



BRIEF COMMUNICATION

Characterization of patients diagnosed with vulvar lichen sclerosus

Caracterización de las pacientes con diagnóstico de liquen escleroso vulvar

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ABSTRACT

Introduction: Vulvar lichen sclerosus is a chronic and progressive inflammatory dermatosis, underdiagnosed and undertreated, with a tendency to malignancy. Timely diagnosis and early treatment are essential.

Objective: To characterize patients with a diagnosis of vulvar lichen sclerosus attended in the health areas of Chambas municipality, Ciego de Avila.

Methods: An observational, descriptive, cross-sectional, case series study was conducted. The sample coincides with the universe so the sampling was intentional non-probabilistic. Women with vulvar lichen sclerosus, treated in Chambas municipality from January 2015 to September 2022, were included, patients with incomplete medical histories were excluded. Absolute and relative frequencies (percentage) were used as a summary measure.

Results: There was a predominance of the age group 60-69 years (66,7 %) and white skin color (71,4 %). 52,4 % belonged to the white group. The 52,4 % belonged to the Julio Castillo area. The mean age was 64,8±6,217 years. Vulvar pruritus and hypopigmentation were present in 100 % of the patients. Differentiated vulvar intraepithelial neoplasia developed in 61,9 % of the women. In 48,2 % of the cases the time of evolution was longer than three years. They received surgical treatment 71,4 %.

Conclusions: The disease predominated in the elderly; Hypopigmentation and vulvar pruritus were a constant in the patients studied, most developed a differentiated vulvar intraepithelial neoplasia with more than three years of evolution of the lesions and surgical treatment was the most applied behavior.

Keywords: Vulvar Lichen Sclerosus; Vulvar Pruritus; Vulvar Neoplasms.

RESUMEN

Introducción: el liquen escleroso vulvar es una dermatosis inflamatoria crónica y progresiva, infradiagnosticada y subtratada, con tendencia a la malignización. El diagnóstico oportuno y tratamiento precoz son fundamentales.

Objetivo: caracterizar las pacientes con diagnóstico de liquen escleroso vulvar.

Métodos: se realizó un estudio observacional, descriptivo, transversal, de serie de casos. La muestra coincide con el universo por lo que el muestreo fue no probabilístico de tipo intencional. Se incluyeron las mujeres con liquen escleroso vulvar, atendidas en el municipio Chambas desde enero de 2015 hasta septiembre de 2022, se excluyeron las pacientes con historias clínicas incompletas. Como medida de resumen de la información se utilizaron frecuencias absolutas y relativas (porcentaje).

Resultados: existió predominio del grupo de edades 60 – 69 años (66,7 %) y color de piel blanca (71,4 %). El 52,4 % perteneció al área Julio Castillo. La media de edad fue de 64,8±6,217 años. El prurito vulvar e hipopigmentación estuvieron presentes en el 100 % de las pacientes. El 61,9 % de las mujeres desarrollaron una neoplasia vulvar intraepitelial diferenciada. En el 48,2 % de los casos el tiempo de evolución fue mayor de tres años. Recibieron tratamiento quirúrgico el 71,4 %.

Conclusiones: la enfermedad predominó en la tercera edad; la hipopigmentación y el prurito vulvar fueron una constante en las pacientes estudiadas, la mayoría desarrolló una neoplasia intraepitelial vulvar diferenciada con más de tres años de evolución de las lesiones y el tratamiento quirúrgico fue la conducta más aplicada.

Palabras clave: Liquen Escleroso Vulvar; Prurito Vulvar; Neoplasias de la Vulva.

