A Case Report of Extragenital Lichen Sclerosus with Anogenital Sparing

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Abstract
Lichen sclerosus is an uncommon, chronic inflammatory condition primarily affecting the anogenital region; however, it can have concurrent or sole extragenital involvement. The etiology is controversial, and lichen sclerosus, specifically the anogenital type, has been linked to future development of squamous cell carcinoma. We present a case of extragenital lichen sclerosus, along with a review of the literature to discuss presentation, etiology, diagnosis, management, and risk of future squamous cell carcinoma.

Introduction
In most cases, lichen sclerosus is identified in the anogenital region, but solely extragenital presentations are not unheard of. When present, most extragenital lesions are visualized on the superior trunk, axillae, buttocks, and extremities, often in conjunction with anogenital lesions. This patient presents with a case of classical-appearing lichen sclerosus on the inferior, posterior aspect of the trunk with anogenital sparing.

Review of the literature revealed other reported cases of lichen sclerosus devoid of anogenital involvement, including those with bullous presentations and those following the lines of Blaschko on the face. In addition, a rare case of extragenital lichen sclerosus involving the oral mucosa has previously been documented.

Lichen sclerosus, like many dermatologic diseases, has multiple proposed etiologies, including those related to genetics, infection, and autoimmune disease. The patient presented in this case lacked any of the above predispositions and was placed on topical corticosteroids, which are the mainstay of therapy and are generally known to be effective. However, regardless of the simplicity or complexity of the patient’s medical history, choosing effective therapy can prove to be quite challenging when patients do not respond to topical corticosteroids. Recalcitrant lichen sclerosus may require additional therapies including trials of calcineurin inhibitors, methotrexate, cyclosporine, and phototherapy.

Case Presentation
An 83-year-old Caucasian female presented to the clinic complaining of an “itchy discomfort” on her lower back that had previously bled. She denied discomfort elsewhere. She reported a history of hypertension, thyroid disease, and arthritis but denied any history of skin cancer or other dermatologic issues. On physical exam, she was found to have large, hypopigmented, atrophic plaques extending over the central lower thoracic and lumbar areas of the back with a cigarette-paper-like, wrinkled texture (Figure 1). The area was devoid of blisters, erosions, and excoriations. Concomitant vulvar disease was not present.

A 4-mm punch biopsy was taken and revealed subepidermal clefting with homogenization and edema of the papillary dermis with expansion, as well as mild superficial lymphocytic and eosinophilic infiltrate in the superficial dermis (4x).

The clinical and pathological findings were consistent with lichen sclerosus et atrophicus. The patient was managed with topical triamcinolone acetonide lotion and a tapered dose of prednisone along with a ceramide moisturizer. Phototherapy and methotrexate are being considered if symptoms persist.

Discussion
Lichen sclerosus is a chronic, relapsing,
inflammatory condition affecting mucocutaneous junctions and cutaneous surfaces, leading to epidermal atrophy.\textsuperscript{3,4} This lymphocyte-driven condition has a predilection for the anogenital area; however, extragenital lichen sclerosus has been reported to occur in 15\% to 20\% of patients with lichen sclerosus.\textsuperscript{6,6} Of note, this condition is commonly referred to as lichen sclerosus without the ‘atrophicus’ portion.\textsuperscript{6} This is attributed to findings that lichen sclerosus can result in hypertrophic rather than atrophic epithelium.\textsuperscript{6}

The etiology of lichen sclerosus is unknown. Studies have determined that there is a change in the expression of certain proteins, specifically extracellular matrix protein-1 (ECM-1), and connective-tissue growth factor in patients with lichen sclerosus.\textsuperscript{7} Some studies have proposed autoimmune, endocrine, and even an infectious etiology. To elaborate, studies have located tissue-specific antibodies, and clinicians have persistently recognized its association with other autoimmune conditions such as vitiligo, thyroid disease, and pernicious anemia in genetically predisposed individuals.\textsuperscript{5,6,8} With prepubescent children and postmenopausal women being affected, many have also attributed the onset to decreased estrogen levels.\textsuperscript{5,8} Although controversial, several reports suggest the disease may be triggered by an infectious agent, specifically \textit{Borrelia burgdorferi}.\textsuperscript{4,5,9} In addition, koebnerization has been described in the development of extragenital lesions.\textsuperscript{6}

Lichen sclerosus can affect individuals across all age groups.\textsuperscript{5} However, the condition typically demonstrates a bimodal onset among prepubertal children and postmenopausal women, with the latter being the most commonly affected group.\textsuperscript{5,7} The incidence of lichen sclerosus is difficult to assess because of the large number of asymptomatic individuals and the multi-specialty patient coverage.\textsuperscript{6}

Lesion distribution is most commonly in the vulvar, perianal, and groin regions.\textsuperscript{9} Extragenital lesions are usually visualized on the upper trunk, axillae, buttocks, extremities or lateral thighs.\textsuperscript{9} The most commonly reported presenting symptoms are pruritus and irritation, especially at night, though many individuals are asymptomatic.\textsuperscript{5,6,9} Symptoms can also be location-dependent. For example, patients with vulvar lichen sclerosus often experience dyspareunia, dysuria, or bleeding secondary to irritation and introital narrowing, whereas those with perianal lichen sclerosus may experience dyschezia due to similar mechanisms.\textsuperscript{5,6,9} The predominance of chronic pruritus inevitably leads to lichenification, fissures, and future atrophy, scarring, and potential malignancy.\textsuperscript{5,8}

