

Primary Cutaneous Lymphoma. About A Patient

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ABSTRACT

Introduction: Primary cutaneous lymphomas are defined as non-Hodgkin's lymphomas, which manifest in the skin without evidence of systemic involvement. Fungoidal mycosis is the most common cutaneous T-cell lymphoma along with Sezary syndrome. **Method:** A descriptive qualitative study was carried out using the bibliographic review technique, through analysis, from an integrative vision for which the Medline, Lilacs, Crossref, SciELO and academic Google databases were used, the keywords were: primary cutaneous lymphoma, fungoidal mycosis, non-Hodgkin's lymphoma, the time period was limited from 2018 to 2022. A total of 65 records were obtained, to which full articles referring to primary cutaneous lymphoma of the fungoidal mycosis type were applied as selection criteria, 12 articles were found that corresponded to it, which were selected for the study. **Objective:** To describe the behavior of primary cutaneous lymphoma of the fungoidal mycosis type through the presentation of a clinical case. **Case presentation:** We present the case of a 45-year-old patient with a history of psoriasis, which 10 months ago began with a chronic, macular-type skin lesions, with the presence of scaling and exulcerations scattered throughout the body. It is decided to enter for better study and treatment. **Conclusion:** It was of great importance to perform a biopsy in the presence of persistent hypopigmented lesions or those with unusual appearance to achieve timely diagnosis and early treatment.

Key words: Primary cutaneous lymphoma, Fungoidal mycosis, Non-Hodgkin's lymphoma

INTRODUCTION

Primary cutaneous lymphomas (PCLs) are a heterogeneous group of malignant lymphoproliferative processes that initially manifest in the skin without evidence of extracutaneous involvement at the time of diagnosis. They have a low incidence (7-10 cases × 106/year).⁽¹⁾ Primary cutaneous lymphomas are defined as non-Hodgkin's lymphomas, which manifest in the skin without evidence of systemic involvement. Fungoidal mycosis is the most common cutaneous T-cell lymphoma along with Sezary syndrome; represents 50% of cases and is characterized by

proliferation of atypical T lymphocytes with epidermotropism.^(2,3) In 2021, an annual incidence of cutaneous lymphomas was reported in Europe of 10 per million inhabitants.^(4,5) The type The most common form of cutaneous lymphoma is fungoidal mycosis, representing about 50% of all primary cutaneous lymphomas, which is widely described in the literature as a type of low-grade lymphoma with an indolent course due to its slow progression.^(4- 6) of the investigations published in the last two years, only cases were reported worldwide in Spain,⁽⁷⁾ Regionally in Mexico,^(2,3,6,7-9) Argentina (4,10,11) Costa Rica,⁽⁵⁾ and Cuba (12). The case reported in Cuba was in Havana, (12) the province of Villa Clara did not report any cases.

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The typical and characteristic clinical presentation is the progression of lesions that start as a patch, evolving into plaques and finally tumor lesions. Most cases develop in people over 50 years of age. (3, 6, 7)

Epidemiologic studies have failed to consistently identify viral-associated or environmental risk factors for most subtypes of cutaneous T-cell lymphoma. (4, 7)

The diagnosis and classification of primary cutaneous lymphomas are based on clinical characteristics and the results of histopathological, immunophenotypic and genetic studies. (7,8) Recent classifications include typical Fungoidalmycosis and its three variants: folliculotropic, reticulosispagetoid and granulomatous cutis laxa.(3,5,7,9)

The selection of treatment will be based on the corresponding staging. Skin-directed therapies are generally found to be appropriate for limited disease, whereas systemic agents are used for treatment of disease disseminated to blood and nodes.(10)

Due to the low incidence of primary cutaneous lymphomas at the global, regional and national levels, it is necessary to disseminate the research that is carried out regarding this pathology in order to use it by the medical community in a research teaching role, which has favorably repercussions in health care.

METHODS

An updated electronic search was carried out in bibliographic sources on topics related to primary cutaneous lymphoma of the fungoidalmycosis type, the Medline, Lilacs, Crossref, SciELO and academic Google databases were consulted. The following keywords were used: primary cutaneous lymphoma, fungoidalmycosis, and non-Hodgkin's lymphoma. The time period was limited from 2018 to 2022, a total of 65 records were obtained, to which complete articles referred to primary cutaneous lymphoma of the fungoidalmycosis type were applied as a selection criterion, 12 articles were found that corresponded to the same, which were selected for the study.