INTRODUCTION

Lichen sclerosus (LS) is a chronic, progressive,⁽¹⁾ and multifactorial inflammatory dermatosis of unknown etiology,⁽²⁾ affecting the anogenital region or other part of the body surface.⁽³⁾ It is underdiagnosed and therefore undertreated, so it tends to atrophy, scarring, functional deterioration and risk of malignant evolution, developing a differentiated vulvar intraepithelial neoplasia (VIN) or a spinocellular carcinoma (SCC).⁽⁴⁾

Francois Hallopeau first described the disease in 1887, as an atrophic form of lichen planus. Ferdinand Darier, five years later, named it lichen planus sclerosus. It received several denominations according to the specialty consulted and in 1976 the International Society of Vulvovaginal Disease adopted the term lichen sclerosus with which this disease is currently recognized.⁽⁵⁾

The disease is more frequent in childhood and peri- or post-menopause.^(4,6) The true prevalence is unpredictable because it is under-diagnosed. It is said to be 1 in 300 in adult women and 1 in 900 in girls.⁽⁶⁾ LE in 85-98 % of cases is located in the anogenital region and only 15-20 % in the extragenital region.^(6,7)

Symptoms include pruritus associated with irritation and tenesmus, dyspareunia, dysuria and anal symptoms.^(4,7) The cutaneous picture is characterized by papules or maculo-papules, flat, depressed with atrophy, fragility, excoriations and lichenification with edema of the labia minora; telangiectasias and purpura. In final stages, the labia majora and labia minora are sealed with partial or total occlusion of the vaginal introitus.^(4,7)

Considering the repercussions of this entity on skin, urinary and sexual health, as well as the potential risk of developing a differentiated VIN or CEC if not diagnosed and treated early, the interest arises to conduct the present research, with the aim of characterizing patients diagnosed with vulvar lichen sclerosus.

METHODS

An observational, descriptive, cross-sectional, cross-sectional, case series study was carried out on patients with a diagnosis of vulvar lichen sclerosus attended in dermatology and/or gynecology consultations belonging to the health areas of the municipality of Chambas, Ciego de Avila, in the period from January 2015 to September 2022.

The universe was constituted by all the patients attended in the place and period mentioned above (n=21). The sample coincides with the universe so the sampling was non-probabilistic of intentional type. The following inclusion and exclusion criteria were considered.

Inclusion criteria

Female population with a diagnosis of vulvar lichen sclerosus, who were attended in the health areas of the municipality of Chambas during the established period of time.

Exclusion criteria

Patients with incomplete clinical histories.

The following variables were taken into account: age group; skin color; health area; symptomatology; physical examination findings; tendency to malignancy, according to histopathological study; time of evolution of the lesions, taking into account the time elapsed from the onset of symptoms to diagnosis and treatment received.

Methods of obtaining information

The necessary information was collected from the individual clinical histories of the patients, using a data collection instrument created for this purpose by the authors of the research with the objective of organizing the variables to be analyzed, being the result of a correct anamnesis and physical examination of the patients, in adequate conditions of hygiene and illumination.

Statistical processing

The results were entered into a Microsoft Excel database, which was processed in the SPSS 18.0 statistical package for Windows. Descriptive statistics were used. Absolute frequencies, relative percentages, and measures of central tendency and dispersion (mean, median, mode and standard deviation) were used to summarize the information.

Ethical considerations

The research was approved by the institution's Scientific and Ethics Council. It complied with the principles of medical ethics and the aspects established in the Declaration of Helsinki.⁽⁸⁾

RESULTS

There was a predominance of the 60 to 69 years age group with 14 patients representing 66,7 %, those with white skin color (71,4 %), and those belonging to the Julio Castillo health area (52,4 %). The mean age was $64,8 \pm 6,217$ years (Table 1).

Table 1 Distribution of patients diagnosed with vulvar lichen sclerosus according to age group, skin color and health area to which they belonged.

Variables	N (%)
Age group	
50-59	3(14,3)
60-69	14(66,7)
70 y más	4(19,0)
Mean: 64.8 Median: 65 Mode: 67 Standard Deviation: 6,217	
Skin	
White	15(71,4)
black	4(19,0)
Mestizo	2(9,5)
Healthcare	
Polyclinic Julio Castillo	11(52,4)
Polyclinic Camilo Cienfuegos	5(23,8)
Polyclinic Máximo Gómez	5(23,8)

Source. Medical records

About 100 % of the patients presented symptoms of vulvar pruritus and hypopigmentation as physical examination findings. The most affected age group was 60 to 69 years old (Table 2).

Table 2 Distribution of patients according to symptomatology, findings on physical examination and age group.

