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REVIEW – CONTINUING EDUCATION (CPD)

Lichen sclerosus

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Summary: Lichen sclerosus (LS) is a chronic inflammatory disorder of the skin and mucosa, presenting to genitourinary physicians and dermatologists. It affects both sexes and all age groups. Although the exact aetiology is uncertain, genetic predisposition, infections and autoimmune factors have been implicated in its pathogenesis. Symptoms include pruritis and soreness, but asymptomatic presentations are not uncommon. The classical clinical picture is of atrophic white plaques in the anogenital region. Histopathology is specific with basal cell degeneration, upper dermal oedema, homogenization of collagen and a chronic inflammatory infiltrate. Short courses of potent topical corticosteroids form the mainstay of treatment. The condition tends to be remitting and relapsing, with spontaneous regressions reported in a few. In men, the term balanitis xerotica obliterans is sometimes used to describe late and severe LS of the penis. Scarring and progression to squamous cell carcinomas can occur in chronic LS, resulting in significant morbidity. A multidisciplinary approach to care and the need for long-term monitoring cannot be overemphasized.

Keywords: lichen sclerosus, balanitis xerotica obliterans, squamous cell carcinoma

Lichen sclerosus (LS) is a chronic inflammatory disorder of unknown aetiology affecting predominantly the anogenital skin and mucosa in both sexes. It was first reported by Hallopeau in 1887.[1] Although it has been described with the synonyms circumscribed scleroderma, leukoplakic vulvitis, kraurosis vulvae, lichen albus and LS et atrophicus, the International Society for the Study of Vulvovaginal disease favours the term LS.[2]

Epidemiology

LS occurs in all age groups, with reported cases from six months of age to late adulthood.[3] There appears to be a bimodal peak in incidence from prepubertal children to the post-menopausal group in women and from 30–50 years of age in men.[4] It is uncommon under the age of two years and the mean age of onset is the fifth to sixth decade in women.[5] Most patients described in the literature are Caucasian, although this could be due to a reporting bias in epidemiological studies. It also occurs in native Africans,[6] Orientals[7] and other dark-skinned patients.[8] LS predominantly affects women and the female:male ratio varies from nearly 10:1 to 5:1.

The exact prevalence of LS is difficult to estimate. Patients with LS can present to various specialists including dermatologists, genitourinary physicians, gynaecologists and urologists. It can be asymptomatic and therefore some patients are unaware that they have the condition, while others may be embarrassed or frightened and hence not seek any medical intervention. Studies based on epidemiological surveys are therefore likely to underestimate its prevalence, which is reported to vary from one in 300 to one in 1000 of all dermatology referrals.[5]

Aetiology

Even though the exact aetiology is unknown, a variety of theories have been proposed to explain the mechanism of disease.

(1) *Autoimmune theory*: An increased incidence of autoimmune conditions has been found in association with LS, with the frequency ranging from 21 to 74%; associated disorders include thyroid disease, pernicious anaemia, diabetes mellitus, alopecia areata, vitiligo and mucous membrane pemphigoid.[1,9,10] Recently,

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it was proposed that autoantibodies against extracellular matrix protein 1 (ECM1) could be pathogenic. Immunoreactivity to ECM1 was seen in 74% of LS patients, while it was positive in only 7% of controls. ECM1 could therefore be a plausible target antigen for autoimmunity in LS. As it has influences in keratinocyte differentiation, basement membrane sclerosus, and also plays a role in angiogenesis, antibodies against ECM1 can explain the histological changes in LS of epidermal atrophy, liquefactive degeneration of the basal layer and dermal stromal changes, respectively.[11] The findings in this study provide evidence for a specific humoral immune response to ECM1 in LS.

(2) *Genetic factors*: The occurrence of LS in families has been reported.[12,13] Immunogenetic studies have identified an association with HLA class 2 antigen DQ7 (and to a lesser extent, with DQ8 and DQ9) compared with controls.[14] There have also been studies on interleukin-1 receptor antagonist gene polymorphisms related to the severity of LS.[15] This could be a candidate gene or severity factor for LS.

(3) *Infection*: It has been postulated that LS may be a consequence of atypical mycobacterial[16] or spirochaetal (*Borrellia burgdorferi*)[17] infections. Even though spirochaetes have been found in histological specimens of LS, a study using polymerase chain reaction has disproved this link.[18] Human papillomavirus has also been implicated as a possible causative agent.[19]

(4) *Hormonal influences*: The occurrence of LS in women, especially when endogenous oestrogens are low, led to a hormonal hypothesis in its aetiology. However, there is no association of LS with pregnancy, hysterectomy, use of contraceptives or hormone replacement, and no improvement occurs with the use of systemic or topical oestrogen treatment.[20] Defective androgen metabolism has also been postulated to play a part,[21] but treatment with topical testosterone confers only limited benefit.[22]

(5) *Local factors*: LS can Koebnerize at sites of trauma, sunburns, thermal burns[23] and radiation treatment. It also occurs post-vulvectomy and after circumcision.[24] Constant friction and rubbing could trigger LS. Healthy skin grafted on to the vulva becomes affected with LS and a full-thickness graft from diseased vulva transplanted onto the thigh becomes normal, suggesting local factors in the aetiology.[25]

Clinical features

Even though LS predominates in the anogenital area, extragenital LS occurs in up to 15–20% of patients.[1] In women, symptoms of pruritus, dyspareunia, dysuria and vaginal discharge are common. Incontinence can be a presenting feature.[26] Traumatic fissures and tears may occur with sexual intercourse. A significant number of patients are, however, asymptomatic. Children present with soreness in the vulva, labial stenosis or fusion and, sometimes, constipation.[27] Men can experience tightening of the foreskin, painful erections, decreased sensation in the glans, poor urinary stream and urinary obstruction.[28] Extragenital LS is usually asymptomatic although pruritus can occur.

LS begins as white polygonal papules that coalesce to form plaques (Figure 1). The labia majora, labia minora, clitoris, perineum and perianal skin may all be affected, usually symmetrically. It typically forms a 'figure of 8' configuration of lesions in the anogenital region. Oedema of the clitoral foreskin may be an early sign.[29] Fully developed lesions assume a parchment-like wrinkled appearance. With the occurrence of fibrosis and loss of elasticity, fissures may appear (Figure 2). The surface of the lesions can show prominent dilated pilosebaceous or sweat-duct orifices with horn plugs (comedo-like) or evenly spaced dells. If these are marked, the surface of the lesion may be warty.[30] Areas of telangiectasia, purpura, erosions and haemorrhagic blisters can also occur, not only in the skin but also in the genital mucosa (Figure 3). Scarring may obliterate the normal skin texture and disrupt the architecture. In extreme cases, the labia minora may fuse or be resorbed (Figure 4), and the introitus becomes so narrow that sexual intercourse becomes impossible. The vaginal and cervical tissues are not affected.[5] LS follows a relapsing and remitting course and there is poor correlation between the clinical signs and duration and severity of the disease.

