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- ◉ Reducing the risk of venous thromboembolism in patients admitted to hospital (2010;340:c95)
- ◉ Depression in adults, including those with a chronic physical health problem (2009;339:b4108)
- ◉ When to suspect child maltreatment (2009;339:b2689)
- ◉ Early management of persistent non-specific low back pain (2009;338:b1805)
- ◉ Recognition and assessment of coeliac disease in children and adults (2009;338:b1684)

- during invasive or 64 slice (or more) computed tomography coronary angiography
- Reversible myocardial ischaemia is found during non-invasive functional imaging.

Overcoming barriers

This guideline will enhance the understanding of the clinical assessment and investigation of patients with suspected acute coronary syndrome. Healthcare professionals should particularly note the recommendations for the urgency of referral for hospital assessment and the timing of diagnostic testing and ensure these can be implemented. In patients with suspected stable angina, the guideline emphasises the importance of a detailed clinical history to inform an initial triage of whether pain might be cardiac in origin, and mechanisms should be in place to ensure that this is accurately recorded. In patients with non-anginal pain, further diagnostic testing is not generally recommended. In patients with symptoms of typical or atypical angina who require further diagnostic testing, the estimated likelihood of coronary artery disease will determine which tests to use. Healthcare providers should ensure appropriate and timely access to high quality diagnostic testing and interpretation and have systems in place to audit their use.

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- 1 Goodacre S, Cross E, Arnold J, Angelini K, Capewell S, Nicholl J. The health care burden of acute chest pain. *Heart* 2005;91:229-30.
- 2 National Institute for Health and Clinical Excellence. Chest pain of recent onset: assessment and diagnosis of recent onset chest pain or discomfort of suspected cardiac origin (clinical guideline 95). 2010. www.nice.org.uk/guidance/CG95.
- 3 O'Driscoll BR, Howard LS, Davison AG. BTS guideline for emergency oxygen use in adult patients. *Thorax* 2008;63(suppl 6):vi1-68.
- 4 National Institute for Health and Clinical Excellence. Unstable angina and NSTEMI: the early management of unstable angina and non-ST-segment-elevation myocardial infarction (clinical guideline 94). 2010. www.nice.org.uk/guidance/CG94.
- 5 Thygesen K, Alpert JS, White HD. Universal definition of myocardial infarction. *Eur Heart J* 2007;28:2525-38.
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EASILY MISSED?

Lichen sclerosis

Kate Dalziel,¹ Sarah Shaw²

Lichen sclerosis is a chronic inflammatory skin disease usually involving the anogenital skin. It is most common in postmenopausal women but occurs in both sexes at all ages.¹ The cause is unknown, but lichen sclerosis is strongly associated with autoimmune disorders, particularly thyroid disease, in almost 30% of patients.²

Why is it missed?

Lack of familiarity with the condition and failure to examine the genital skin properly can lead to long delays in diagnosis. Reticence and embarrassment on the part of patient and doctor may hinder the taking of a full history and examination. Common mistaken diagnoses in women are candida infection and postmenopausal vulval atrophy. Candida vulvovaginitis is usually confined to women of childbearing years⁵ and is unusual in older women unless there are additional risk factors such as diabetes. The delay to diagnosis in one case series of 327 female patients was 2.2 years in children and 5.3 years in adults.⁶

Lack of familiarity with the condition probably explains why lichen sclerosis is rarely diagnosed as a cause of severe constipation in girls.⁷ Terminology has been confusing in the past, when terms such as vulval dystrophy and balanitis xerotica obliterans have been used instead

CASE SCENARIO

An 89 year old woman presents with severe vulval itch that wakes her at night. She had previously been prescribed topical clotrimazole cream and subsequently oestrogen cream with no response.

Examination shows white plaques, areas of purpura, and excoriation of the skin around the vulva and perianal region. You suspect lichen sclerosis and refer her to the vulval clinic. You also prescribe a moderately potent topical steroid and an emollient and check her thyroid function. At the vulval clinic a diagnosis of vulval lichen sclerosis with scarring is confirmed by vulval biopsy, and very potent topical steroids are prescribed, with excellent clinical response.

