

Lichen sclerosus of the vulva and squamous cell carcinoma

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ABSTRACT

Introduction: Lichen sclerosus is a disease whose features include skin and mucosa involvement. Its location in the vulvar area requires an approach both by gynecologists and dermatologists. An exhaustive monitoring of these patients must be made in order to improve their quality of life, and to prevent the disease transformation into squamous cell carcinoma.

Objectives: To determine the incidence of lichen sclerosus in our study group and its relation to squamous cell carcinoma.

Material and Methods: Our study included 32 patients with genital lichen sclerosus evaluated at the Vulvar Pathology Section along a five-year period.

Results: Lichen sclerosus represented 7.05% of consultations at the Vulvar Pathology Section. The age of the 32 evaluated patients ranged from 50 to 82 years with a mean age of 66 years.

Familial incidence was observed in one case (3.12%) in which both mother and daughter were affected. In 96% of the patients the main symptom was pruritus.

Of the 32 patients with lichen sclerosus, six (18.75%) were associated with squamous cell carcinoma, of whom four (66.7%) were differentiated VIN and two (33.3%) were invasive carcinomas. During follow-up, one patient died of complications arising from her tumor.

Conclusions: Lichen sclerosus is a frequent disease that should be assessed jointly by dermatologists and gynecologists. Malignant transformation in 18.75% of our patients justifies the need for a thorough clinical and histopathological follow-up with the aim of an early detection of neoplasias (Dermatol. Argent., 2011, 17(5): 365-369).

Keywords

lichen sclerosus of the vulva, squamous cell carcinoma.

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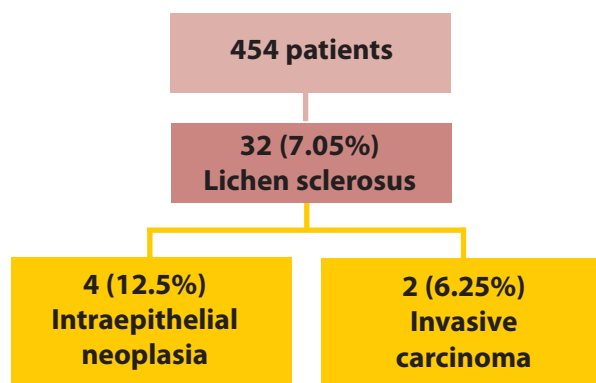
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Photo 1: Atrophic whitish plaque on vulvar and perianal region.

GRAPH 1: Incidence of lichen sclerosus and malignant transformation in our community



Introduction

Lichen sclerosus (LS) is a chronic dermatosis affecting skin and mucosa, especially the genital area¹. Even if it has been reported to affect all age groups indistinctly, a bimodal incidence is observed as it affects prepubertal girls and especially menopausal women².

Its pathogenesis is unknown. Clinically, its main features are whitish papules which converge and form plaques. Its most salient symptoms are pruritus, burning, dyspareunia, dysuria and painful defecation^{3,4}.

Several therapeutic resources are mentioned, the most common of which is topical application of corticosteroids and a transformation into squamous cell carcinoma has been reported in 4 to 6% of cases⁵⁻¹³.

The aims of this paper are to determine the incidence of lichen sclerosus in our study group, its onset age and main symptoms and its relation with squamous cell carcinoma.

Material and methods

Hospital Luis Lagomaggiore in Mendoza has a Vulvar Pathology Section where a dermatologist and a gynecologist work together and an oncologist and gynecological surgeon join them if necessary. This section only receives patients by derivation, so consultation about tumoral pathology is very frequent. Dermatoses are excluded because they are treated at primary attention centers.

A protocolized, descriptive, observational, cross-sectional trial was carried out including the patients with LS diagnosis examined between January 2003 and September 2008.

The diagnosis of LS was clinical. A repeat biopsy was performed on patients who presented suspicious lesions such as erosions, ulcers, hyperkeratotic areas or a change of color.

Follow-up was done every six months for the patients with typical LS lesions and monthly or every three months for the ones who presented any clinical finding which could lead to complications. The diagnosis of squamous cell carcinoma was anatomopathological in all cases.

Results

During the study period, 454 women were evaluated at the Vulvar Pathology Section. Out of all the patients evaluated, 7.05% (n=32) presented LS. Their ages ranged from 50 to 82 years with a mean age of 66 years and in 96% of the cases, the main symptom was pruritus followed by burning and dyspareunia.

Familial incidence was observed in one case (3.22%), in which mother and daughter were affected. No patient

presented clinical lesions compatible with infection by human papilloma virus simultaneously.