Clinical and histological changes of LS are similar in children compared with adults. In all, 10–15% of LS cases occur in childhood, the majority of which involve the female genitalia.[31] The affected skin is sharply delineated and atrophic, which gives a parchment-type appearance in the anogenital area. Localized haemorrhage may be a presenting feature and can raise the suspicion of childhood sexual abuse.[31] Clinically, the appearance of hymenal trauma can help to determine if sexual abuse has occurred, regardless of the presence of LS.[32] Labial fusion can occur in childhood LS.[33] The condition tends to improve during menarche in some cases.

In men, the glans penis and foreskin are most commonly affected (Figure 5), with sparing of the perianal region. Pallor, atrophy and sclerotic plaques occur with a non-retractile prepuce (Figure 6) and erosions are not uncommon (Figure 7). Haemorrhagic bullae, follicular plugging and telangiectasia are inconsistent findings.[28] Even though LS has been considered as synonymous with balanitis xerotica obliterans (BXO), some classify it as an end-stage condition resulting from

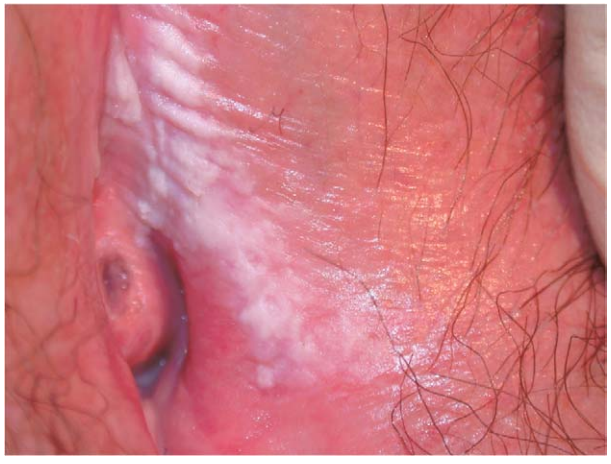

Figure 1 Porcelain white atrophic plaque of lichen sclerosus

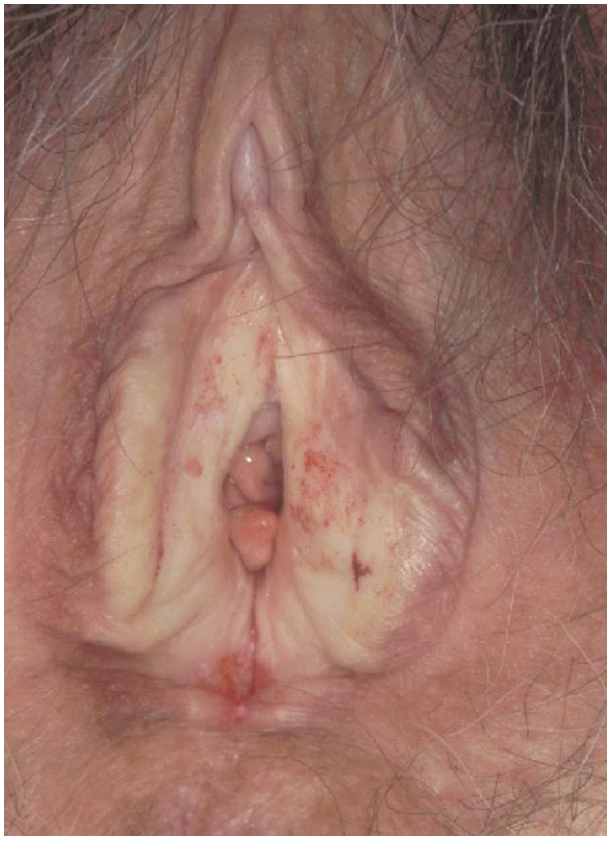

Figure 2 Chronic lichen sclerosus with erosions, purpura and fissures. Dual pathology should be considered, as the fissures here grew herpes simplex type 1

