Answer to Dermacase *continued from page 711*

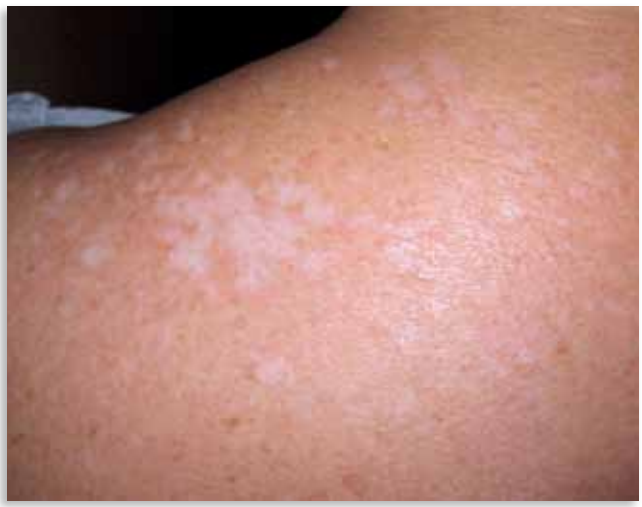
5. Lichen sclerosus

Lichen sclerosus (LS), also known as lichen sclerosus et atrophicus, is a chronic, progressive, inflammatory skin condition that can cause substantial discomfort and morbidity. It tends to affect women more commonly than men, with the female-to-male ratio ranging from 6:1 to 10:1.[1] The highest incidence of LS occurs in women older than 50 years of age, but prepubertal girls, women in their reproductive years, and men of any age are also affected. The actual prevalence of LS in the general population is likely higher than the reported prevalence because of frequent misdiagnosis and failure of asymptomatic patients to seek medical care.[2]

Lichen sclerosus most commonly involves the anogenital area (85% of cases), but extragenital lesions (15% of cases) can also occur.[1] Initially, the lesions of LS appear as ivory-white, well-demarcated, discrete, polygonal, flat-topped papules. With time, the lesions coalesce to form larger atrophic patches, often with overlying superficial telangiectasia, petechial and purpuric hemorrhages, and bullae. Fissures and erosions can also be present.[3] Occasionally, a hyperpigmented border of normal-appearing skin might surround the lesions. The isomorphic or Köbner phenomenon is often demonstrated by LS, as it can develop at sites of trauma, in old scars, and at sites prone to constant friction.[4]

In women, genital LS typically involves the vulvar and perianal regions, with occasional extension to the inner thighs; this often gives the lesions an "hourglass" or "figure-eight" appearance. The clitoral hood, labia majora, and labia minora can all be affected to varying degrees; advanced cases might involve the entire vulvar area (from the clitoris to the anus).[3] Pruritus, burning, and soreness (vulvodynia) are the most common symptoms experienced, although some patients can remain asymptomatic even with severe disease.[1] Additional symptoms are a consequence of anatomic distortions resulting from progressive scarring. Obliteration of the clitoral hood and fusion of the labia minora or majora can cause narrowing of the urethral meatus and vaginal introitus, which can lead to dysuria and dyspareunia, respectively.[5] Further, erosions and fissures in the perianal area can cause pain with defecation. This often leads to constipation, especially in prepubertal girls.[1]

In men, genital LS tends to affect the glans penis and foreskin and, in contrast to females, typically spares the perianal area.[1] Lichen sclerosus affecting the glans in men is also known as balanitis xerotica obliterans. The penile shaft and scrotum are less commonly affected. Pruritus, burning, and soreness are again the most common symptoms experienced.[1] Progressive scarring can cause narrowing of the urethral meatus and atrophy of the foreskin, which can lead to dysuria and phimosis, respectively. Phimosis is a common complication of



LS. In fact, up to 60% of acquired phimosis in prepubertal boys and at least 10% of acquired phimosis in adult males is associated with LS.[5]

Extragenital LS is less common than genital LS and more often tends to be asymptomatic. Although it can appear anywhere on the body, the most common sites of predilection (in both men and women) are the neck, shoulders, upper back, and chest.[3,5] Rare reports of oral mucosal LS also exist.