Out of the 32 patients with lichen sclerosus, 2 (6.25%) were associated with invasive squamous cell carcinoma and 4 (12.5%) with differentiated vulvar intraepithelial neoplasia (VIN) (Graph 1).

Mean time elapsed between diagnosis of LS and neoplasia was 18 months.

Comments

Lichen sclerosus (LS) was first described by Hallopeau in 1887 under the name of atrophic lichen planus. In 1982 Darier observed the histopathologic findings and called this entity lichen planus sclerosus^{14,15}.

It is a chronic dermatosis which affects skin and mucosa, especially the genital area³. Its prevalence in the dermatological consultation is estimated between 1 out of 300 to 1,000 patients³.

Even if it has been reported in all age groups, a bimodal incidence is observed as it affects prepubertal girls and especially menopausal women². It is more frequent in Caucasian patients and it develops both in genital and extragenital areas. In females, genital involvement is known as kraurosis vulvae⁴.

Its etiopathogenesis is unknown. However, hormonal factors, autoimmune disorders, genetic influence, infectious agents such as *Borrelia burgdorferi*, human papilloma virus, hepatitis C virus and trauma (Köebner phenomenon) have been postulated as possible causes³.

Its main symptoms are pruritus, burning, dyspareunia, dysuria and painful defecation⁴.

Clinically its main features are whitish papules which converge into plaques, often with erythema areas, ecchymosis, hyperkeratosis, fissures, excoriations and telangiectasias (Photo 1). Hyperpigmentation, blisters, ulcers and edema on the area may occasionally be observed. During the course of the disease, the skin becomes atrophic, whitish, shiny and fragile and adopts cigarette paper appearance, which may extend to the perianal region in a figure of eight (8) distribution^{4,16}. The final stage is characterized by the disappearance of subcutaneous tissue with a flattening of the labia folds, constriction of the vaginal opening and finally, loss of vulvar architecture, labial fusion and introital stenosis, which hinder coitus and urination³.

Histology evidences a thinned epithelium with flattening of the dermo-epidermal junction and varying degrees of keratinization. There may be spongiosis and vacuolar degeneration of the basal layer. The dermis looks edematous with hyalinization of collagen fibers, intense mixed inflammatory infiltrate and dilated blood and lymphatic vessels with red blood cell extravasation (Photo 2)³.



Photo 2: Atrophic epithelium, dermal edema and inflammatory infiltrate.



Photo 3: Invasive squamous cell carcinoma over lichen sclerosus plaque.

Resolution of LS is usually difficult. However, in some cases, especially in prepubertal girls, it may remit spontaneously².

The therapeutic resource of choice is the topical application of corticosteroids. The next most frequent, of controversial efficacy, are tacrolimus, pimecrolimus, calcipotriol, testosterone, retinoids, antimalarial agents, photodynamic therapy and surgery⁵⁻⁷.

In our paper, given the fact that it is about a Vulvar Pathology Section which only receives patients by derivation, the detected incidence was 32 in 454 women.



Photo 4: Differentiated vulvar intraepithelial neoplasia VIN (arrow) over lichen sclerosus plaque.



Photo 5: Differentiated vulvar intraepithelial neoplasia VIN (arrow) over lichen sclerosus plaque.

The main symptom was pruritus, which at times hindered the patients' sleep and intimacy. One of the patients mentioned the familial antecedent of her mother's death due to long-course lichen sclerosus of the vulva and further transformation into squamous cell carcinoma. This information was confirmed by clinical history.

Out of the 32 patients with LS, 2 (6.25%) were associated with invasive squamous cell carcinoma (Photo 3) and 4 (12.5%) with differentiated intraepithelial neoplasia (VIN) (Photos 4 and 5).

Malignant transformation of LS has been much discussed in the literature, but it is currently accepted that 4 to 6% of patients present lesions which turn into invasive squamous cell carcinoma⁸⁻¹³. The cause of this transformation would be the chronic inflammation brought about by LS, as no associations with human papilloma virus or decrease in the Langerhans cells have been detected which would justify anaplasia¹⁷.

We have observed a transformation into invasive squamous cell carcinoma in 6.25% of the cases in our study group, which together with the VIN findings, compel us to a very thorough follow-up of patients with LS with the aim of preventing the appearance of neoplastic lesions.

The main therapeutic resource we used was fluorinated topical corticoids for 30 days with a progressive reduction in frequency and strength according to clinical evolution. These patients have to be evaluated every six months and biopsies have to be performed of any suspicious areas (hyperkeratosis, erosions which do not heal after therapy or pigmentary changes).

As for the patients with diagnosis of VIN, a local extirpation of the lesion was performed and follow-up was carried out every six months. One of the patients with invasive carcinoma was performed a tumorectomy and the other a radical vulvectomy.

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