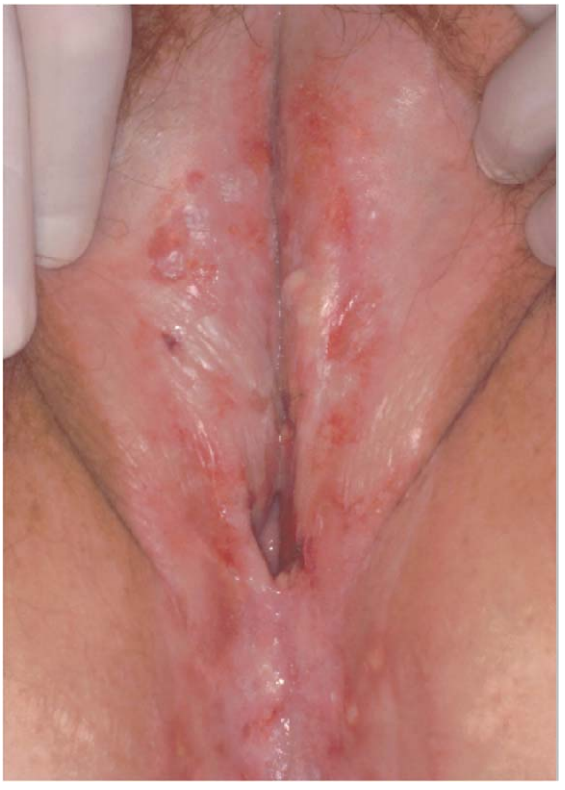

Figure 3 Extensive lichen sclerosus with erosions and excoriations

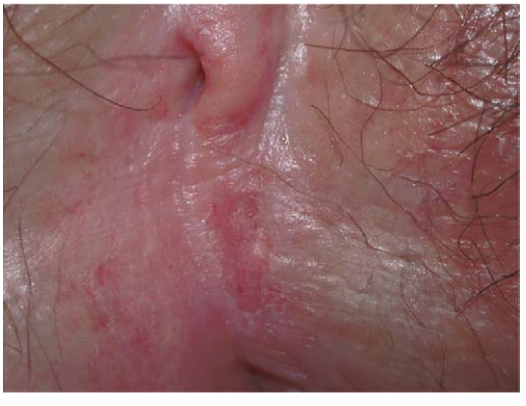

Figure 4 Resorption of labia minora almost sealing the clitoris

Figure 5 Extensive plaques around prepuce and frenulum

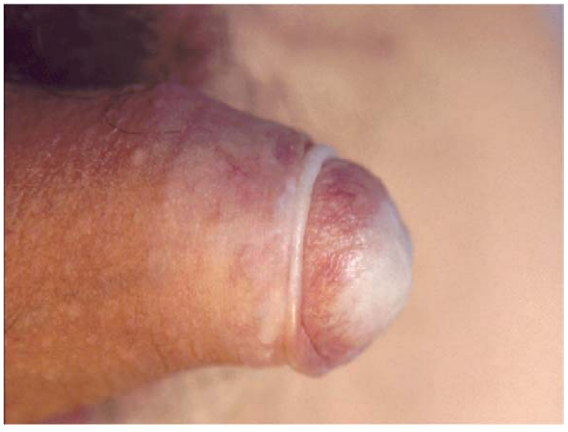

Figure 6 Fibrosis of the tip of glans penis and prepuce with difficulty of prepucial retraction

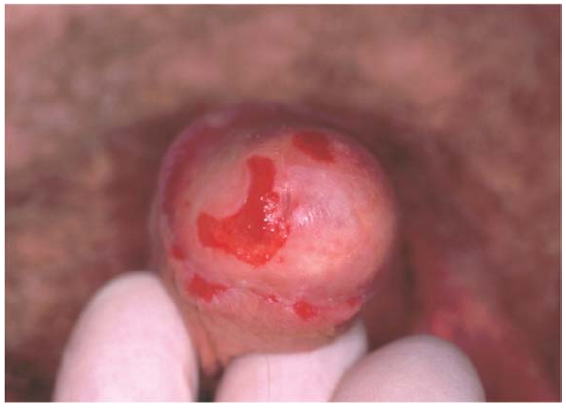

Figure 7 Extensive erosions of glans with distortion of meatal opening

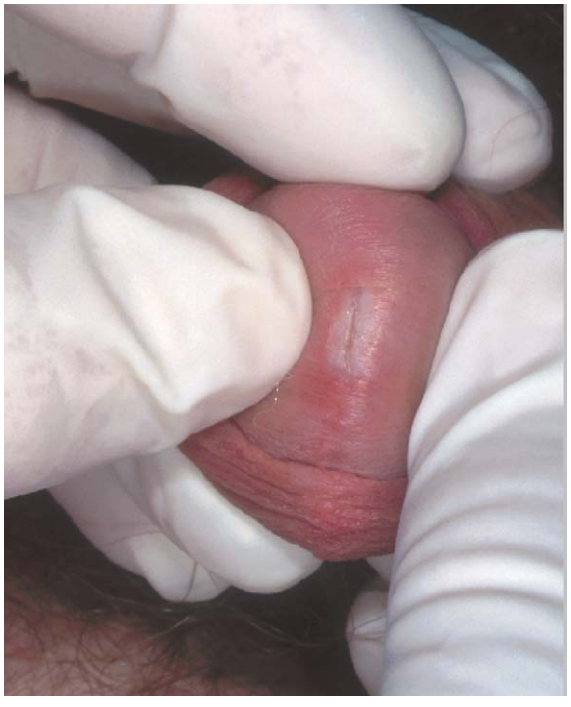

Figure 8 Balanitis xerotica obliterans with obliteration of meatal opening

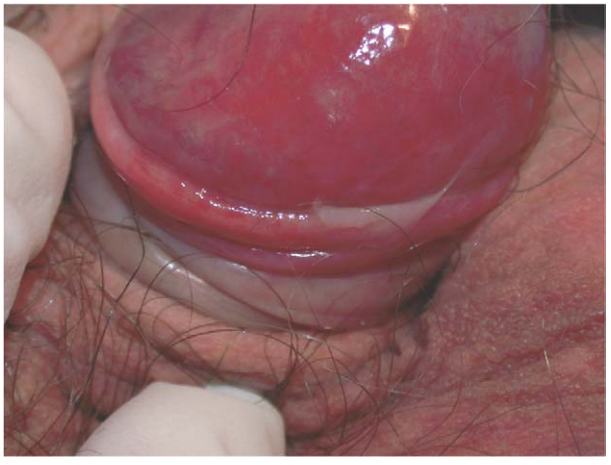

Figure 9 Early lichen sclerosus with soft adhesions of prepuce to glans

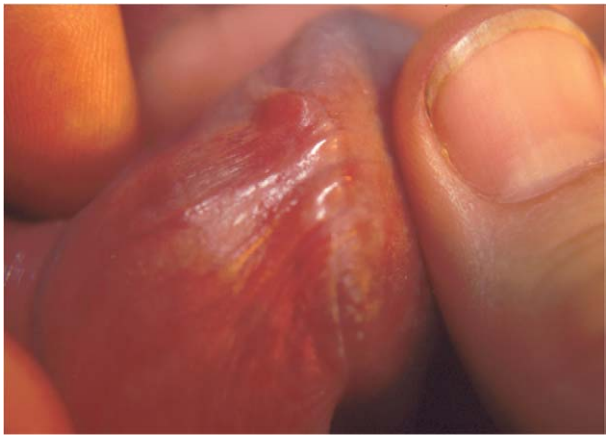

Figure 10 Well-established adhesions of prepuce to glans after untreated lichen sclerosus for six months

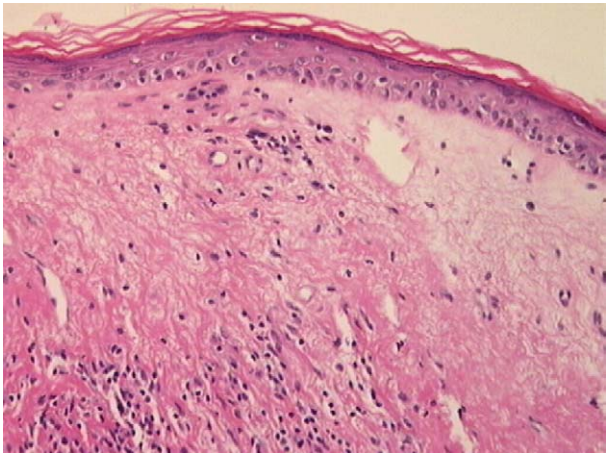

Figure 11 Histopathology showing basal cell degeneration, upper dermal oedema and lymphocytic infiltrate (H&E, $\times$ 200)

a larger spectrum of diseases, the commonest being LS (Figure 8).[28] Acute urinary obstruction due to atrophy and scarring in the glans penis can be a presenting feature.[34] LS in the anterior urethra has also been reported.[35] Adhesions of the prepuce to the glans has also been observed (Figures 9, 10). In young boys, the condition is usually asymptomatic. In one series, 14 of 100 boys referred for medical circumcision for prepucial scarring and meatal stenosis had LS.[36] Lesions in the glans are rare. Penile purpura can be a presenting feature.[37]