Variables	Age groups							
	50 – 59		60 – 69		70 and more		Total	
	n	%	n	%	n	%	n	%
Symptomatology *								
Vulvar itching	3	14,3	14	66,7	4	19,0	21	100
Dispareunia	1	4,8	8	38,1	1	4,8	10	47,6
Dysuria	0	0,0	3	14,3	1	4,8	4	19,0
Anal symptoms	0	0,0	1	4,8	1	4,8	2	9,5
Findings on physical examination *								
Hypopigmentation	3	14,3	14	66,7	4	19,0	21	100
Areas of leukoplakia	0	0,0	9	42,9	3	14,3	12	57,1
Keratotic or parchymal plaques	0	0,0	6	28,6	1	4,8	7	33,3
Image in 8	0	0,0	8	38,1	2	9,5	10	47,6
Lichenification and/or ulceration	0	0,0	6	28,6	2	9,5	8	38,1
Echymotic plaques	1	4,8	2	9,5	0	0,0	3	14,3
Vulvar anatomical changes	0	0,0	2	9,5	1	4,8	3	14,3
* There were patients with more than one symptomatology and physical examination findings.								

Source. Medical records

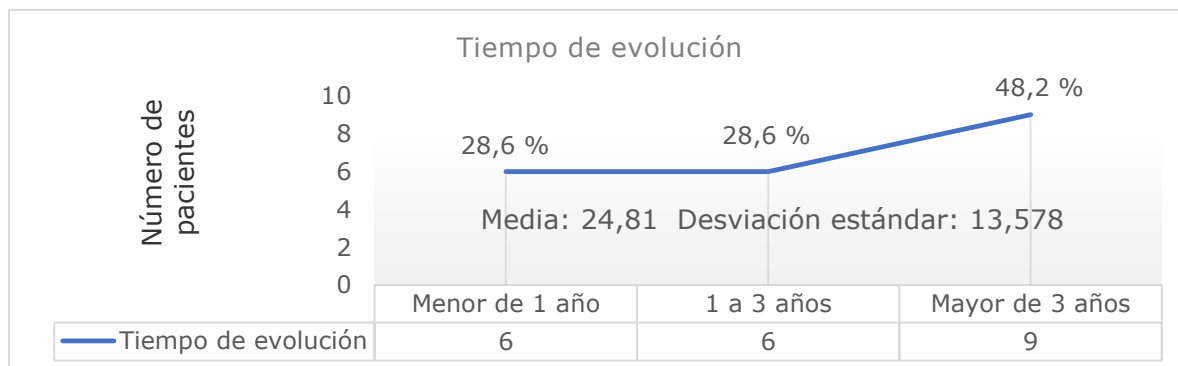
It was observed that 61,9 % of the women developed a differentiated vulvar intraepithelial neoplasia. (Table 3).

Table 3 Distribution of patients according to tendency to malignization of the lesions

Malignant tendency	Frequency	
	n	%
Differentiated vulvar intraepithelial neoplasia	13	61,9
Spinocellular carcinoma of the vulva	2	9,5
Lichen sclerosus without malignancy	6	28,6

Source. Medical records

About 48,2 % of patients presented more than 3 years of lesion evolution. The mean time of evolution of the lesions was $24,81 \pm 13,578$ months (Figure 1).



Graph. 1 Distribution of patients according to the time of evolution of the lesions.

Fifteen patients received surgical treatment, representing 62,5 %. (Graph 2)



Graph. 2 Distribution of patients according to treatment received.

DISCUSSION

LE has two peaks of incidence in prepubertal girls and in perimenopausal or postmenopausal women.^(4,9) It occurs in 10 % to 15 % in prepubertal patients.^(3,9) In this study, the women treated with LE are older than 50 years, with a predominance between 60 and 69 years of age, a fact that coincides with the literature consulted.

Of the three health areas that make up the municipality of Chambas, the highest incidence of the disease occurred in the Julio Castillo health area, as it is the area with the largest population.

The disease has a higher incidence in white-skinned women,^(1,4,7) as was the case in this study. In a study carried out in Brazil by Castro do Rego Barros,⁽¹⁰⁾ 83,1 % of LE predominated in black women, due to the fact that the research was carried out in an area with an Afro-American population.

