Lichen Sclerosus With Vaginal Involvement
Report of 2 Cases and Review of the Literature
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Importance
Lichen sclerosus (LS) is an uncommon chronic inflammatory disease that most commonly affects anogenital skin of postmenopausal women. It typically manifests as atrophic white plaques, which may be accompanied by purpura or fissuring. Rarely, LS has been observed to affect mucosal tissues in the mouth and the penile urethra. It is generally taught that LS does not affect the vagina, unlike lichen planus. To our knowledge, only one case report of LS with vaginal involvement exists in the literature.

Observations
Two cases of severe vulvar LS with vaginal involvement are reported. Both cases exhibited characteristic features of LS on vaginal biopsy, and both patients were followed up clinically without further treatment of the vagina.

Conclusions and Relevance
Vaginal LS may be more common than previously thought and may be underdiagnosed. Patients with more severe disease or with significant vaginal atrophy may be more likely to have involvement of the vagina. In addition, patients with pelvic organ laxity may be at increased risk if their vaginal walls are chronically exposed because of prolapse. Physicians managing patients with vulvar LS should be aware of the possibility of vaginal involvement so that vaginal lesions may be diagnosed and followed up appropriately.

Report of Cases
Case 1
A 59-year-old white woman was seen with vulvovaginal itching and vulvar rash associated with a sensation of rawness and burning for 1½ years. Physical examination revealed several well-demarcated white plaques. Purpura and fissuring were also commonly seen because of skin fragility. In women, a figure-eight pattern is classically described involving the vulvar and perianal skin. In a case series from the Southeast Vulvar Clinic, Charlotte, North Carolina (L.E., unpublished data, 2006), extragenital lesions were seen in approximately 6% of patients. Rarely, LS has been reported to affect the oral and penile urethral mucosa as well. It is generally taught that LS does not affect the vagina, unlike lichen planus.

We report 2 cases of LS involving both the vulva and the vagina. Using PubMed, a review of the English literature since June 1937 revealed one case report of LS with vaginal involvement by Longinotti et al in 2005. Our objective is to add 2 additional cases to the scant existing literature to learn more about the pathogenesis and natural history of this disease entity.

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tern. Also present were hyalinization of the papillary dermis and a bandlike infiltrate in the upper reticular dermis, diagnostic of LS.

During the next 4 years, the patient was treated with a combination of potent topical corticosteroid ointments, intral esional corticosteroid injections, and vaginal estrogen therapy. She had significant difficulty achieving and maintaining control of her symptoms because of the severity of her disease and multiple socioeconomic factors. She also experienced several cutaneous adverse effects during her therapy, including contact dermatitis in response to medication use, corticosteroid dermatitis and atrophy, and secondary infection, which were treated as needed.

Several years after her initial visit, routine examination revealed a purpuric patch on the distal anterior vaginal wall (Figure 2). Biopsy of this lesion was performed and showed classic features of LS (Figure 3). She has continued a maintenance regimen of topical corticosteroid therapy 3 times weekly and vaginal estrogen therapy, with fairly good control. The purpura resolved after biopsy.

Case 2
A 76-year-old white woman was seen with vulvovaginal itching, pain, rawness, and occasional bleeding of 2 years' duration. She had tried multiple ineffective therapies during this period, including vaginal estrogen cream, topical and vaginal antifungal creams, topical corticosteroid creams, and hydroxyzine hydrochloride.

Physical examination revealed a purpuric plaque with a thin, crinkled border covering the vulva, with hypopigmentation of the perineal body. Significant scarring was present,
with resorption of the labia minora (Figure 4). Vaginal examination showed mild redness of the vaginal mucosa but no other abnormalities. A rectocele was present. Her wet mount showed normal vaginal secretions. No oral lesions were present. A clinical diagnosis of LS was made.

During the next 3 years, the patient was managed with ultrapotent topical corticosteroid ointments and vaginal estrogen cream. She used oral fluconazole and oral antibiotics as needed for yeast and secondary bacterial infection, respectively. Control of her vulvar disease waxed and waned. About 3 years after her initial presentation, routine internal examination revealed a 2-cm white, cobblestoned plaque on the distal posterior vaginal wall (Figure 5). Biopsy of this lesion revealed abnormal hyperkeratosis, with hyalinization of the mucosal subepithelium, diagnostic of LS (Figure 6).

Discussion

Two cases of LS involving not only the vulva but also the vagina are reported, adding to the one previous case report by Longinotti et al. Histologic features of vaginal biopsy in each case were diagnostic of LS. This brings into question the true incidence of vaginal lesions in this disease. Is vaginal involvement as rare as the literature suggests, or are we underdiagnosing? Significantly more case reports of mucosal LS involving the oral cavity exist in the dental literature. Oral lesions are most commonly found on the lip or buccal mucosa, and approximately half of the patients are symptomatic. Mucosal LS is also described in the urologic literature. Barbagli et al demonstrated histologic involvement not only of the squamous urethral epithelium but also of the penile mucosal urethra in 12 of 16 male patients with LS.

In the same article, Barbagli et al propose that when LS involves a mucous membrane, it is always after the mucous membrane has become “squamatized.” It is postulated that in this process chronic irritation changes the mucous epithelium to a metaplastic squamous epithelium. With continued inflammation or irritation, the squamatized epithelium becomes hyperkeratotic and epidermis-like. If their theory is true, this may be a permissive factor in the ability of LS to affect mucosal tissues. Notably, both of our patients had significant pelvic organ prolapse so that the affected portions of their vaginal wall were more chronically exposed than would be normal. This brings into question whether squamatization of the vaginal mucosa may have had a role in the development of their vaginal lesions.

The risk of scarring and malignant neoplasm in vaginal LS is unknown but is likely small because neither has been reported to occur. Therefore, optimal management has not yet...
been defined. In general, if a vaginal lesion is identified, an initial biopsy specimen should be obtained for diagnosis (Figure 7). The lesion should then be followed up clinically to assess for early signs of malignant transformation. Therapy with topical or intralesional corticosteroids or vaginal estrogen therapy may be considered if patients are symptomatic.

In our opinion, attempts at self-monitoring of vaginal lesions in this patient population would be of low yield and would likely invoke unnecessary patient anxiety. The incidence of vaginal squamous cell carcinoma is exceedingly low. Even in the setting of vulvar lichen planus, a disease that routinely involves vaginal inflammation and scarring, there are no reports of vaginal squamous cell carcinoma. No evidence exists that vaginal lesions in LS lead to scarring or have any medical significance. Therefore, the focus for these patients should continue to be on achieving compliance and control of their vulvar disease. In addition, a speculum examination should be performed at each visit to improve detection and surveillance of vaginal lesions. Ideally, this would be done by a dermatologist, who is more likely to recognize inflammatory skin disease. However, if this is impossible, patients should be referred to a gynecologist for complete speculum examination.

In conclusion, LS of the vagina occurs and may be more common than previously thought because the vagina may not be examined carefully for LS or because lesions may be subtle or atypical. Lichen sclerosus may be more likely in patients with vaginal mucosa that is exposed because of prolapse. The risk of scarring and malignant neoplasm is unknown. Therefore, patients should be followed up carefully, including routine speculum examination, with the goal of optimizing patient outcomes and further defining the clinical spectrum of LS.

**REFERENCES**