Extra-genital LS appears as small, ivory or porcelain-white, shiny, round macules or papules with prominent dilated pilosebaceous or sweat duct orifices. It usually forms a solitary plaque but can be annular,[38] follows the lines of Blaschko,[39] occurs in a linear[40] or band-like arrangement.[41] The most frequent sites of involvement are the upper portion of the trunk, neck and shoulders. Other uncommon sites include the scalp (resulting in scarring alopecia),[42] palms and soles,[43] areola[44] and the eyelids.[45] Extra-genital LS can Koebnerize at vaccination sites,[46] in surgical scars[47] and peristomally.[48,49] Infrequently, bullous, haemorrhagic,[50] keratotic or nodular[51] lesions may be the presenting feature. Oral LS is rare and manifests as whitish, flat lesions on the buccal, labial and palatal mucosa.[52] Localized loss of periodontal attachment can occur.[53] Nail involvement in LS has also been reported.[54]

Histology

Established lesions show hyperkeratosis, follicular plugging, thinning of the epidermis and vacuolar degeneration of the basal layer. There is a zone of subepidermal oedema with homogenous collagen (Figure 11), which becomes eosinophilic and more sclerotic in older lesions.[55] A band of mononuclear cells is seen just beneath the homogenous zone and is most evident in the mid-dermis, although it is sparse in established lesions. The infiltrate in the sclerotic region contain CD4 and CD8 lymphocytes in equal proportions, macrophages and mast cells. Squamous hyperplasia can occur in up to a third of biopsies and this could be an indicator for increased risk of malignant transformation.[56] Elastic fibres are pushed downwards by the oedematous zone and subsequently destroyed.[57] Electron microscopy (EM) shows degeneration of superficial dermal collagen and the presence of collagen in the intercellular spaces in the epidermis.[58] Other EM changes that have been reported include abnormalities in the basement membrane zone and condensation of tonofilaments in the basal epidermal cells.[59]

Associations

A variety of dermatological and systemic disorders have been associated with LS. Systemic conditions associated with LS include diabetes mellitus,[60] thyroid diseases,[61] systemic lupus erythematosus,[62] systemic sclerosus and CREST syndrome,[63] primary biliary cirrhosis,[64] polymyalgia rheumatica,[65] prolactinoma,[66] pernicious anaemia[67] and chronic hepatitis C infection.[68] Dermatological disorders that have been reported with LS include morphea,[69] vitiligo,[70] alopecia areata,[65] atrophoderma of Pasini and Pierini,[71] lichen planus[72] and erythermalgia.[73]

Differential diagnosis

The differential diagnosis for vulval LS includes cicatricial pemphigoid[74] and lichen planus.[75] However, they are distinct conditions that can be differentiated by their clinical features on cutaneous surfaces. Cicatricial pemphigoid presenting only on the vulva could lead to diagnostic confusion. Similarly, lichen planus shares clinical and pathological features with LS. Recently, authors have suggested that LS and lichen planus form a spectrum of a single disease.[76] In men, lichen planus or Zoon's plasma cell balanitis can be mistaken for LS. Extra-genital LS needs to be distinguished from discoid lupus erythematosus,[77] morphea and atrophic lichen planus.[72]

Complications

LS can be frequently superinfected with candida. However, the main complication is physical scarring, which can be associated with significant morbidity. The introitus can be narrowed, making intercourse impossible. Adhesions of the clitoral hood sometimes result in the formation of pseudocysts. Psychosexual problems like dyspareunia, difficulty in achieving orgasms and vulvodynia can develop. Loss of interest in sexual activity is common, associated with feeling of guilt and anxiety. In one patient, the occurrence of LS necessitated a caesarian section for delivery of the child.[78] In men, phimosis can occur, sometimes requiring circumcision. Meatal stenosis occasionally manifests as an altered urinary stream.

Squamous cell carcinomas (SCC) can occur in the vulva in 4–5% of patients with LS.[5] In excised samples of vulval SCC, evidence of LS can be found in adjacent skin between 25% and 61% of cases.[79,80] It is not clear what the risk for SCC is in treated patients with LS. It has been recommended that long-term follow-up is essential for most LS patients.[81] In men, only anecdotal reports of SCC occurring in LS exist and the exact risk is unknown.[82] However, in one series, 11 of 20 cases of penile SCC had a clinical history and/or histological evidence of LS.[83] Aberrant p53 expression occurs in vulval skin (as in premalignant and dysplastic skin disorders) but not in extragenital lesions.[84] Even though the pathogenesis of SCC in LS is not certain, it could be attributed to oncogenic

human papilloma viruses.[85] As topical corticosteroids are potential triggers for latent human papilloma viruses, it is not known if the risk of malignancy is changed with local control of the disease.[86] Other cancers reported with LS include verrucous carcinoma,[87] basal cell carcinomas[88] and melanomas.[89]

Rarely, extra-genital LS can give rise to local complications including ocular tendon involvement (resulting in diploplia) and gingival erosion (causing maxillary bone involvement).[90]

Treatment

A multidisciplinary team would be ideal in managing patients with LS, preferably in a dedicated clinic. Treatment should aim to control any symptoms, prevent progression, manage complications and offer surveillance for malignancy. It should be emphasized that there is no cure, although spontaneous remission can occur. Information sheets and support groups provide patients with the help required to understand the condition (www.lichensclerosus.org). Ideally, in adult women, a punch biopsy under local anaesthesia is needed to confirm the diagnosis. The British Association of Dermatologists has issued guidelines on the management of LS.[91]

Ultrapotent topical steroids (clobetasol propionate) remains the accepted and recommended treatment.[92] It is used once a night for four weeks, then alternate nights for four weeks and the third month, twice weekly. Fissuring and erosions subside with this regimen, but the atrophy and colour change remain. Even asymptomatic patients with clinically active LS (ecchymosis, hyperkeratosis, progressive atrophy) should be treated. Topical emollients could be added as soap substitutes.[93] Recently, topical tacrolimus has been found to be effective in anogenital[94] and vulval LS.[95] It is generally well tolerated and, unlike topical steroids, application is not associated with skin atrophy. Topical oestrogen[96] and progesterone[97] were suggested as possible treatments and, even though anecdotally successful, their effects have not been proved in comparative trials. Topical androgens (2% testosterone) can be used but result in androgenic side effects like hirsutism, clitoral enlargement and amenorrhea.[98] Topical tretinoin (0.025%) has been claimed to be effective in one open study on 22 patients with LS.[99] Even though there was slight histological improvement of epidermal atrophy with topical cyclosporin, there was no clinical benefit.[100] Intralesional triamcinolone can also be used in LS.[101]

Among the oral agents, acitretin (20–30 mg/day) has been effective in severe vulval LS.[102] It has also been used successfully in extragenital LS.[103] There have been case reports of stanazolol,[104] hydroxychloroquine,[105] potassium para-aminobenzoate[106] and calcitriol[107] controlling vulvar and extragenital LS. If associated with significant pain, LS can be treated with amitryptiline. Stool softening agents are helpful to prevent constipation that may occur secondary to anal fissures.

