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Review

Lichenoid vulvar disease: A review☆

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ABSTRACT

Vulvar dermatoses are common, potentially debilitating conditions that can be seen by a variety of medical specialists. Lichenoid vulvar diseases, namely lichen sclerosus (LS), lichen planus (LP), and lichen simplex chronicus (LSC), can all negatively impact patients' quality of life and LS and LP also have an association with squamous cell carcinoma. It is essential that dermatologists are familiar with the unique features of each of these conditions to ensure the appropriate management and follow up. Herein, we provide an update on the epidemiology, clinical presentation, histopathology, and treatment of patients with vulvar LS, LP, and LSC.

Introduction

Lichenoid vulvar dermatoses, which include lichen sclerosus (LS), lichen planus (LP), and lichen simplex chronicus (LSC), are chronic inflammatory conditions that can manifest with a variety of symptoms, most commonly pruritus or pain. Although there are overlapping clinical characteristics of LS, LP, and LSC, each condition also has its own unique features (Table 1). An accurate diagnosis is essential for appropriate disease management and patient education because the natural history of these conditions differs. In this article, we provide an update on the epidemiology, clinical characteristics, and treatment of patients with lichenoid vulvar dermatoses with an emphasis on clinical pearls for dermatologists who may not be familiar with the management of vulvar disease.

Lichen sclerosus

Nomenclature

LS is a chronic inflammatory disease that was first identified in 1887 (Fistarol and Itin, 2013). It has since been referred to as kraurosis vulvae, vulvar dystrophy, white spot disease, lichen albus, guttate scleroderma, and lichen sclerosus et atrophicus (Fistarol and Itin, 2013; Schlosser and Mirowski, 2015).

Epidemiology

The exact prevalence of LS is unknown. It is estimated that the prevalence ranges from 0.1% to 1.7% with a higher prevalence reported in gynecology practices compared with dermatology practices (Goldstein et al., 2005; Wallace, 1971). A recent study determined the incidence of vulvar LS at 14.6 per 100,000 woman-years (Bleeker et al., 2016). Because LS can be asymptomatic and not always identified by physicians, its prevalence is likely greater than what has been reported. Although LS can occur at any age, it typically has a bimodal onset in prepubertal or postmenopausal women and is the most frequent among women who are postmenopausal (Wallace, 1971). Prepubertal LS was previously believed to resolve by puberty; however, several studies have refuted this notion (Powell and Wojnarowska, 2002; Smith and Fischer, 2009). In Smith and Fischer's prospective study of 12 patients with prepubertal LS, 75% of patients still had active disease after menarche (Smith and Fischer, 2009).

Etiology

The exact etiology of LS has not been fully elucidated; however, it is thought to be an immune-mediated disorder. LS has been

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associated with autoimmune diseases in up to 28% of patients (Cooper et al., 2008). Autoimmune thyroiditis is the most common concomitant autoimmune disorder, followed by alopecia areata, vitiligo, and pernicious anemia (Birenbaum and Young, 2007; Cooper et al., 2008). A familial predisposition has been reported in 12% of patients and an association with human leukocyte antigen DQ7, DQ8, and DQ9 has also been reported (Powell and Wojnarowska, 2001; Sherman et al., 2010). Although autoantibodies that target extracellular matrix protein may be elevated in the sera of patients with anogenital LS, it is thought that these autoantibodies are secondary and not directly pathogenic in the disease process (Fistarol and Itin, 2013; Oyama et al., 2003). Additionally, Howard et al. (2004) found that one third of patients in their cohort of 96 patients with vulvar LS had autoantibodies that targeted the basement membrane zone (BMZ). However, Gambichler et al. (2011) later reported no significant difference in BMZ autoantibody levels between patients with LS and control subjects.

Clinical findings

LS primarily affects the vulvar and perianal area and spares the vagina, although there have been rare reports of biopsy-proven LS that involve the vagina (Zendell and Edwards, 2013). LS can occur extragenitally in up to 13% of patients and presents exclusively extragenital in 6% of cases (Thomas et al., 1996; Wallace, 1971). Classically, vulvar LS appears as porcelain-white papules or plaques, which may have associated purpura, hyperkeratosis, fissures, erosions, or ulcerations (Figs. 1 and 2; Fistarol and Itin, 2013). A wrinkled or cigarette paper-like appearance is characteristic (Schlosser and Mirowski, 2015). Clitoral hood edema may be present as well as lichenification if patients scratch affected areas (Schlosser and Mirowski, 2015). Vulvar LS usually involves the interlabial sulci, labia minora, labia majora, clitoris, clitoral hood, perineum, and perianal area, forming a figure-of-eight pattern (Fistarol and Itin, 2013). Additionally, LS can extend into the genitocrural folds, buttocks, and thighs (Fistarol and Itin, 2013).