The methods of analysis, synthesis and systematization were used, which made possible the interpretation of the bibliography found and the organization of knowledge. Through the bibliographic review technique, an exhaustive analysis of the selected articles was carried out, which fully address the aspects related to primary cutaneous lymphoma of the fungoidalmycosis type, from an integrative perspective.

Presentation of the case:

Reason for admission: "skin lesions"

History of current illness:

A 45-year-old female patient, black, housewife, with a personal pathological history of psoriasis without a family pathological history, who denies toxic habits; who goes to the emergency room of the "Mártires del 9 de Abril" Hospital in Sagua la Grande, Villa Clara, because ten months ago she began with macular-type skin lesions, a chronic, with the presence of peeling and exulcerations, pruritic, scattered throughout the body. She is examined and it is decided to enter for better study and treatment.

Positive data on physical examination:

– Macular type lesions, a chronic, with the presence of desquamation and exulcerations of generalized topography. (Figures 1, 2 and 3).



Figure 1. Achromic macular lesions in the abdomen



Figure 2. Achromic and scaly macular lesions in the right upper limb



Figure 3. Achromic and scaly macular lesions in the lower limbs.

Complementary Exams

Hemoglobin: 13.8g/L

Leukogram: 9.4 x 10⁹/dL: Polymorph 0.64 Lymphocytes 0.45
Eosinophils: 0.01

Antibody for Hepatitis C Negative

Serology Non-reactive

HIV Negative

Peripheral Lamina

large leukocytosis of 32x10⁹ with lymphocyte predominance, with a large number of spindle lymphocytes and the presence of brain cells.

Medullogram

scarce inconclusive sample

Given the clinical and paraclinical findings, two possible approaches were expressed:

1-Primary cutaneous lymphoma with bone marrow infiltration?

2- Bone marrow lymphoproliferative process that infiltrates skin?

Skin Biopsy of Lesion

Epidermis with regular acanthosis and mild to moderate spongiosis, isolated apoptotic bodies and epidermotropism of occasional macrokaryotic lymphocytes and hyperchromatic and cerebriform nuclei. In the dermis, a diffuse band-like mononuclear inflammatory infiltrate interspersed with isolated lymphocytes with similar characteristics to those described above was identified.

Diagnosis

Cutaneous lymphoproliferative process compatible with fungoidalmycosis.

Immunohistochemistry

Predominance of CD8⁺ cells over CD4⁺, KI67 10%, CD20 negative, CD3 positive, CD5 positive.

Nosological Diagnosis

Primary cutaneous lymphoma of the fungoidalmycosis type the case is registered with an oncology consultation who assumes therapeutic behavior with polychemotherapy.

Favorable clinical evolution at this time. (Figure 4)



Figure 4. Achromic spots, no scaly lesions or exulcerations are observed

DISCUSSION

Primary cutaneous hypopigmented fungoidalmycosis lymphoma is a rare form of presentation. Most of the cases correspond to mixed forms, that is, in conjunction with erythematous plaques or tumors,(4,6-10) in the case of this patient the lesions were a chronic with scaling.

It occurs more frequently in children and young people, unlike the common fungoidalmycosis, which is diagnosed during the fifth or sixth decades of life, (5,6,10-12) the case presented was a 45-year-old patient of age, so it does not fit the most common age of onset.

It is more frequent in people with dark photo types or Asian individuals, (4, 10, 12) as in the case presented, which involved a black woman.

A complication sometimes associated with this disease is ulceration, (2, 4-11) which was not observed in the case presented.

The patient in the present case did not show lymphadenopathy or organomegaly on palpation. Differential diagnoses were: Hansen's disease, atopic dermatitis, vitiligo, extensive pityriasis alba, and pityriasis lichenoides chronica. From the histological point of view, the lymphocytic infiltrate with slight epidermotropism is characteristic, (4, and 8) as evidenced in this case. The predominance of the CD8⁺ phenotype of suppressor T cells in epidermal infiltrates is described as characteristic of the hypopigmented variety, (4, 8) which coincides with what was found in this patient.

Biopsy and histological study are of great importance for the diagnosis and treatment of this disease.

CONCLUSION

Primary cutaneous lymphomas have a low incidence in the world and within it fungoidalmycosis is the most frequent. It is of great importance to perform a biopsy in the presence of persistent hypopigmented lesions or of unusual appearance to achieve timely diagnosis and early treatment.

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