Surgical treatment for LS is reserved for patients with post-inflammatory sequelae or malignancy. Vulvectomy,[108] cryosurgery,[109] flash lamp-pumped pulsed dye laser[110] and carbon dioxide lasers[111] have been advocated as treatment options. Photodynamic therapy was effective in 10 of 12 patients in one series.[112] Perineoplasty can be considered in patients with introital stenosis, as it can improve dyspareunia associated with LS.[113] The recurrence of labial and clitoral hood adhesions in adolescents with LS can be prevented by surgical lysis and application of Surgicel® (oxidized regenerated cellulose gauze) to the affected area.[114] Tangential partial thickness excision can be considered in extragenital LS and has been successfully used in bullous LS.[115] In men, circumcision is helpful in LS of the foreskin and meatal dilation, meatotomy or meatoplasty for meatal stenosis.[116,117] Extra-genital LS can be responsive to low-dose ultraviolet (UV) A1 phototherapy.[118]

Treatment failure occurs due to non-compliance (from steroid phobia), secondary candidiasis or development of malignancy, due to sensory problems like dysaesthetic vulvodynia or as a consequence of scarring.[91] The psychosocial aspect of LS also needs to be addressed and counselling from specially trained staff may be needed in patients unable to have an active sexual life in spite of no anatomical abnormalities. Younger patients need to be reassured that there is no long-term disturbance in sexual function.[93]

Conclusion

LS is a condition that could present to various specialities including genitourinary medicine, dermatology, gynaecology and urology. From recent studies, it seems likely that LS is an autoimmune disorder, probably, to ECM1. Clinically, it can appear as atrophic white plaques in the genital region. Extra-genital lesions, however, are not uncommon. Long-term follow-up is necessary because of its infrequent, but well-recognized association with SCC in the genitals – mainly in the vulva. Management of LS would ideally require input from many disciplines and a dedicated clinic can offer a high quality of service for these patients.

