MEDOVATE
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Lichen Sclerosus

Lichen sclerosus is a common chronic skin disorder that most often affects genital and perianal areas.

Older names for lichen sclerosus (LS) include lichen sclerosus et atrophicus, kraurosis vulvae (in women) and balanitis xerotica obliterans (in males).

Who gets lichen sclerosus?

Lichen sclerosus can start at any age, although it is most often diagnosed in women over 50.

- Lichen sclerosus is 10 times more common in women than in men.
- 15% of patients know of a family member with lichen sclerosus.
- It may follow or co-exist with another skin condition
- People with lichen sclerosus often have a personal or family history of other autoimmune conditions such as thyroid disease (about 20% of patients), pernicious anaemia, or alopecia areata.

What causes lichen sclerosus?

The cause of lichen sclerosus is not fully understood, and may include genetic, hormonal, irritant, traumatic and infectious components.

Lichen sclerosus is often classified as an autoimmune disease. Autoimmune diseases are associated with antibodies to a specific protein.

- Extracellular matrix protein-1 (ECM-1) antibodies have been detected in 60–80% of women with vulval lichen sclerosus.
- Antibodies to other unknown proteins may account for other cases, explaining differing presentations of lichen sclerosus and response to treatment.
- However, these antibodies could be epigenetic, ie, the results of disease rather than the cause of disease.

Male genital lichen sclerosus is rare in men circumcised in infancy. It has been suggested that it may be caused by chronic, intermittent damage by urine occluded under the foreskin.

As onset in women is commonly postmenopausal, a relative lack of estrogen may be important.

What are the clinical features of lichen sclerosus?

Lichen sclerosus presents as white crinkled or thickened patches of skin that have a tendency to scar.

Vulval lichen sclerosus

Lichen sclerosus primarily involves the non-hair bearing, inner areas of the vulva.

- It can be localised to one small area or extensively involve perineum, labia minora (inner lips) and clitoral hood.
- It can spread onto the surrounding skin of the labia majora and inguinal fold and, in 50% of women, to anal and perianal skin.
- Lichen sclerosus never involves vaginal mucosa.

Lichen sclerosus can be extremely itchy and/or sore

Sometimes bruises, blood blisters and ulcers appear after scratching, or from minimal friction (eg tight clothing, sitting down).

- Urine can sting and irritate.
- Sexual intercourse can be very uncomfortable and may result in painful fissuring of the posterior fourchette at the entrance to the vagina.
- It may cause discomfort or bleeding when passing bowel movements.

Lichen sclerosus causes adhesions and scarring.

- The clitoris may be buried (phimosis).
- The labia minora resorb/shrink.
- The entrance to the vagina tightens (introital stenosis).

Penile lichen sclerosus

In men, lichen sclerosus usually affects the tip of the penis (glans), which becomes white, firm and scarred.

- The urethra may narrow (meatal stenosis), resulting in a thin or crooked urinary stream.
- The foreskin may become difficult to retract (phimosis).
- Sexual function may be affected, because of painful erections or embarrassment.

Extragenital lichen sclerosus

Extragenital lichen sclerosus refers to lichen sclerosus at other sites.

- Extragenital lichen sclerosus affects 10% of women with vulval disease.
- Only 6% of men and women with extragenital lichen sclerosus do not have genital lichen sclerosus at the time of diagnosis.

One or more white dry plaques may be found on the inner thigh, buttocks, lower back, abdomen, under the breasts, neck, shoulders and armpits.

- Lichen sclerosus resembles cigarette paper, as the skin is dry, wrinkled and thin (atrophic).
- Hair follicles may appear prominent, containing dry plugs of keratin.
- Bruises, blisters and ulcers may appear without noticeable trauma.

What are the complications of lichen sclerosus?

Lichen sclerosus of anogenital sites is associated with an increased risk of vulval, penile or anal cancer (squamous cell carcinoma, SCC).

- Cancer is estimated to affect up to 5% of patients with vulval lichen sclerosus.
- Cancer is more likely if the inflammatory disease is uncontrolled.
- Invasive SCC presents as an enlarging lump, or a sore that fails to heal.

Extragenital lichen sclerosus does not appear to predispose to cancer.

How is lichen sclerosus diagnosed?

An experienced clinician can often diagnose lichen sclerosus by its appearance. Skin biopsy is frequently recommended.