In a retrospective cohort study of 81 women, 76% of patients had labial involvement including 70% who had concurrent clitoral involvement (Lorenz et al., 1998). Perineal and anal involvement were present in 68% and 32% of patients, respectively (Lorenz et al., 1998). LS can leave scars later in the disease course and result in the fusion of the labia minora with the labia majora, complete resorption of the labia minora, fusion of the clitoral hood, and/or scarring of the introitus, which may lead to sexual dysfunction and in extreme cases interference with urination (Fistarol and Itin, 2013; Schlosser and Mirowski, 2015).

In children, LS often presents with vulvar purpura or ecchymosis, which may mimic sexual abuse (Powell and Wojnarowska, 2001). Additionally, children with LS may present with gastrointestinal complaints due to perianal involvement. In a study of 18 girls with anogenital LS, 89% reported at least one gastrointestinal complaint of whom 67% experienced constipation (Edwards, 2011; Maronn and Esterly, 2005).

LS can lead to vulvar lentiginosis, which appears as dark brown or black macules with irregular borders (Fistarol and Itin, 2013). Vulvar lentiginosis can be readily distinguished from melanoma on the basis of biopsy results. However, it is important to note that genital nevi in the setting of LS can mimic melanoma both clinically and histologically. Thus, pathologists should be informed if a genital nevus has been biopsied in the setting of LS. Although vulvar melanoma can occur in setting of LS, it is extremely rare (Fistarol and Itin, 2013).

Patients with LS can experience debilitating symptoms with pruritus as a hallmark of this condition (Fistarol and Itin, 2013). LS can also cause pain, burning, and dyspareunia (Fistarol and Itin, 2013). LS is asymptomatic in up to 39% of adult patients (Goldstein et al., 2005). In contrast, LS in children is usually symptomatic (Powell and Wojnarowska, 2001).

Pathology

Pathognomonic histological changes in patients with LS include hyperkeratosis, epidermal atrophy, and homogenization of collagen in the papillary dermis (Schlosser and Mirowski, 2015). Histopathology in patients with early LS, however, may be nonspecific and difficult to differentiate from LP because both involve a lymphocytic, lichenoid interface dermatitis (Fung and LeBoit, 1998). Fung and Leboit (1998) identified light microscopic criteria to help differentiate early LS from early LP. A psoriasiform lichenoid pattern, epidermotropism, atrophy, basement membrane thickening, and a decrease in papillary dermal elastic fibers with vertically oriented fibers were more common in LS lesions compared with LP ones (Fung and LeBoit, 1998).

Diagnosis

A diagnosis of LS can be made clinically; however, a biopsy can aid in the diagnosis or rule out other conditions in the differential diagnosis. As described previously, histology may be nonspecific, particularly in patients with early LS. In children with LS, a biopsy should ideally be restricted to treatment-refractory cases.

Disease course

LS is a chronic, relapsing condition that can result in loss of vulvar architecture and development of vulvar squamous cell carcinoma (VSCC). A recent study of 3038 women with histologically confirmed

Table 1
Distinguishing characteristics of vulvar lichen sclerosus, erosive lichen planus, and lichen simplex chronicus

	Lichen Sclerosus	Erosive Lichen Planus	Lichen Simplex Chronicus
Age of Onset	All ages with common onset in both prepubertal and postmenopausal patients	Not in children	All ages
Extragenital Involvement	Rarely	Yes	Yes
Associated Symptoms	Pruritus	Pain	Pruritus
Clinical Features	Porcelain-white papules or plaques; can have associated purpura, hyperkeratosis, fissures, erosions, or ulcerations	Erosions with white reticulations	Lichenified plaques
Common Distribution in Genital Area	Interlabial sulci, labia minora, labia majora, clitoris, clitoral hood, perineum, and perianal area	Medial aspect of labia minora and vestibule	Labia majora
Vaginal Involvement	No	Yes	No
Histopathology	Hyperkeratosis, epidermal atrophy, and homogenization of collagen in the papillary dermis, overlying a lymphocytic infiltrate	Lymphocytic and lichenoid dermatitis, wedge shaped hypergranulosis, numerous cytoid bodies, and pointed rete ridges	Epidermal thickening, hyperkeratosis, spongiosis, and acanthosis
Risk of Progression to Squamous Cell Carcinoma	Yes	Yes	No