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Case Report

PRIMARY CUTANEOUS FOLLICLE CENTER LYMPHOMA TREATED WITH RITUXIMAB: A CASE REPORT

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Abstract

Rationale: Cutaneous B-cell lymphomas comprises a group of lymphoproliferative neoplasms of the B-cell non-Hodgkin's lymphoma type with extra nodal location, this type of neoplasms are indolent, so mainly it affects the cutaneous tissue, without any other type of systemic clinical manifestation. **Patient concerns:** A 53-year-old male with no chronic degenerative history, who came for evaluation due to the appearance of dermal lesions at the thorax**. Diagnoses:** Within the classification of cutaneous lymphomas, primary cutaneous follicle center lymphoma is mainly characterized by the appearance of dermal lesions of papules and erythematous plaques which expresses a phenotype with CD20+, CD79a+, CD5-, CD10+/-, BCL 6+ and MUM-1/IRF-4 negative. **Outcomes**. The use of biological agents (rituximab) combined with a surgical excision as the treatment for this neoplasm have a complete response in this type of primary lymphoma.

Keywords: *Non-Hodgkin lymphoma,* Primary cutaneous follicle center lymphoma*, CD 20+, Rituximab, Case report.*

INTRODUCTION

Non-Hodgkin's lymphomas are a group of hematological neoplasms that were described for the first time in 1832, initially identified in lymph nodes, and although most of the time they are identified in lymph nodes, they can also be found in extranodal tissue mainly in the central nervous system, gastrointestinal tract, urinary tract and skin (Goyal *et al.,* 2019; Ganapathi *et al.,* 2021). Cutaneous B-cell lymphomas comprise a group of lymphoproliferative neoplasms of the B-cell non-Hodgkin's lymphoma type with extranodal location, this neoplasm mainly affects the cutaneous tissue, without having another type of systemic clinical manifestation, currently these neoplasms represent approximately 25% of all cutaneous lymphomas (Cortés and Prins, 2014; Lara-Endara *et al.,* 2021). They are classified in three subgroups, according to the World Health Organization (WHO), which in 2007 divided them in three subgroups: 1. Primary cutaneous marginal zone, 2. Primary cutaneous diffuse large cell lymphoma of the legs, and 3. Primary cutaneous follicle center lymphoma, and in there is another type, named intravascular large B-cell lymphoma, which is considered as a different entity, characterized by a mucocutaneous ulcer consequence of the infection of Epstein-Barr virus, emphasizing that for the adequate classification of these, an immunohistochemical study must be done (Table 1) (Vitiello *et al.,* 2020). The diagnosis essentially requires a histopathologic study with adequate immunohistochemistry for this classification. As mentioned, this type of neoplasm is usually asymptomatic and its definitive treatment is surgical resection of the lesion, however, sometimes it behaves more aggressively. As for the variant discussed in the clinical case, it is the primary cutaneous follicle center lymphoma, which is a lymphoma that behaves in an indolent manner, this means that in most cases it is benign and its main clinical manifestation is the presence of skin lesions, and due to this benign behavior, the current treatment is focused on surgical excision for the purpose of complete removal of the lesion (Wilcox *et al.,* 2018).

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CASE

53-year-old male with no chronic degenerative history, who presents with the appearance of dermatosis in the anterior thorax, characterized by erythematous and erythematous-violaceous plaques, confluent at sternal level, with a rough and opaque appearance, with the largest being approximately 3 cm in diameter and the smallest approximately 1 cm (Figure 1 A and 1 b), non-pruritic, of three years of evolution, also reporting a weight loss of approximately 10 kg in 3 months. He went for evaluation to the internal medicine service, where he was approached to study these lesions, performing biopsy of these lesions, with hematoxylin and eosin staining, reporting a solution of continuity of the epidermis by a lymphoid neoplasm located in papillary dermis, atypical and scarce cytoplasm, some plasma cells and vascular congestion, resulting compatible with non-Hodgkin's lymphoma (Figure 2), with subsequent immunohistochemistry study, reporting the following pattern: CD20+, CD5-, CD10+, BCL 6+, BCL2- (Figure 3); compatible for primary cutaneous follicle center cell lymphoma.

