Lichen Sclerosus

Lichen Sclerosus is a skin condition wherein patches of smooth, shiny, distinctly white skin appear that have a thin, papery, porcelain-like appearance. While there may be no additional symptoms, these patches may grow into areas that have a wrinkled, porcelain-like appearance. While severe itching, tenderness, may be severe itching, tenderness, or bruising and in severe cases, this bruising may lead to bleeding, blistering or ulceration. It is thought to be an inflammatory disorder and not mediated by any infectious agent.

Lichen Sclerosus (LS) can affect anyone, and while it is most common in post-menopausal women, young girls, boys and men are also affected, but these are much less common. In women and girls, the most common sites of LS is the vulval-anal region, while in men and boys the condition generally affects the foreskin. Here, in severe cases, loss of elasticity in the tissue can require circumcision. In both girls and boys, the condition can resolve spontaneously around puberty.

While the cause of LS is presently undefined, its etiologic suggests that estradiol may protect women of child-bearing age. There is also reasonable evidence that LS has an autoimmune component. Neither of these two observations however, indicates any particular triggering event; nonetheless, previous skin damage at an LS site has been posited as an initiating factor.

LS can occur on skin outside the anogenital area (extragenital LS) and here, it may be treated by observation. Anogenital symptoms however are usually treated to diminish itching and the consequent risk of infection. Topical glucocorticoids applied over several weeks is usually the first approach and if this is unsuccessful, more powerful immunomodulators may be used.

Common Features and Presentation

LS is defined as a chronic, inflammatory dermatosis, resulting in white plaques with epidermal atrophy. It is relatively common, although the distribution of patches between dermatologists, gynecologists, urologists and other clinical disciplines makes it hard to determine the true frequency. In female patients, genital LS has two peaks of presentation, prepubertal and post-menopausal. A similar bimodal onset is observed in males.

As previously stated, LS presents genitally and extragenitally. While LS is the common term for all aspects of the disease, it may be described as ‘lichen sclerosus et atrophicus’ in a dermatological context, or ‘Balinitis xerotica obliterans’ if the head of the penis is involved. There are few data that suggest an association with malignant disease and precancerous (such as lichenoid or HPV) lesions are undefined. General presentation (vulval and perianal) can be similar to extragenital, extrapenile LS, with LS regions on the male abdomen, chest and arms.

In certain cases, there is a poor response to topical glucocorticoid treatment. It may be difficult to differentiate between LS and the closely-related Lichen Planus (LP) and these ‘overlap syndromes’ patients are the ones who are at risk of squamous cell carcinoma. In certain cases where there is a poor response to topical glucocorticoid treatment, it may be difficult to differentiate between LS and the closely-related Lichen Planus (LP) and these ‘overlap syndromes’ patients are the ones who are at risk of squamous cell carcinoma.

The Adult Female Patient: This is the most common patient. Her typical LS regions will be ‘balinitis-xerotica’-like and associated with redness and bruising. She will most likely be experiencing severe itching and may have introduced tears through scratching of the thinned skin, leading to pain and possible opportunistic infection. LS regions never affect mucosal tissue so, unlike LP, LS is not found inside the vagina or on the cervix. Characteristic sites include the perineum, the major and minor labia and the clitoris and bilateral clitoral hood. Since LS is intrinsically a scarring condition, loss of elasticity and even sewing of the clitoral hood may occur. Scarring at the entrance to the vagina can make intercourse difficult and painful, as can lesions arising in the thinned skin. Perianal LS occurs in 30% of cases and again, lesions may result in difficult bowel movements and infection.

The Juvenile Female Patient: The regions affected in juveniles are very similar to those in adult females. However, bruising can be very pronounced and perianal lesions may be more common, leading to patients presenting with constipation as the primary symptom.

The Adult Male Patient: Perianal LS is rarely found in males. LS is almost completely confined to the penis, particularly the foreskin prepuce and the glans. In certain cases, where there is a poor response to topical glucocorticoid treatment, it may be difficult to differentiate between LS and the closely-related Lichen Planus (LP) and these ‘overlap syndromes’ patients are the ones who are at risk of squamous cell carcinoma. In certain cases where there is a poor response to topical glucocorticoid treatment, it may be difficult to differentiate between LS and the closely-related Lichen Planus (LP) and these ‘overlap syndromes’ patients are the ones who are at risk of squamous cell carcinoma.

Epidemiology

Epidemiology of LS is unclear, but autoimmune mechanisms are being called upon more frequently. Associations with other autoimmune diseases has been noted (in particular, Thyroid Disease), there is a recognized tissue-specific autoantibody profile and MHC class-II associations, all indicative of an autoimmune background. This is supported by the presence of circulating extracellular matrix proteins, and basement membrane zone protein 1/180 may be emerging as important at both T-cell and B-cell levels.