The exact etiology of LS has yet to be determined, but genetic, mechanic, infectious, and autoimmune factors are all thought to play a role in its pathogenesis.[1] Lichen sclerosus is strongly associated with several autoimmune disorders, including alopecia areata, vitiligo, thyroid disease, pernicious anemia, and type 1 diabetes mellitus.[1-3,5] This association occurs in about 20% to 33% of women with LS; it is present to a lesser degree in men.[5] Genital LS is also associated with the risk of developing vulvar and penile squamous cell carcinoma (SCC). The overall lifetime risk of developing vulvar SCC for females with genital LS is approximately 4% to 6%.[6] Extragenital LS is not associated with the development of SCC.[7]

Diagnosis

Although LS can be diagnosed on the basis of clinical appearance alone, a skin biopsy, ideally from the edge of a lesion, should be performed to confirm the diagnosis and rule out other entities that can mimic LS, especially a malignancy in cases of genital LS. The differential diagnosis of genital LS includes vitiligo, lichen planus, lichen simplex chronicus, vulvar intraepithelial neoplasia or SCC in situ, SCC, extramammary Paget disease, and scarring secondary to physical or sexual abuse. The differential diagnosis of extragenital LS includes morphea, scleroderma, vitiligo, tinea versicolor, and lichen planus. Owing to the strong association of LS with autoimmune disorders, screening complete blood count with vitamin B12, serum thyroid stimulating hormone, and

blood glucose levels should be ordered. For cases of genital LS, a skin swab should be considered, as concurrent infections are not uncommon; further, any nonhealing erosive or wart-like lesions should undergo biopsy to rule out malignancy.[1]

Treatment

Although a definitive cure for LS has yet to be discovered, the recommendation is that all patients (even those who are asymptomatic) must be treated to prevent the development of anatomic distortions from scarring and potential progression to malignancy.[2] Over the years, multiple treatment modalities have been employed for LS. These include a wide variety of topical, oral, light, and surgical therapies.

High-potency topical corticosteroids remain the mainstay of treatment. Despite the lack of evidence to support the use of one particular corticosteroid over another, clobetasol propionate 0.05% has become the preferred agent. Several different dosing regimens have been employed successfully, and no one regimen has proven more effective than another. One approach is to apply clobetasol to affected areas twice daily for 1 month, then once daily at bedtime for 2 months, followed by twice weekly for 3 months or as needed.[3] Another approach is to use clobatesol twice daily for 1 month, then once daily at bedtime for 2 months, followed by the use of a mid-potency corticosteroid for several months with eventual tapering to a low-potency corticosteroid for long-term use.[3] Regardless of dosage, the use of topical corticosteroids can substantially improve LS in children and adults, in terms of both symptomatic relief and clinical and histologic regression.[2] For genital LS, ointments are preferred over creams, as they are less likely to cause irritant contact dermatitis.[1]

For patients not responding to or developing irritant contact dermatitis from topical corticosteroids, there are a number of alternative treatment options that can be considered. Other topical therapies for LS include calcineurin inhibitors (tacrolimus and pimecrolimus), retinoids (tretinoin), and vitamin D3 analogues (calcitriol).[3,5] Topical testosterone, although previously the favoured treatment of LS, is no longer used owing to its lack of efficacy as well as its virilizing adverse effects in females (acne, hirsutism, clitoromegaly, and menstrual irregularities).[8,9] Intralesional injections of corticosteroids, such as triamcinolone, can be helpful for resistant disease. Oral therapies that have demonstrated some success include antimalarials (chloroquine and hydroxychloroquine), retinoids (acitretin), cyclosporine, and potassium para-aminobenzoate. Recently, phototherapy with low-dose UVA1 and 5-aminolevulinic acid photodynamic therapy have shown efficacy in treating extragenital LS.[3,5]

Surgery is rarely indicated for LS. Cryosurgery and laser ablation have both been employed, with varying success. Circumcision is the treatment of choice for phimosis; however, it is not curative for all cases of genital LS in uncircumcised male patients, as LS exhibits the Köbner phenomenon and often recurs in circumcision scars. Vulvectomy is only indicated in women with genital LS who have anatomic distortions, vulvar intraepithelial neoplasms, or cancer.[3] Additional agents might be required to treat the symptoms of LS. Topical or oral antihistamines, or both, can be used to relieve pruritus, with sedating antihistamines at bedtime being especially helpful in preventing nocturnal pruritus. Burning and soreness might respond to topical lidocaine or a low-dose tricyclic antidepressant, such as amitriptyline.[1,3]

Further treatment measures are required in patients with genital LS. Concurrent vaginitis and balanitis should be treated with appropriate antibiotics and antifungals. In addition, all patients should be advised to wash with bland emollients, avoid using strong soaps and detergents, avoid wearing occlusive clothing at night, and use adequate lubrication during sexual intercourse.[1] Patients should also be encouraged to perform monthly self-examinations of affected areas for suspicious skin changes that could be indicative of malignancy.

Given the substantial discomfort and morbidity associated with this condition, considerable stress and anxiety are likely to result. Issues relating to chronic pain, sexual dysfunction, and reduced quality of life should be addressed. Support groups and sexual counseling might be of benefit to some patients. Long-term follow up is recommended in all cases.

Ms Murynka is a third-year medical student at the University of Calgary in Alberta. **Dr Prajapati** is a first-year dermatology resident at the University of Alberta in Edmonton. **Dr Barankin** is a dermatologist practising in Toronto, Ont.

Competing interests
None declared

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