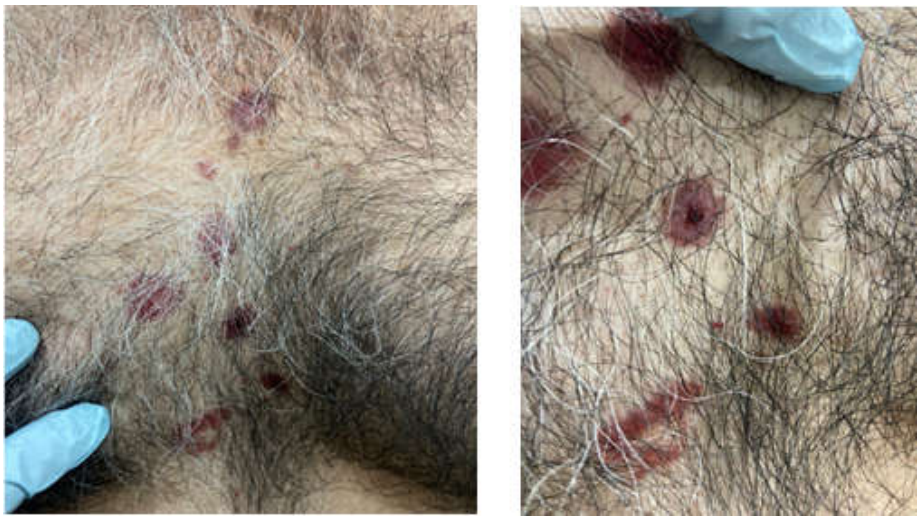
Due to these findings, it was decided to manage in conjunction with medical oncology, performing extension studies, with no report of lymph node or organic metastasis, so it was decided to completely remove with surgical approach of the reported lesions and then treated with 30 mg of rituximab per week for 2 months, with subsequent follow-up by the oncology service, currently without any report of appearance or extension of the lesions.

DISCUSSION

In the most used classification of cutaneous B-cell lymphomas proposed in 2007 by the WHO organization; the primary cutaneous follicle center lymphoma, is a lymphoma that behaves in a benign manner, represents approximately 60% of cutaneous B-cell lymphomas, its incidence has been increasing in recent years, it has an incidence of 4 per one million inhabitants.

Table 1. Classification of primary cutaneous B lymphomas according to the WHO, with their main clinical, topographic, behavioral, immunohistochemical pattern and frequency characteristics (Modified from: Oncol Front. 2020)

Histological type	Clinical presentation	Topography	Behavior	Phenotype	Frequency
Primary cutaneous marginal zone lymphoma	Small, solitary or multiple, purplish-red papules or nodules	Thorax, arms or head	Indolent	CD20+, CD79a+, BCL2+, CD5-, CD10-, BCL 6-, MAMÁ 1-	9%
Primary cutaneous follicle center lymphoma	Erythematous or erythematous-violaceous papules, plaques and/or nodules. Solitary or in clusters	Thorax, head or neck	Indolent	CD20+, CD79a+, CD5-, CD10+/-, BCL 6+, BCL2-, MUM-1/IRF-4 negative	12%
Primary cutaneous diffuse large cell lymphoma of the legs	Fast- growing erythematous- cyanotic plaques and/ or nodules	Legs	Agressive	CD20+, CD79a+, BCL2+, CD10-, BCL 6+/-, FOX-P1 y MUM-1/IRF-4 positive	4%



(A) **(B)**

Figure 1. A) Dermatosis located in the anterior thorax, with confluent erythematous-violaceous plaques of 3cm, 2cm and 1.5 cm in diameter. B) Dermatosis in zoom, with presence of erythematous macule at sternal level