Relationship with Malignant Disease

It may be quite difficult to navigate the literature regarding LS and malignant disease, in order to separate descriptions of patients with both conditions, and those that address whether LS confers an additional risk. A significant literature exists regarding the role of papilloma viruses in
The treatment may be pharmacological or surgical, the choice depending
on the extension of the involved area, the histological pattern and the level
of disease activity. Our study is the first large prospective study that
has been carried out in a defined cohort of patients. The treatment
options are aimed at ameliorating the symptoms, preventing the
ulcers and reducing disease progression. The study was designed to
evaluate the pathological and histological characteristics of the
disease, with a view to developing a more targeted and effective
therapy in the future.

Topical Immunomodulators in Lichen Sclerosus

IL-20 has been shown to prevent the development of
ulcerating cutaneous lupus erythematosus. Moreover, it has been
shown to reduce the expression of IL-17 and TNF-alpha, which are
important cytokines in the pathogenesis of lichen sclerosus. In a
recent study, IL-20 was shown to significantly reduce the number
of CD8+ T cells in the basal layer of the skin, which is associated
with the reduction of disease activity.

Surgical treatment is normally not a good option for women. When
the lichen sclerosus involves the vulva, it can be removed surgically
by POEM. However, this is usually not necessary and it is
recommended to continue with topical treatments until the disease is
under control. If the treatment fails to improve, POEM may be
considered, but it should be noted that the recurrence rate is high.

Pharmacological treatment is also an option, but it is not always
effective. Topical steroids, such as hydrocortisone, are commonly
used, but they may cause skin thinning and other side effects.

In conclusion, lichen sclerosus is a chronic inflammatory disease that
affects the skin and underlying tissues. It is characterized by
white plaques, atrophy, and pruritus. The treatment options include
topical and systemic therapies, as well as surgical interventions.

Research is ongoing to develop new and more effective treatments
for this condition.
Effective November 26, 2012...

- We will require that all specimen containers be labeled at the time of specimen collection with two patient identifiers. These identifiers must correspond to information provided on the requisition form or accompanying documents.
- As of November 26, 2012, we will begin to note on result reports if a specimen is received without the required two patient identifiers.

Preferred first identifier

Preferred second identifier

Other acceptable identifiers

- Social security number
- Date of birth
- Patient Identification number

Additional desirable information

- Unique random number
- Accession number
- Medical record number

Sincerely,

Kelly D. Smith
Director of Quality Control

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Medical Diagnostic Laboratories L.L.C.

Specimen Labeling Policy...

The College of American Pathologists (CAP) guidelines state that all primary clinical specimen containers be labeled with two patient identifiers at the time of specimen collection.

GEN.40491 Specimen Labeling Phase II

Primary specimen containers are labeled by at least 2 identifiers.

NOTE: All primary specimen containers must be labeled with 2 identifiers at the time of collection. Submitted slides may be labeled with a single identifier, but two identifiers are preferred. Examples of acceptable identifiers include but are not limited to: patient name, date of birth, hospital number, social security number, accession number, unique random number. The ‘primary’ specimen container is the innermost container received by the laboratory that actually holds the specimen. It is good laboratory practice to use two identifiers.

Effective November 26, 2012...

- We will require that all specimen containers be labeled at the time of specimen collection with two patient identifiers. These identifiers must correspond to information provided on the test requisition form or accompanying documents.
- As of November 26, 2012, we will begin to note on report failures if a specimen is received without the required two patient identifiers.

Preferred first identifier

- Patient’s first & last name

Preferred second identifier

- Patient date of birth

Other acceptable identifiers

- Social security number
- Requisition number
- Patient Identification number
- Medical record number
- Accession number
- Unique random number

Additional desirable information

- Date of collection
- Time of collection
- Specimen type
- Specimen source

Sincerely,

Kelly O. Smith
Director of Quality Control

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Recent Publications

MDL: Research & Development Peer-Reviewed Papers:


Oncoveda Cancer Research Center™: Research & Development Peer-Reviewed Papers:


Question:

I received the results from testing I ordered for a patient. The tests were negative so I would like to request some additional testing. How do I do this?

Answer:

Testing can be added if the specimen volume permits and it is within an allowable time period from specimen collection. For our swab platforms OneSwab®, UroSwab®, and NasoSwab® test addition requests can typically be honored up to 30 days from receipt of the specimen. For other specimen types, it is best to contact our Client Services Department toll free at 877.269.0090. All test addition requests MUST be received in writing on either a Test Addition Request Form, office stationary, or on a script. These requests should be faxed to MDL’s QA/QC Department at 609.570.1020.

If you have a question you would like addressed in future issues, please email your question(s) to QA&A@mdlab.com

Quality Assurance Q&A

Question:

LS may be observed by practitioners in which of the following medical specialties?