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References

1 Meffert JJ, Davis BM, Grimwood RE. Lichen sclerosus. *J Am Acad Dermatol* 1995;**32**:393–416

2 Ridley CM, Frankman O, Jones IS, *et al*. New nomenclature for vulvar disease: International Society for the study of vulvar disease. *Hum Pathol* 1989;**20**:495–6
3 Aaronson LD, Baler G, Schiff BL. Lichen sclerosus et atrophicus occurring in childhood. *Arch Dermatol* 1962;**85**:746–7
4 Tasker GL, Wojnarowska F. Lichen sclerosus. *Clin Exp Dermatol* 2003;**28**:128–33
5 Powell JJ, Wojnarowska F. Lichen sclerosus. *Lancet* 1999;**353**:1777–83
6 Dogliotti M, Bentley-Phillips CB, Schmaman AS. Lichen sclerosus et atrophicus in the Bantu. *Br J Dermatol* 1974;**91**:81–5
7 Tagami H. Eosinophilic infiltration in lichen sclerosus et atrophicus. *Arch Dermatol* 1972;**105**:606–7
8 Datta C, Dutta SK, Chaudhuri A. Histological and immunological studies in a cohort of balanitis xerotica obliterans. *J Indian Med Assoc* 1993;**91**:146–8
9 Meyrick Thomas RH, Ridley CM, McGibbon DH, Black MM. Lichen sclerosus et atrophicus and autoimmunity – a study of 350 women. *Br J Dermatol* 1988;**118**:41–6
10 Harrington CI, Dunsmore IR. An investigation into the incidence of auto-immune disorders in patients with lichen sclerosus and atrophicus. *Br J Dermatol* 1981;**104**:563–6
11 Oyama N, Chan I, Neill SM, *et al*. Autoantibodies to extracellular matrix protein 1 in lichen sclerosus. *Lancet* 2003;**362**:118–23
12 Meyrick Thomas RH, Kennedy CT. The development of lichen sclerosus et atrophicus in monozygotic twin girls. *Br J Dermatol* 1986;**114**:377–9
13 Cox NH, Mitchell JN, Morley WN. Lichen sclerosus et atrophicus in non-identical female twins. *Br J Dermatol* 1986;**115**:743
14 Marren P, Yell J, Charnock FM, Bunce M, Welsh K, Wojnarowska F. The association between lichen sclerosus and the antigens of the HLA system. *Br J Dermatol* 1995;**132**:197–203
15 Clay FE, Cork MJ, Tarlow JK, *et al*. Interleukin 1 receptor antagonist gene polymorphism association with lichen sclerosus. *Hum Genet* 1994;**94**:407–10
16 Cantwell AR. Histologic observations of pleomorphic, variably acid-fast bacteria in scleroderma, morphea, and lichen sclerosus et atrophicus. *Int J Dermatol* 1984;**23**: 45–52
17 Tuffanelli D. Do some patients with morphea and lichen sclerosus et atrophicans have a Borrelia infection? *Am J Dermatopathol* 1987;**9**:371–3
18 Dillon WI, Saed GM, Fivenson DP. Borrelia burgdorferi DNA is undetectable by polymerase chain reaction in skin lesions of morphea, scleroderma, or lichen sclerosus et atrophicus of patients from North America. *J Am Acad Dermatol* 1995;**33**:617–20
19 Drut RM, Gomez M, Drut R, Lojo MM. Human papilloma virus is present in some cases of childhood penile lichen sclerosus: an *in-situ* hybridization and SP-PCR study. *Pediatr Dermatol* 1998;**15**:85–90
20 Marren P, Cherry C, Day A. Lichen sclerosus: the patients, the hormonal influences and the disease impact. *Br J Dermatol* 1995;**133**:21 (abstract)
21 Friedrich Jr EG, Kalra PS. Serum levels of sex hormones in vulval lichen sclerosus, and the effect of topical testosterone. *N Engl J Med* 1984;**310**:488–91
22 Sideri M, Origoni M, Spinaci L, Ferrari A. Topical testosterone in the treatment of vulvar lichen sclerosus. *Int J Gynecol Obstet* 1994;**46**:53–6
23 Meffert JJ, Grimwood RE. Lichen sclerosus et atrophicus appearing in an old burn scar. *J Am Acad Dermatol* 1994;**31**:671–3
24 Todd P, Halpern S, Kirby J, Pembroke A. Lichen sclerosus and the Kobner phenomenon. *Clin Exp Dermatol* 1994;**19**:262–3
25 Whimster IW. The natural history of endogenous skin malignancy as a basis for experimental research. *Trans St Johns Hosp Dermatol Soc* 1973;**59**:195–224
26 Owen CM, Yell JA. Genital lichen sclerosus associated with incontinence. *J Obstet Gynaecol* 2002;**22**:209–10
27 Clark JA, Muller SA. Lichen sclerosus et atrophicus in children. *Arch Dermatol* 1967;**95**:476–82
28 Ledwig PA, Weigand DA. Late circumcision and lichen sclerosus et atrophicus of the penis. *J Am Acad Dermatol* 1989;**20**:211–14
29 Kamarashev JA, Vassileva SG. Dermatological diseases of the vulva. *Clin Dermatol* 1997;**15**:53–65
30 Criado PR, Lima FH, Miguel DS, Valente NY, Vasconcellos C, Sittart JA. Lichen sclerosus – a keratotic variant. *J Eur Acad Dermatol Venereol* 2002;**16**:504–5
31 Wood PL, Bevan T. Child sexual enquiries and unrecognized vulval lichen sclerosus et atrophicus. *BMJ* 1999;**319**:899–900
32 Warrington SA, de San Lazaro C. Lichen sclerosus et atrophicus and sexual abuse. *Arch Dis Childh* 1996;**75**: 512–16
33 Gibbon KL, Bewley AP, Salisbury JA. Labial fusion in children: a presenting feature of genital lichen sclerosus? *Pediatr Dermatol* 1999;**16**:388–91
34 Hawkswell J, Nathan M. Lichen sclerosus and acute urinary obstruction. *Genitourin Med* 1992;**68**:177–8
35 Barbagli G, Lazzeri M, Palminteri E, Turini D. Lichen sclerosus of the male genitalia involving anterior urethra. *Lancet* 1999;**354**:429
36 Chalmers RJ, Burton PA, Bennett RF, Goring CC, Smith PJ. Lichen sclerosus et atrophicus: a common and distinctive cause of phimosis in boys. *Arch Dermatol* 1984;**120**:1025–7
37 Barton PG, Ford MJ, Beers BB. Penile purpura as a manifestation of lichen sclerosus. *Pediatr Dermatol* 1993;**10**:129–31
38 Adams BB, Mutasim DF. Annular lichen sclerosus et atrophicus. *Cutis* 2001;**67**:249–50
39 Libow LF, Coots NV. Lichen sclerosus following the lines of Blaschko. *J Am Acad Dermatol* 1998;**38**:831–3
40 Kaur S, Thami GP, Kanwar AJ, Mohan H. Linear oro-facial lichen sclerosus. *Clin Exp Dermatol* 2002;**27**:467–70
41 Izumi T, Tajima S. Lichen sclerosus et atrophicus in a band-like arrangement. *J Dermatol* 1995;**22**:764–6
42 Foulds IS. Lichen sclerosus et atrophicus of the scalp. *Br J Dermatol* 1980;**103**:197–200
43 Petrozzi JW, Wood MG, Tisa V. Palmar-plantar lichen sclerosus et atrophicus. *Arch Dermatol* 1979;**115**:884
44 Starzycki Z. Lichen sclerosus et atrophicus confined to the areola. *Br J Dermatol* 1993;**129**:748–9
45 Laws HW, Kalz F. Lichen sclerosus et atrophicus of the eyelid. *Canad J Ophthalmol* 1968;**3**:39–42
46 Anderton RL, Abele DC. Lichen sclerosus et atrophicus in a vaccination site. *Arch Dermatol* 1976;**112**:1787
47 Pass CJ. An unusual variant of lichen sclerosus et atrophicus: delayed appearance in a surgical scar. *Cutis* 1984;**33**:405–8
48 Farrar CW, Dowling P, Mendelsohn SD. Peristomal lichen sclerosus developing posturostomy. *Clin Exp Dermatol* 2003;**28**:223–4
49 Ah-Weng A, Charles-Holmes R. Peristomal lichen sclerosus affecting colostomy sites. *Br J Dermatol* 2000;**142**: 1778
50 Di Silverio A, Serri F. Generalised bullous and haemorrhagic lichen sclerosus et atrophicus. *Br J Dermatol* 1975;**93**:215–7
51 Kossard S, Pang B. Nodular lichen sclerosus. *J Am Acad Dermatol* 1994;**31**:817–8
52 Jensen T, Worsaae N, Melgaard B. Oral lichen sclerosus et atrophicus: a case report. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 2002;**94**:702–6
53 Jimenez Y, Bagan JV, Milian MA, Gavalda C, Scully C. Lichen sclerosus et atrophicus manifesting with