In lichen sclerosus, lesions evolve with time. Classically, preliminary lesions appear as small, minimally raised, pink- or ivory-colored papules.\textsuperscript{9} These early lesions are usually flat-topped with white or brown follicular plugs referred to as “delling.”\textsuperscript{9} Over time, these papules become confluent, forming white, porcelain-like plaques with a characteristic atrophic and wrinkled appearance.\textsuperscript{9} Dermoscopy of extragenital lesions often reveals scaling and keratotic plugs.\textsuperscript{4} Interestingly, an erythematous halo visible on dermoscopy is indicative of an active lesion.\textsuperscript{10}

On histology, lichen sclerosus usually reveals an edematous papillary dermis, effaced epidermis, and blured border of separation between the two layers.\textsuperscript{8,9} The damage and subsequent atrophy of the epidermis results in contraction of the skin and therefore the characteristic wrinkled appearance of the skin lesions.\textsuperscript{5} Additional histopathological features consistent with both genital and extragenital lesions include hyalinosis in the upper dermis, vascular ectasia, and basal vacuolization.\textsuperscript{5,7}

The use of biopsy to establish a diagnosis for lichen sclerosus has been controversial for some time. Many clinicians discourage biopsy, arguing that lichen sclerosus is more of a clinical diagnosis. However, should biopsy be pursued in questionable cases, it is important to be aware that biopsies of lichen sclerosus are not always straightforward and that an inconclusive biopsy does not necessarily rule out the diagnosis of lichen sclerosus.\textsuperscript{5,9} Cases where biopsy is warranted include those that have signs of neoplasia such as chronic ulceration, fissuring, or hyperplasia.\textsuperscript{5} Biopsy is also recommended in lesions unresponsive to treatment, lesions with pigmentation, and in lesions that must be differentiated from other conditions that present similarly to lichen sclerosus, including lichen planus, lichen simplex chronicus, guttate morphea, discoid lupus erythematosus, vitiligo, anetoderma, cutaneous T-cell lymphoma, chronic graft-versus-host disease and extramammary Paget's disease.\textsuperscript{4,6,9,11} In addition, it is important to consider the social history, especially in children with anogenital lesions, as sexual abuse must be ruled out.

Preliminary management of lichen sclerosus is with high-potency topical steroids.\textsuperscript{4} Clobetasol propionate ointment 0.05\% has been found to be effective for symptomatic relief across all age groups and has been shown to induce recovery of skin changes such as atrophy and even reverse the histological changes.\textsuperscript{5} It is unclear as to how topical steroids recover skin atrophy, but it can be assumed that this reversal is a product of steroid action on reducing inflammation, edema, and subsequent tissue damage that would lead to skin atrophy. This high-potency topical steroid is commonly used twice daily for one month, followed by once daily for another month, followed by tapering until a three-month follow-up is scheduled with the dermatologist.\textsuperscript{9} A majority of patients do benefit from therapy with topical steroids.\textsuperscript{5} Moisturizing agents aid in symptomatic relief, are often used in conjunction with topical steroids, and are a mainstay of long-term supportive therapy. In patients who have failed therapy with or those with contraindications to topical steroids, a trial of a topical calcineurin inhibitor such as tacrolimus ointment 0.1\% or pimecrolimus cream 1\% is reasonable.\textsuperscript{9} Calcineurin inhibitors are appropriate for management of genital or extragenital lichen sclerosus and have been shown to decrease exacerbations.\textsuperscript{5,9} Other considerations include methotrexate for cases of widespread disease and cyclosporine for refractory cases.\textsuperscript{9} Narrow-band UVB phototherapy and low-dose psoralen-UVA has also been shown to be effective for widespread extragenital lichen sclerosus.\textsuperscript{12} In consideration of the possibility of infection by \textit{Borrelia burgdorferi} as a cause of lichen sclerosus, some patients have benefited from treatment of intramuscular penicillin G following failed attempts of management with topical steroids.\textsuperscript{9}

Finally, surgery is a last resort and is generally limited to patients with severe cases of genital lichen sclerosus who have failed medical therapy.\textsuperscript{5}

The development of squamous cell cancer is a feared complication seen in 4.5\% of individuals affected by lichen sclerosus.\textsuperscript{5} These neoplastic changes are most commonly seen with female genital lichen sclerosus and are not a part of the natural history of extragenital lesions.\textsuperscript{11} This oncologic manifestation is seen an average of 10 years following diagnosis of lichen sclerosus.\textsuperscript{5}

Management and follow-up is fairly individualized due to the variability of the course of lichen sclerosus. Interestingly, most cases seen in children usually improve.\textsuperscript{4} However, it is imperative that patients with anogenital involvement be evaluated every six months to monitor for progression of disease.\textsuperscript{13}

**Conclusion**

Lichen sclerosus is a chronic inflammatory disorder that presents with genital involvement, extragenital involvement, or both. ECM-1 has been linked to lichen sclerosus; however, other autoimmune and endocrine etiologies, as well as infection by \textit{Borrelia burgdorferi}, are still being considered. Lesions have a predisposition for the anogenital region, are pruritic, and most commonly affect postmenopausal women. Biopsy is generally unnecessary but may be required to differentiate lichen sclerosus from other pathologies, including squamous cell carcinoma. There is generally a good response to high-potency topical steroids combined with emollients, but other options are available for resistant cases.

**References**

4. Larre Borges A, Tiodorovic-Zivkovic D, Lallas...


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