Primary cutaneous follicle center lymphoma*

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Abstract: Cutaneous lymphomas are classified according to their cellular origin into T-cell lymphoma and B-cell lymphoma. The annual incidence rate is 0.3 per 100,000 population. We report a case of a 56-year-old male patient who presented with a two-month history of nodules of varying sizes, some ulcerated, on the face, abdomen, and upper limbs. Histopathological examination and immunohistochemical study confirmed the diagnosis of primary cutaneous centrofollicular lymphoma. Studies have shown an increased incidence of non-Hodgkin lymphomas in the last decade. We report an infrequent case that should be kept as a differential diagnosis of patients with nodules and cutaneous papules.
Keywords: Lymphoma; Skin; Skin neoplasms

INTRODUCTION

Cutaneous lymphomas are classified according to their cellular origin into T-cell lymphoma and B-cell lymphoma. The annual incidence rate is 0.3 per 100,000 population. 65% of cases are T cells, 25% are B-cells, and 10% are true histiocytic lymphomas or other rare types of lymphomas. From the dermatological point of view, B-cell lymphomas are characterized by few lesions, which are in general nodules or infiltrates, presenting relatively fast growth. Unlike T-cell lymphomas, they show no pruritus.[1] *Primary cutaneous follicle-center* B-cell *lymphoma* is the most common subtype, representing about 55% of cases.[2]

CASE REPORT

A 56-year-old brown male patient, trader, noticed the appearance of "balls" on the abdomen 2 months before. He reported progressive growth and the emergence of new similar lesions associated with mild local pain. He denied itching or secretion drainage, as well as previous treatment. On physical examination, we found nodules of varying sizes, with a smooth and glossy surface, some ulcerated and erythematous on the face, abdomen, and upper limbs (Figures 1 and 2). A biopsy of one of the nodules revealed on histopathological examination atrophic epidermis and nodular neoplastic infiltration of the dermis under the Grenz zone at all levels. The subcutaneous tissue revealed dense aggregates of epithelioid cells with scant eosinophilic cytoplasm, vesicular nuclei, more than two nucleoli, nuclear-cellular pleomorphism, and frequent atypical mitosis figures. These findings were compatible with atypical lymphoid infiltrate. Therefore, we performed an immunohistochemical study to complete diagnosis, as well as blood count, chest, abdomen, head, and neck CT scans. These complementary exams showed no change (Figure 3). Immunohistochemical examination showed positivity for CD20, PAX-5, CD10, BCL-2, and BCL-6 (Figure 4). The patient was diagnosed with primary *cutaneous follicle-center lymphoma* and referred to a state oncology reference hospital. There, he was submitted to six chemotherapy sessions with the R-CHOP regimen (Rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) with complete remission of the skin lesions, which remained unchanged to imaging tests (Figure 5). The patient has been followed without signs of relapsing.

DISCUSSION

In the last decade, studies show increased incidence of non-Hodgkin lymphoma compared to other subtypes of skin cancer. Cutaneous lymphomas can be primary or secondary. The former is that of exclusively cutaneous involvement at the time of diagnosis and up to six months after, as evidenced in the present case by physical examination and complementary imaging tests. Although

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the two variants are identical in morphological appearance, they have different clinical behaviors. Primary cutaneous lymphoma features a more indolent natural history than the secondary form of the disease, with good prognosis.[3] It manifests itself at an average age of 58 years, consistent with our patient's age, with a 5-year survival rate of 95%. Cutaneous manifestations can be papules or nodules and their most frequent location are the head, scalp, and trunk.[4] Some authors believe that this type of lymphoma can be caused by

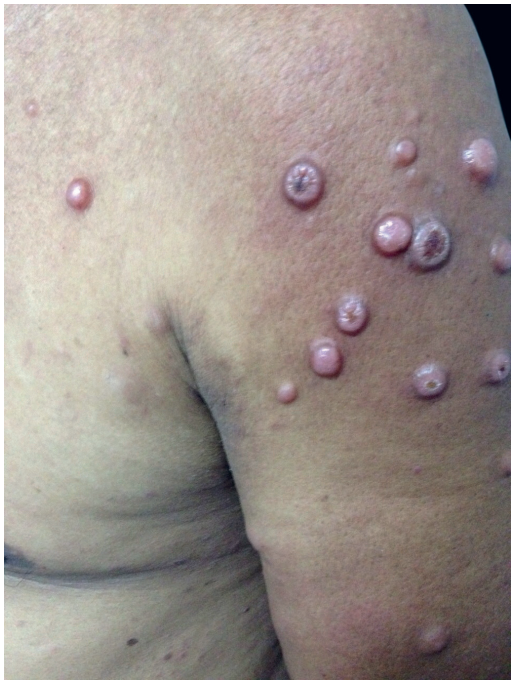

Figure 1: Erythematous nodules of varying sizes, some ulcerated, others covered by smooth and shiny skin