localized loss of periodontal attachment. *Oral Dis* 2002; **8**:310–3
54 Ramrakha-Jones VS, Paul M, McHenry P, Burden AD. Nail dystrophy due to lichen sclerosus? *Clin Exp Dermatol* 2001;**26**:507–9
55 Weedon D, In: *Pathology of the Skin*. 1st edn. London: Churchill Livingstone publications, 1997;295–7
56 Weber P, Rabinovitz H, Garland L. Verrucous carcinoma in penile lichen sclerosus et atrophicans. *J Dermatol Surg Oncol* 1987;**13**:529–32
57 Steiglerder GK, Raab WP. Lichen sclerosus et atrophicans. *Arch Dermatol* 1961;**84**:219–26
58 Mann PR, Cowan MA. Ultrastructural changes in four cases of lichen sclerosus et atrophicans. *Br J Dermatol* 1973;**89**:223–31
59 Kint A, Geerts ML. Lichen sclerosus et atrophicus. An electron microscopic study. *J Cut Pathol* 1975;**2**:30–4
60 Garcia-Bravo B, Sanchez-Pedreno P, Rodriguez-Pichardo A, Camacho F. Lichen sclerosus et atrophicus. A study of 76 cases and their relation to diabetes. *J Am Acad Dermatol* 1988;**19**:482–5
61 Poskitt L, Wojnarowska F. Lichen sclerosus as a cutaneous manifestation of thyroid disease. *J Am Acad Dermatol* 1993;**28**:665
62 Thomas RH, Ridley CM, Black MM. Lichen sclerosus et atrophicus associated with systemic lupus erythematosus. *J Am Acad Dermatol* 1985;**13**:832–3
63 Fitzgerald EA, Connelly CS, Purcell SM, Kantor GR. Familial lichen sclerosus et atrophicus in association with CREST syndrome: a case report. *Br J Dermatol* 1996;**134**:1144–6
64 Lavery HA, Pinkerton JHM, Callender M. Association between lichen sclerosus et atrophicus and primary biliary cirrhosis. *Br J Dermatol* 1985;**112**:729–30
65 Faergemann J. Lichen sclerosus et atrophicus generalisata, alopecia areata, polymyalgia rheumatica found in the same patient. *Cutis* 1979;**23**:757–8
66 Chang SN, Lee SH, Ahn SK. Lichen sclerosus associated with prolactinoma. *Br J Dermatol* 1994;**131**:448–9
67 Cunliffe WJ, Hall R, Newell DJ, Stevenson CJ. Vitiligo, thyroid disease and autoimmuntiy. *Br J Dermatol* 1968;**80**:135–9
68 Boulinguez S, Bernard P, Lacour JP, *et al*. Bullous lichen sclerosus with chronic hepatitis C virus infection. *Br J Dermatol* 1997;**137**:474–5
69 Shono S, Imura M, Ota M, Osaku A, Shinomiya S, Toda K. Lichen sclerosus et atrophicus, morphea, and coexistence of both diseases. *Arch Dermatol* 1991;**127**:1352–6
70 Osborne GE, Francis ND, Bunker CB. Synchronous onset of penile lichen sclerosus and vitiligo. *Br J Dermatol* 2000;**143**:218–19
71 Heymann WR. Coexistent lichen sclerosus et atrophicus and atrophoderma of Pasini and Pierini. *Int J Dermatol* 1994;**33**:133–4
72 Connelly MG, Winkelmann RK. Coexistence of lichen sclerosus, morphea, and lichen planus. *J Am Acad Dermatol* 1985;**12**:844–51
73 Hammar H. Plantar lesions of lichen sclerosus et atrophicus accompanied by erythermalgia. *Acta Dermatol Venerol* 1978;**58**:91–2
74 Marren P, Walkden V, Mallon E, Wojnarowska F. Vulval cicatricial pemphigoid may mimic lichen sclerosus. *Br J Dermatol* 1996;**134**:522–4
75 Holmes S. Burden. Lichen sclerosus and lichen planus: a spectrum of disease? Report of two cases and review of literature. *Clin Exp Dermatol* 1998;**23**:129–31
76 Marren P, Millard P, Chia Y, Wojnarowska F. Mucosal lichen sclerosus/lichen planus overlap syndromes. *Br J Dermatol* 1994;**131**:118–23
77 Chorzelski TP, Jablonska S, Blaszyczyk M, Fabjanska L. Annular atrophic plaques of the face. *Arch Dermatol* 1976;**112**:1143–5
78 Dalziel KL. Effects of lichen sclerosus on sexual function and parturition. *J Reprod Med* 1995;**40**:351–4
79 Zaino RJ, Husseinzadeh N, Nahhas W, Mortel R. Epithelial alterations in proximity to invasive squamous carcinoma of the vulva. *Int J Gynecol Pathol* 1982;**1**:173–84
80 Leibowitch M, Neill S, Pelisse M, Moyal-Baracco M. The epithelial changes associated with squamous cell carcinoma of the vulva; a review of the clinical, histological and virological findings in 78 women. *Br J Obstet Gynaecol* 1990;**97**:1135–9
81 Kagie MJ, Kenter CG, Hermans J, Trimbos JB, Fleuren GJ. The relevance of various vulval epithelial changes in the early detection of squamous cell carcinoma of the vulva. *Int J Gynecol Cancer* 1997;**7**:50–7
82 Pride HB, Miller OF, Tyler WB. Penile squamous carcinoma arising from balanitis xerotica obliterans. *J Am Acad Dermatol* 1993;**29**:469–73
83 Powell J, Robson A, Cranston D, Wojnarowska F, Turner R. High incidence of lichen sclerosus in patients with squamous carcinoma of the penis. *Br J Dermatol* 2001;**145**:85–9
84 Tan SH, Derrick E, McKee PH, Hobbs C, Ridley M, Neill S. Altered p53 expression and epidermal cell proliferation is seen in vulval lichen sclerosus. *J Cut Pathol* 1994;**21**:316–23
85 Ansink AC, Krul MR, De Weger RA, *et al*. Human papilloma virus, lichen sclerosus, and squamous cell carcinoma of the vulva: detection and prognostic significance. *Gynecol Oncol* 1994;**52**:180–4
86 von Krogh G, Dahlman-Ghozlan K, Syrjanen S. Potential human papilloma virus reactivation following topical corticosteroid therapy of genital lichen sclerosus and erosive lichen planus. *J Eur Acad Dermatol Venereol* 2002;**16**:130–3
87 Brisigotti M, Moreno A, Murcia C, Matias-Guiu X, Prat J. Verrucous carcinoma of the vulva. A clinicopathologic and immunohistochemical study of 5 cases. *Int J Gynecol Pathol* 1989;**8**:1–7
88 Thomas RH, McGibbon DH, Munro DD. Basal cell carcinoma of the vulva in association with vulval lichen sclerosus et atrophicus. *J R Soc Med* 1985;**78**:16–18
89 Egan CA, Bradley RR, Logsdon VK, Summers BK, Hunter GR, Vanderhooft SL. Vulvar melanoma in childhood. *Arch Dermatol* 1997;**133**:345–8
90 Dalziel K, Reynolds AJ, Holt PJ. Lichen sclerosus et atrophicus with ocular and maxillary complications. *Br J Dermatol* 1987;**116**:735–7
91 Neill SM, Tatnall FM, Cox NH. British Association of Dermatologists. Guidelines for the management of lichen sclerosus. *Br J Dermatol* 2002;**147**:640–9
92 Dalziel K, Millard PR, Wojnarowska F. The treatment of vulvar lichen sclerosus with a very potent steroid (clobetasol propionate 0.05%) cream. *Br J Dermatol* 1991;**124**:461–4
93 Neill SM, Ridley CM. Management of anogenital lichen sclerosus. *Clin Exp Dermatol* 2001;**26**:637–43
94 Bohm M, Frieling U, Luger TA, Bonsmann G. Successful treatment of anogenital lichen sclerosus with topical tacrolimus. *Arch Dermatol* 2003;**139**:922–4
95 Kunstfeld R, Kirnbauer R, Stingl G, Karlhofer FM. Successful treatment of vulvar lichen sclerosus with topical tacrolimus. *Arch Dermatol* 2003;**139**:850–2
96 Lascano EF, Montes LF, Mazzini MA. Tissue changes in lichen sclerosus in children following application of oestrogens. *Br J Dermatol* 1964;**76**:496–7
97 Jasionowski EA, Jasionowski PA. Further observations of the effect of topical progesterone on vulvar disease. *Am J Obstet Gynecol* 1979;**134**:565–7
98 Joura EA, Zeisler H, Bancher-Todesca D, Sator MO, Schneider B, Gitsch G. Short-term effects of topical testosterone in vulvar lichen sclerosus. *Obstet Gynecol* 1997;**89**:297–9