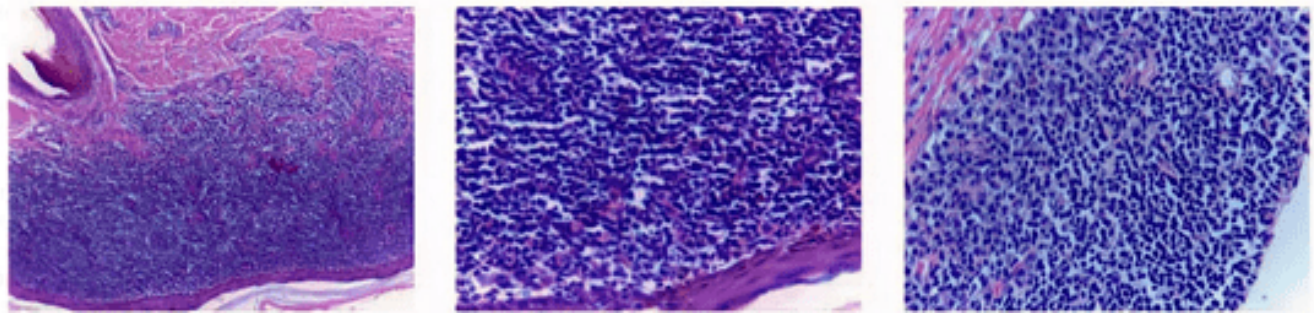


Figure 2. Hematoxylin and eosin staining characterized by a neoplasm of lymphoid origin, located in the papillary dermis with diffuse proliferation of atypical ovoid cells and scant cytoplasm

Figure 3.Immunohistochemistry of skin biopsy, positive for CD20, CD10, BCL, and negative forCD5 and BCL2, with a proliferation index of 95%, corresponding to a primary cutaneous follicle center lymphoma lymphoma, with an inmunnoblastic phenotype

It has a higher incidence in adults over 50 years of age, male and of Hispanic descent, likewise certain risk factors have been identified by antigenic stimulation such as some infectious processes, mainly by human immunodeficiency virus (HIV), human herpes virus type 8 (HHV-8), some bacterial infections, such as Borrelia burgdorferi and some autoimmune diseases such as in the context of Systemic Lupus Erythematosus (Skala *et al.,* 2018).

Currently the mechanisms related to malignant proliferation in the dermis is unknown, however, it represents a malignant proliferation of germinal center cells with the presence of centrocytes and centroblasts limited to the skin, it is characterized mainly by the appearance of dermal lesions such as papules and erythematous plaques or purpuric characteristic, may occur as solitary lesions or together but predominantly restricted to a specific region, mainly in the trunk, head and

neck, rarely presents with ulceration of the same (Olszewska-Szopa *et al.,* 2021). The diagnosis is based on a histopathological study of the lesions, mainly by excisional skin biopsy with immunohistochemical analysis, although dermoscopy can also help. The histopathological study is characterized by dermal infiltrate in different patterns that can be with follicular, follicular-diffuse and diffuse, these different patterns do not impact on the behavior of the disease, and the immunohistochemical study helps to determine the phenotype of the neoplasm, the characteristic pattern of this lymphoma is: CD20+, CD79a+, CD5-, CD10+/-, BCL 6+ and MUM-1/IRF-4 and BCL-2 negative (Willemze *et al.,* 2019; Alcocer-Gamba *et al.,* 2015). For therapy in solitary lesions, is required radiotherapy or complete surgical excision with complete curative intent, and intralesional therapies with corticosteroids or topical treatment with imiquimod or cryotherapy can also be considered; rarely a more aggressive therapy with chemotherapy R-CHOP will be required, or in its absence with rituximab, a chimeric monoclonal antibody against the CD20+, that in recent years has had great boom in the alternative therapy against lymphomas (Malachowski *et al.,* 2019; Fava *et al.,* 2022). For the follow-up of these patients, a clinical and nodal examination is required every 6 months. But they are generally considered lymphomas with good prognosis, they have been associated with a survival rate of more than 95% at 5 years of diagnosis, and even without treatment dermal lesions can remain stable without growth or even in some case reports have reported spontaneous disappearance, however, on the contrary there are also factors of poor prognosis, such as the location of the lesions in the lower extremities, which decreases survival to only 41% at 5 years. Recurrences are frequently observed in 30 to 50%, almost always limited to skin tissue (Fernández-Guarino *et al.,* 2013; Gamo *et al.,* 2008).

Conclusion

The aim of communicating this case was to highlight the clinical manifestation of cutaneous follicular center B-cell lymphoma in an immunocompetent patient in the fifth decade of life, due to the infrequency of primary cutaneous lymphomas. It also aimed to highlight the relevance of early clinical and histopathological diagnosis, and the different treatments that are nowadays available for a favorable outcome in these patients.

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