- Histopathology may confirm the suspected diagnosis of lichen sclerosus.
- Another skin condition may be diagnosed or coexist with lichen sclerosus.
- A focal area may undergo biopsy to assess for cancer.

Biopsy may also be recommended at follow-up, to evaluate areas of concern or to explain poor response to treatment.

What is the treatment for lichen sclerosus?

Patients with lichen sclerosus are best to consult a doctor with a special interest in the condition for accurate diagnosis and treatment recommendations.

They are advised to become familiar with the location and appearance of their lichen sclerosus.

- Women may use a mirror when applying topical therapy.
- Photographs may help in monitoring activity and treatment.

General measures for genital lichen sclerosus

- Wash gently once or twice daily.
- Use a non-soap cleanser, if any.
- Try to avoid tight clothing, rubbing and scratching.
- Activities such as riding a bicycle or horse may aggravate symptoms.
- If incontinent, seek medical advice and treatment.
- Apply emmolients to relieve dryness and itching, and as a barrier to protect sensitive skin in genital and anal areas from contact with urine and faeces.

Topical steroid ointment

Topical steroids are the main treatment for lichen sclerosus. An ultrapotent topical steroid is often prescribed, eg clobetasol propionate 0.05%. A potent topical steroid, eg mometasone furoate 0.1% ointment, may also be used in mild disease or when symptoms are controlled.

- An ointment base is less likely than cream to sting or to cause contact dermatitis.
- A thin smear should be precisely applied to the white plaques and rubbed in gently.
- Most patients will be told to apply the steroid ointment once a day. After one to three months (depending on the severity of the disease), the ointment can be used less often.
- Topical steroid may need to be continued once or twice a week to control symptoms or to prevent lichen sclerosus recurring.
- Itch often settles within a few days but it may take weeks to months for the skin to return to normal (if at all).
- One 30-g tube of topical steroid should last 3 to 6 months or longer.

The doctor should reassess the treated area after a few weeks, as response to treatment is quite variable.

Topical steroids are safe when used appropriately. However, excessive use or application to the wrong site can result in adverse effects. In anogenital areas, these include:

- Red, thin skin
- Burning discomfort
- Periorificial dermatitis
- Candida albicans infection (eg vulvovaginal thrush/yeast)

It is most important to follow instructions carefully and to attend follow-up appointments regularly.

Other topical therapy

Other topical treatments used in patients with lichen sclerosus include:

- Intravaginal estrogen cream or pessaries in postmenopausal women. These reduce symptoms due to atrophic vulvovaginitis (dry, thin, fissured and sensitive vulval and vaginal tissues due to hormonal deficiency).
- Topical calcineurin inhibitors tacrolimus ointment and pimecrolimus cream instead of or in addition to topical steroids. They tend to cause burning discomfort (at least for the first few days). Early concern that these medications may have the potential to accelerate cancer growth in the presence of oncogenic human papilloma virus (the cause of genital warts) appears unfounded.
- Topical retinoid (eg tretinoin cream) is not well tolerated on genital skin but may be applied to other sites affected by lichen sclerosus. It reduces scaling and dryness.

Internal medications

When lichen sclerosus is severe, acute, and not responding to topical therapy, systemic treatment may rarely be prescribed. Another option includes intralesional steroids.

Surgery

Surgery is essential for high-grade squamous intraepithelial lesions or cancer.

In males, circumcision is effective in lichen sclerosus affecting prepuce and glans of the penis. It is best done early if initial topical steroids have not controlled symptoms and signs. If the urethra is stenosed or scarred, reconstructive surgery may be necessary.

In females, release of vulval and vaginal adhesions and scarring from vulval lichen sclerosus may occasionally be performed to reduce urination difficulties and allow intercourse if dilators have not proved effective. Procedures include:

- Simple perineotomy (division of adhesions)
- Fenton procedure (an incision that is repaired transversely)
- Perineoplasty (excison of involved tissue and vaginal mucosal advancement)

Unfortunately, lichen sclerosus sometimes closes up the vaginal opening again after surgery has initially appeared successful. It can be repeated.

What is the outlook for lichen sclerosus?

Lichen sclerosus is a chronic disease and usually persists for years.

- Extragenital lichen sclerosus is more likely than genital disease to clear.
- Early treatment occasionally leads to complete and long-term remission.
- Scarring is permanent.

Long-term follow-up is recommended to monitor the disease, optimise treatment and ensure early diagnosis of cancer.