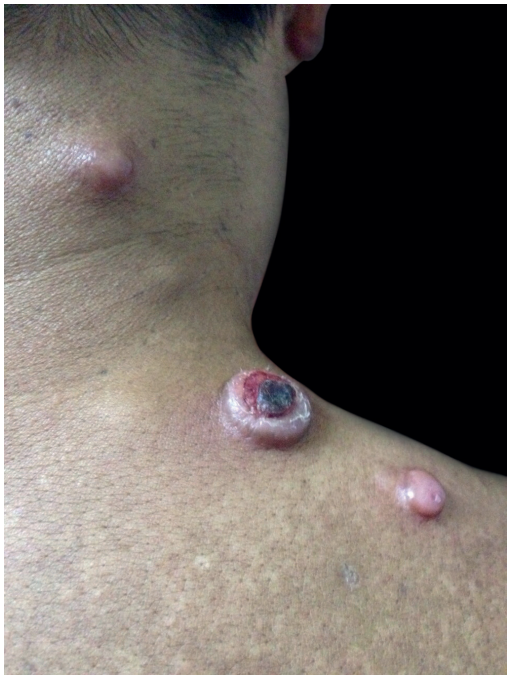

Figure 2: Nodules on the back

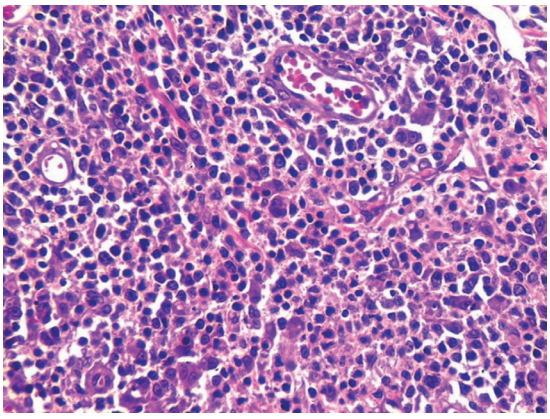

Figure 3: Dense aggregates of epithelioid cells with scant eosinophilic cytoplasm, vesicular nuclei, more than two nucleoli, nuclear-cellular pleomorphism, and frequent atypical mitosis figures. Hematoxylin & eosin, X40

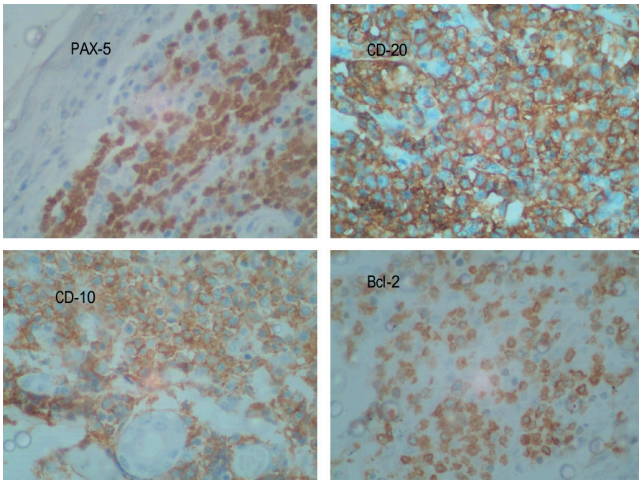

Figure 4: Immunohistochemistry. Magnification X40

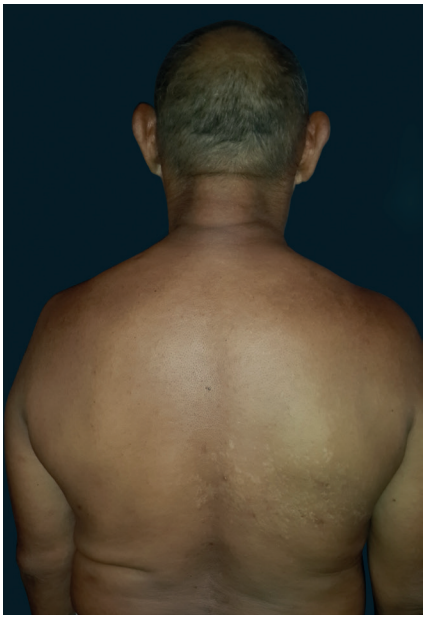

Figure 5: Remission of the skin lesions after treatment

prolonged antigenic stimulation possibly caused by chronic infection by specific microorganisms, especially by species of Borrelia. It should also be noted that these types of bacteria have been described in HIV patients and in patients undergoing methotrexate therapy, thereby suggesting that immune dysregulation may act in the development of these neoplasms.[5] Clinical history and physical examination help the diagnosis of primary cutaneous B-cell lymphomas. Confirmation, however, is obtained essentially by histological and immunohistochemical examinations.[2] Follicular center cells usually express CD20+, CD79a+, BCL-6+, and BCL-2- (possibly with a weak BCL-2 expression in a minority of B cells), with variable CD43 and CD106 expression. Treatment of choice is based on histology, anatomical location, and number of tumor lesions.[2] Radiation therapy is one of the preferred options in cases of a few lesions or if they are restricted to a single area. Although it can also be used in cases of late recurrences, the optimal dose and the number of treatment sessions has not yet been well established.[6] Chemotherapy with cyclophosphamide, hydroxydoxorubicin, vincristine, prednisone, and rituximab (R-CHOP) is another option, particularly for patients with multifocal and extensive skin disease and for patients who develop extra-cutaneous manifestations. In the present case, due to the large amount and distribution of nodules, we opted for chemotherapy, resulting in a complete remission of the skin lesions.[7] We report an infrequent case that should be kept as a differential diagnosis of patients with nodules and cutaneous papules. ❑

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