It is described that vulvar pruritus and hypopigmentation of the genital area are a constant in most patients.^(4,7,10) Cabrera-Acea G et al.,⁽¹¹⁾ found that the clinical aspects of LE are diverse, but the predominant symptom is a pruritus of variable intensity, intermittent or permanent, at vulvar and/or perianal level. Barchino Ortiz L,⁽¹²⁾ found pruritus as the predominant symptom in 85,9 % of patients, followed by itching, dyspareunia, pain and presence of fissures. This fact coincides with the present investigation where pruritus is a constant symptom followed by dyspareunia.

The most frequent cardinal sign in this disease is hypopigmentation, followed by leukoplakia plaque, as described by Cortés-Ros O et al.⁽¹³⁾ and Cabrera-Acea G et al.,⁽¹¹⁾ in their respective investigations, which coincides with this study. Another research that yields similar results is the Doctoral Thesis of Barchino Ortiz L,⁽¹²⁾ where pearly-white plaques predominated (93,39 %).

LE appears to be an independent promoter of human papillomavirus (HPV) for carcinogenesis, which corroborates its potential for malignant degeneration,^(14,15) hence the importance of long-term follow-up of these patients.⁽¹⁵⁾

Gutiérrez-Pascual M et al,⁽¹⁶⁾ refer to the relationship of differentiated VIN with genital CEC. Both are entities highly linked to genital LE and independent of HPV infection. In addition, oncogenic molecular alterations independent of HPV infection have been demonstrated that could explain the malignant potential of LE itself.

Differentiated VIN is not causally related to HPV, this entity is associated with chronic inflammatory dermatoses such as lichen sclerosus and lichen simplex chronicus. Immunosuppression, advanced age and chronicity of lesions are associated with an increased risk of progression to CEC vulva.^(17,18,19) Falcón M F et al,⁽¹⁷⁾ agree that differentiated VIN occurs in postmenopausal women and is usually associated with lichen sclerosus.

All of the above discussed corresponds to what was found in this investigation where 61,9 % of the women with a diagnosis of vulvar LE developed a differentiated VIN.

The association between genital LE and CEC is widely documented in the literature, the estimated risk is 5 % of the cases, mainly in those of long evolution,^(16,17) in the present investigation most of the women had a period of more than three years of evolution of the disease, which is considered to be related to the development of malignant processes.

The use of high potency steroids, such as clobetasol propionate 0,05 % is considered first line treatment.^(14,20) In refractory cases methotrexate, cyclosporine and other immunosuppressants are used. Physical therapy is applied in cases of malignization or scar sequelae.⁽¹⁰⁾ Cabrera-Acea G et al.,⁽¹¹⁾ presented a case with LE, successfully treated with high potency local steroids, conjugated estrogens and psychological support.

Surgery remains the first-line therapy in the face of the development of a differentiated VIN, a CEC and to correct scar defects that severely interfere with the patients' quality of life.^(14,16)

In the present investigation the highest percentage of patients underwent surgical treatment because of the tendency of the lesions to malignize, which was in correspondence with the late diagnosis of the disease and the absence of treatment.

It is not possible to say whether medical treatment can prevent progression to malignization, but early diagnosis and initiation of treatment of these patients could not only reverse the lesions and reduce the rate of malignant transformation, but also prevent scarring and long-term sequelae. The present research is one of the outputs of an Institutional Project entitled: Predictive factors of malignant transformation in vulvar lichen sclerosus.

Being an infrequent pathology, a limitation of the study was the small size of the universe, which prevents the extrapolation of the results with other investigations.

The highest frequency of vulvar lichen sclerosus was in elderly patients, white and belonging to the Julio Castillo health area; hypopigmentation and vulvar pruritus were a constant in the patients studied, most of them developed a differentiated vulvar intraepithelial neoplasia with more than three years of evolution of the lesions and surgical treatment was the most applied conduct.

Conflict of interest

The authors declare that there is no conflict of interest with the preparation of this paper.

Authors' contribution

TAM: Project Management, Supervision, Visualization.

TAM, OE of LG: Conceptualization, Formal analysis, Validation, Writing the original draft, Writing, revising and editing.

LG SOs, DMHA, AGL: Data curation, Acquisition of funds, Research.

JCAH: Methodology, Resources, Software.

All approved the final version of the manuscript.

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