safe/cheap method will reduce the topical steroid-dosage requirement. Dysesthesia will not respond to topical steroids, and if there is no response to 5% lignocaine ointment, gabapentin/amitriptyline is justified. Surgical correction of the distorted vulva may help (Ridley, 1987; Warrington & de San Lazaro, 1996).

* 1. **Rx in pre-pubertal girls**

*Betamethasone dipropionate* is successful in vulvar LS without the need for maintenance therapy, and subsequent studies have reported the effectiveness of *Dermovate* at 2/day for 6–8 weeks. Topical estrogen was beneficial in one study (Fischer & Rogers, 1997; Garzon & Paller, 1999; Gurumurthy et al., 2012; Ridley, 1987).

* 1. **New & unlicensed Rx**

Topical *Calcineurin* inhibitors (pimecrolimus and tacrolimus) are promising as steroid-sparing alternatives, but unlicensed, and are best used in unresponsive cases to potent topical steroids. However, these immunosuppressants’ long-term safety (neoplastic transformation induction and *HPV* activation) is unknown, and they are better used in short courses. For response achieved within 16–24 weeks, one protocol showed the efficacy of 0.1% tacrolimus at 2/day for 16 weeks. Contraindications are pregnancy and breastfeeding (Goodfield et al., 2010; Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987; Thomas, 2009).

Penneys et al. reported good LS improvement at various sites after using *Potassium para-aminobenzoate* at 4–24 gm/day, and others reported the benefits of anabolic steroids, anti-histamines and anti-pruritics (Ridley, 1987; Shelley et al., 2006). The role of *Borrelia* has yet to be confirmed; however, Shelley et al. (2006) recommend intramuscular ceftriaxone every 3 weeks or intramuscular penicillin every 2–3 weeks in addition to oral penicillin/cephalosporin, which may be a sufficient treatment in some cases. Similarly, the use of prophylactic *HPV* vaccine may play a role (August & Milward, 1980; Prowse et al., 2008). Cryotherapy can be effective in vulvar LS that is unresponsive to topical steroids. CO2 laser/photodynamic therapy/PUVA were reported to be effective. High-intensity focused ultrasound (HIFU) may be safe and effective (August & Milward, 1980; Goodfield et al., 2010; Hillemans et al., 1999; Kartamaa & Reitamo, 1997; Prowse et al., 2008; Ridley, 1987; Ruan et al., 2010; Thomas, 2009).

1. **Follow-up** (Goodfield et al., 2010; Kirtschig et al., 2011; Neill et al., 2010; Ridley, 1987; Thomas, 2009)

Patients' follow-up is essential, and long-term follow-up in a specialized clinic is reserved for complicated diseases (Table 6).

Table 6

*General Guidelines for Follow-Up of LS*

|  |
| --- |
| After 2–3 months for assessing the response to treatment, if the patient’s response is good, another assessment is scheduled after 6 months. |
| Active disease to be assessed when needed. |
| Annual review for stable cases is recommended. |
| If patient reports a suspicious lump/change, an urgent medical consultation is to be made. |

Source: Goodfield et al., 2010; Ridley, 1987; Thomas, 2009

**11. Conclusion**

LS is a chronic relapsing skin disease with very high female predisposition and tropism to the ano-genital area. Its pathogenesis is still unclear. Malignant transformation can occur, but is rare. Its histology is diagnostic. Patients can experience psycho-social and sexual dysfunctions. The mainstay treatment is with potent topical steroids. However, topical *Calcineurin* inhibitors are promising alternatives.

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