HOW COMMON IS LICHEN SCLEROSUS?

No good prevalence data exist for lichen sclerosis in either sex

Lichen sclerosis is one of the most frequently seen conditions in vulval clinics.¹ It is the commonest underlying skin disease in elderly women presenting with squamous carcinoma of the vulva³

It has been found in 60% of boys requiring circumcision for acquired phimosis⁴

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This is a series of occasional articles highlighting conditions that may be commoner than many doctors realise or may be missed at first presentation. The series advisers are Anthony Harnden, university lecturer in general practice, Department of Primary Health Care, University of Oxford, and Richard Lehman, general practitioner, Banbury. If you would like to suggest a topic for this series please email us (easilymissed.bmj@bmjgroup.com).



Fig 1 | Vulval lichen sclerosus showing white plaques and scarring with loss of the labia minora

of lichen sclerosus. The introduction of internationally agreed terminology⁸ and the advent of multidisciplinary vulval clinics have improved the recognition of lichen sclerosus and the use of correct nomenclature.

Why does this matter?

Lichen sclerosus is highly symptomatic. Vulval, penile, and perianal disease can cause severe intractable itching and soreness. Scarring may follow, leading to loss of vulval anatomy (fig 1), clitoral phimosis, and vulval stenosis in women.¹ Sexual function may be severely impaired.⁹ Phimosis and anterior urethral obstruction can occur in men and boys. The development of squamous cell carcinoma on genital lichen sclerosus is a well recognised complication in both sexes and is estimated to occur in about 5% of women. Sexual abuse is sometimes suspected in children with lichen sclerosus and, although the condition does not exclude abuse, which must always be considered, this has led to mistaken diagnosis.¹⁰

How is it diagnosed?

Clinical features

Adult women

The commonest presenting symptom of lichen sclerosus is severe vulval itching, and many patients will already have self medicated with over the counter antifungal or anti-itch creams. Typical changes of lichen sclerosus include areas of white skin that may be small and numerous or confluent over larger areas affecting the labia minora, labia majora, and adjacent skin of the perineum and groin creases. Perianal disease is common, giving a “figure of eight” pattern of affected skin around the vulva and perianal region. The white skin often looks thin, wrinkled and fragile, with red or purple areas of bleeding into the skin. Excoriations are common. Sometimes there are thickened plaques or even warty areas. In a series of 253 women with lichen sclero-

sis, 177 had atrophic change and 94 had hyperkeratotic (warty) areas.⁶

In the patients with scarring (about 70% of women)⁶ symptoms can include difficulty in passing urine, especially in men, and in women, dyspareunia, anorgasmia and splitting of the skin on attempted intercourse. Women may notice the change in their vulva with loss of the labia minora (fig 1) and fusion of the labia over the clitoris. Occasionally the first presentation of lichen sclerosus will be when squamous carcinoma develops: a plaque, ulcer or nodule can arise and enlarge very quickly, sometimes in a matter of weeks.

Adult men

Itching and soreness of the glans and prepuce are common. White areas and purpura can be seen on the glans and foreskin but may be hidden by a tight phimosis (fig 2). Squamous cell carcinoma can arise in the affected area, with features similar to those described above for women.

Children

Prepubertal girls with lichen sclerosus often present with vulval and perianal itching and show the typical skin changes as described for adult women, but they may also present with intractable constipation, soiling, anal fissures, and bleeding. Boys tend to present with phimosis, and this may hide the typical skin changes.

Differential diagnosis

Other chronic itchy skin diseases need to be considered. Various types of eczema that affect the perineal and perianal areas, such as contact dermatitis and seborrhoeic eczema, are common. Another auto-immune skin condition, lichen planus, can also show a predilection for genital skin. It too may cause scarring and squamous carcinoma. Plaques of vulval intraepithelial neoplasia may be white, but the other typical features of lichen sclerosus will be absent.



Fig 2 | Lichen sclerosus of glans penis in adult showing whiteness and purpura

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“There is another fact that may be easily missed when visiting a patient affected by LS, ie, the possibility that LS may involve also extragenital sites. Extragenital involvement may in fact be even more easily missed since these lesions are generally not symptomatic, unlike the genital ones.”