99 Virgili A, Corazza M, Bianchi A, Mollica G, Califano A. Open study of topical 0.025% tretinoin in the treatment of vulvar lichen sclerosus. One year of therapy. *J Reprod Med* 1995;**40**:614–18

100 Carli P, Cattaneo A, Taddei G, Giannotti B. Topical cyclosporine in the treatment of vulvar lichen sclerosus: clinical, histologic, and immunohistochemical findings. *Arch Dermatol* 1992;**128**:1548–9

101 Mazdisnian F, Degregorio F, Palmieri A. Intralesional injection of triamcinolone in the treatment of lichen sclerosus. *J Reprod Med* 1999;**44**:332–4

102 Bousema MT, Romppanen U, Geiger JM, *et al*. Acitretin in the treatment of severe lichen sclerosus et atrophicus of the vulva: a double-blind, placebo-controlled study. *J Am Acad Dermatol* 1994;**30**:225–31

103 Basak PY, Basak K. Lichen sclerosus et atrophicus of the scalp: satisfactory response to acitretin. *J Eur Acad Dermatol Venereol* 2002;**16**:183–5

104 Parsad D, Saini R. Oral stanozolol in lichen sclerosus et atrophicus. *J Am Acad Dermatol* 1998;**38**:278–9

105 Wakelin SH, James MP. Extensive lichen sclerosus et atrophicus with bullae and ulceration – improvement with hydroxychloroquine. *Clin Exp Dermatol* 1994;**19**: 332–4

106 Penneys NS. Treatment of lichen sclerosus with potassium para-aminobenzoate. *J Am Acad Dermatol* 1984;**10**: 1039–42

107 Ronger S, Viallard AM, Meunier-Mure F, Chouvet B, Balme B, Thomas L. Oral calcitriol: a new therapeutic agent in cutaneous lichen sclerosus. *J Drugs Dermatol* 2003;**2**:23–8

108 Abramov Y, Elchalal U, Abramov D, Goldfarb A, Schenker JG. Surgical treatment of vulvar lichen sclerosus: a review. *Obstet Gynecol Surv* 1996;**51**:193–9

109 August PJ, Milward TM. Cryosurgery in the treatment of lichen sclerosus et atrophicus of the vulva. *Br J Dermatol* 1980;**103**:667–70

110 Rabinowitz LG. Lichen sclerosus et atrophicus treatment with the 585-nm flashlamp-pumped pulse dye laser. *Arch Dermatol* 1993;**129**:381–2

111 Kartamaa M, Reitamo S. Treatment of lichen sclerosus with carbon dioxide laser vapourisation. *Br J Dermatol* 1997;**136**:356–9

112 Hillemanns P, Untch M, Prove F, Baumgartner R, Hillemanns M, Korell M. Photodynamic therapy of vulvar lichen sclerosus with 5-aminolevulinic acid. *Obstet Gynecol* 1999;**93**:71–4

113 Rouzier R, Haddad B, Deyrolle C, Pelisse M, Moyal-Barracco M, Paniel BJ. Perineoplasty for the treatment of introital stenosis related to vulvar lichen sclerosus. *Am J Obstet Gynecol* 2002;**186**:49–52

114 Breech LL, Laufer MR. Surgicel® in the management of labial and clitoral hood adhesions in adolescents with lichen sclerosus. *J Pediatr Adolesc Gynecol* 2000;**13**: 21–2

115 Klein LE, Cohen SR, Weinstein M. Bullous lichen sclerosus et atrophicus: treatment by tangential excision. *J Am Acad Dermatol* 1984;**10**:346–50

116 Khezri AA, Dounis A, Dunn M. Balanitis xerotica obliterans. *Br J Urol* 1979;**51**:229–31

117 Campus GV, Ena P, Scuderi N. Surgical treatment of balanitis xerotica obliternas. *Plast Reconstr Surg* 1984;**73**:652–7

118 Dawe RS. Ultraviolet A1 phototherapy. *Br J Dermatol* 2003;**148**:626–37

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CPD TEST QUESTIONS

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Based on: Yesudian P D, Sugunendran H, Bates C M, O'Mahony C. Lichen sclerosus. *Int J STD AIDS* 2005; **16**: 465–474

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For each of the following statements, circle whether it is True or False:

1. The aetiological factors implicated in lichen sclerosus include:
   (a) Presence of an autoantibody against extracellular matrix protein 1 True/False
   (b) An association with HLA B27 True/False
   (c) Infection with human herpes simplex virus 2 True/False
   (d) A low level of endogenous testosterone in women True/False
   (e) Local factors like trauma and thermal burns True/False

2. Clinically, lichen sclerosus can manifest as:
   (a) Violaceous flat topped papules True/False
   (b) Oedema of the clitoral skin True/False
   (c) Resorption of the labia minora True/False
   (d) Areas of erosions and telangiectasia True/False
   (e) Stenosis of the cervical os True/False

3. Lichen sclerosus can be complicated by:
   (a) Squamous cell carcinoma of the penis in 20% of affected males True/False
   (b) Super-infection with *Staphyloccocus aureus* True/False
   (c) Meatal stenosis resulting in altered urinary stream True/False
   (d) Malignant melanoma of the affected area True/False
   (e) Psychosexual problems like vulvodynia True/False

4. Lichen sclerosus in males:
   (a) Can often spare the perianal area True/False
   (b) Sometimes manifests as haemorrhagic bullae True/False
   (c) Usually affects the foreskin True/False
   (d) Is a hereditary condition True/False
   (e) Rarely presents as penile purpura True/False

5. Differential diagnoses of lichen sclerosus includes:
   (a) Cicatricial pemphigoid in the vulva True/False
   (b) Zoon's plasma cell balanitis in males True/False
   (c) Morphea when lesions are extragenital True/False
   (d) Vulval lichen planus True/False
   (e) Lymphogranuloma venereum of the penis True/False

Regulations

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