A. Post-menopausal women
B. Boys and men
C. Boys and men
D. All of the above

Answer:

D. All of the above

REFERENCES:

1. True or False. Lichen Sclerosus (LS) is mediated by an infectious process.

2. LS affects the following groups:
   A. Post-menopausal women
   B. Young girls
   C. Boys and men
   D. All of the above

3. LS is believed to play a protective role against LS in women of child-bearing age.

4. True or False. Although in women there appears to be a bimodal onset as observed in males.

5. LS may be observed by practitioners in which of the following medical specialties?
   A. Dermatologists
   B. Urologists
   C. Gynecologists
   D. All of the above

For results to the electronic Epidemiology Quiz, please visit www.mdlab.com and click on the e-Quiz link.
Diagnostic usefulness of dermatoscopy in differentiating lichen sclerosis et atrophicus from morphea


Morphea and lichen sclerosus et atrophicus (LSA) are inflammatory skin disorders that may present similar clinical features. Unilateral, progressive, nodular, or plaque-like scalloped lesions, resembling morphea, may be mistaken for lichen sclerosus. Dermoscopy, which has been found useful in differentiating morphea from chronic actinic dermatitis, is also useful in distinguishing LSA from morphea.

In patients with lichen sclerosus, focal, atrophic, and erythematous plaques are present, with a typical appearance: comedo-like openings and whitish patches. These findings can be present in both lichen sclerosus and morphea. Dermoscopy can help in differentiating LSA from morphea by showing the presence of comedo-like openings in LSA, which are not present in morphea.

The use of dermoscopy can help in making a more accurate diagnosis, which may lead to more appropriate treatment and improve patient outcomes. Dermoscopy can be a valuable tool in the management of patients with LSA and morphea, especially in cases where clinical differentiation is difficult.
Lichen Sclerosus

Overview

Lichen Sclerosus is a skin condition wherein patches of smooth, shiny, distinctly white skin appear that have a thin, papery, porcelain-like appearance. While there may be no additional symptoms, these patches may grow into areas that have a wrinkled, blistery appearance; there may be severe itching, tenderness, or pain from bruising or tearing and in severe cases, this tearing may lead to bleeding, blistering or ulceration. It is thought to be an inflammatory disorder and not mediated by any infectious agent.

Lichen Sclerosus (LS) can affect anyone, and while it is most common in post-menopausal women, young girls, boys and men are also affected, but these are much less common. In women and girls, the most common site of LS is in the vulval-anal region, while in men and boys the condition generally affects the foreskin. Here, in severe cases, loss of elasticity in the tissue can require circumcision. In both girls and boys, the condition may resolve spontaneously around puberty.

While the cause of LS is generally undefined, its etiology suggests that extrudal may protect women of child-bearing age. There is also reasonable evidence that LS has an autoimmune component. Neither of these two observations however, indicates any particular triggering event; nonetheless, previous skin damage at an LS site has been postulated as a risk factor.

LS can occur skin outside the anogenital area (extragenital LS) and here, it may be treated by observation. Anogenital symptoms however are usually treated to diminish itching and the consequent risk of infection. Topical glucocorticoid applied over several weeks is usually the first approach and if this is unsuccessful, more powerful immunomodulators may be used.

Common Features and Presentation

LS is defined as a chronic, inflammatory dermatosis, resulting in white plaques with epidermal atrophy. It is relatively common, although the distinction of patches between dermatologists, gynecologists, urologists and other clinical disciplines makes it hard to determine the true frequency. In female patients, genital LS has two peaks of presentation, prepubertal and post-menopausal. A similar bimodal onset is observed in males.

As previously stated, LS presents generically and extragenitally. While LS is the common term for all aspects of the disease, it may be described as ‘lichen sclerosus et atrophicus’ in a dermatological context, or ‘Balinitis xerotica obliterans’ if the head of the penis is involved. There are few data that suggest an association with malignant disease, and the nomenclature (such as psoriasis vulgaris or PV) are undefined. General presentation (vulval and perineal) outnumber those of genital LS; blistering may be present. A similar bimodal onset is observed in males.

Extra-anogenital LS is described as ‘porcelain-white’ and associated with the children of patients with both conditions, and those that address the relationship with Malignant Disease. Emerging as important at both T-cell and B-cell levels, there is a recognized tissue-specific autoantibody profile and MHC class-II associations, all of which are undefined. Genital presentations invariably show significant leukocytic infiltration.

Confirmatory Histological Examination

This is not always necessary, unless there is reason to suspect metaplastic or necrapsid changes (for example, erythema, wart-like lesions and/or hyperkeratosis that does not resolve with glucocorticoid treatment). Histological analyses invariably show significant leukocytic infiltration.

Evidence for an Autoimmune Component

The etiology of LS is unclear, but autoimmune mechanisms are being called upon more frequently. Associations with other autoimmune diseases have been noted (in particular, Thyroid Disease), there is a recognized tissue-specific autoreactivity profile and HLA class-II associations, all indicative of an autoimmune background. This is supported by the presence of circulating extracellular matrix proteins, and basement membrane zone protein 180 may be emerging as important at both T-cell and B-cell levels.

Relationship with Malignant Disease

It may be quite difficult to navigate the literature regarding LS and malignancy, disease, in order to separate descriptions of patients with both conditions, and those that address whether LS confers an additional risk. A significant literature exists regarding the role of papilloma viruses in

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