Samuele Naviglio in a rapid response. To read all rapid responses or to send a response, visit bmj.com

KEY POINTS

Always examine postmenopausal women with vulval itching for lichen sclerosus and other itchy skin disorders

Consider lichen sclerosus in girls with severe constipation and in boys with phimosis

Lichen sclerosus is often diagnosed by the clinical appearance, but if uncertainty exists then it is essential that a biopsy is done before very potent topical steroids are used

Check thyroid function, and remember that a patient with lichen sclerosus is also more likely to have other autoimmune disorders

Consider using topical steroids to treat phimosis caused by lichen sclerosus as this may avoid the need for circumcision

Advise patients about the small (5%) risk of malignant change and to report immediately any change, such as an ulcer or lump

Investigations

No diagnostic biochemical or immunological investigations exist, but a skin biopsy will often provide the diagnosis. The histology of lichen sclerosus is distinctive, and biopsy will help to differentiate it from other skin diseases that cause anogenital itching and scarring. However, use of a very potent topical steroid (such as clobetasol propionate) may completely correct the clinical and histological changes, so such a steroid must not be used before a definitive diagnosis is made. Accurate diagnosis is important because patients with lichen sclerosus will need follow-up and education about ongoing treatment and the risk of cancer development.

Check thyroid function in all adult patients with lichen sclerosus, given the association with autoimmune thyroid disease.

How is it managed?

Evidence based guidelines for the management of lichen sclerosus state that if clinical doubt exists then a biopsy should be performed to confirm the diagnosis.¹¹ The general practitioner may wish to refer the patient to a specialist with an interest in vulval disease or to an established vulval clinic (based in dermatology, genitourinary medicine, or gynaecology departments). Pending the definitive diagnosis, emollients can be used as soap substitutes and to help repair the skin barrier, and a moderate potency topical steroid such as clobetasol butyrate ointment can be used sparingly. After diagnosis, the accepted treatment is with the very potent topical steroid clobetasol propionate ointment.¹¹ No controlled trials have been published to establish the best regime, but the guidelines suggest a single application at night for four

weeks followed by alternate nights for four weeks and then twice a week for four weeks. Once symptoms are well controlled, the steroid ointment is used as required. Some patients will achieve complete remission; others will require ongoing treatment. In women, surgery is reserved for correcting the effects of scarring and for removal of cancer. Surgery is not otherwise appropriate for benign disease as it is mutilating and there is rapid relapse of the lichen sclerosus.

In men, circumcision may sometimes be needed for phimosis. However, treatment with a very potent topical steroid may avoid the need for circumcision in both men¹² and boys.¹³ In children, if the diagnosis is not certain then referral to a specialist is indicated rather than attempting a biopsy.

Useful support and advice for patients can be found on the National Lichen Sclerosus Support Group's website www.lichensclerosus.org.

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- ▶ Constipation in people with learning disability (2010;340:c222)
- ▶ Long QT syndrome (2010;340:b4815)
- ▶ Ovarian cancer (2009;340:b4650)
- ▶ Slipped capital femoral epiphysis (2009;339:b4457)

Always take a careful history

Unusually for Lyme Regis, on England's south coast, there had been a severe episode of heavy snow that lasted for several days and I was snowbound at home. I lived a mile or so from the town centre, and my practice, at the top of a steep hill. Everything was at a standstill, and I could not get my car on the road.

On the third day I was surprised to see that the milkman had arrived. He had managed to obtain chains to fix to the wheels of his van. He told me of the difficulties he had encountered on his round and mentioned that "poor old Mrs Jones has bad water trouble." She was one of my patients, so I donned suitable clothing and boots, grabbed my medical case, and trudged and slithered the mile down hill in a blizzard to poor old Mrs Jones's house.

On my arrival, Mrs Jones welcomed me with great surprise: "Fancy you turning out on a day like this, doctor. Do come in from the cold." I explained that the milkman had advised me of her water trouble. She nodded and pointed up to the ceiling, where a large patch of damp was to be seen. However, the plumber was on his way, so I commiserated, and we had a cup of tea and a chat. I then trudged back up the hill, treating some children on the way by joining in with a snowball fight.

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