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A CD20 and CD3 negative variant of Lymphoma Cutis: A diagnostic challenge

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Abstract

Introduction: Cutaneous lymphomas (CLs) represent the clonal proliferation of neoplastic T or B lymphocytes and rarely of natural killer (NK cells) or plasmacytoid dendritic cells the diagnosis and classification of PCLs should always be based on a combination of clinical, histological, immunophenotypically and genetic. Treatment should be selected, taking into account the type of lymphoma and its stage and should be adapted to risk. Skin-directed therapies are the first choice for treating the early stages of the disease.

Case presentation: We present a case of cutaneous lymphoma on a 32-year-old woman. The diagnosis was established based on history, physical examination, histopathology, and immunohistochemistry (IHC) examination, which showed CD20 negative and CD3 negative. Patients were treated with an emollient and topical corticosteroid, as the results of the hyperpigmented plaques lesions regressed, and the patients are symptom-free at regular follow up 6-month later.

Conclusion: Topical corticosteroids can be used to treat refractory cases in early-stage disease. An important point in the diagnosis and classification of Primary Cutaneous Lymphoma (PCL) is based on a combination of clinical data and histopathological.

Keywords: Cutaneous Lymphoma; Cutaneous T-Cell Lymphoma; Cutaneous B-Cell Lymphoma

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Introduction

Lymphomas are a several group of malignancies that cover many different histological categories. Cutaneous lymphomas (CLs) are a proliferation of neoplastic T or B lymphocytes and rarely from natural killer cells (NK) [1]. There are two types of cutaneous lymphoma (CL), namely primary skin lymphoma (PCL) and secondary skin lymphoma, due to systemic lymphoma (SCL). Primary cutaneous lymphoma is diagnosed when there is cutaneous involvement, without evidence of lympho proliferative disease at another location at the time of diagnosis [2,3]. Primary cutaneous lymphoma of B-cell (CBCL) and T-cell (CTCL) is a heterogeneous group of non-Hodgkin's lymphoma (NHL) [4]. Cutaneous lymphoma is classified based on variations in clinical manifestations, histopathology, immune phenotyping, and genetic examination [1]. Here we report this rare a cutaneous lymphoma where immunohistochemical examination results showed negative CD20 and negative CD3.

Case Presentation

A 32-year-old woman was consulted from the Internal Medicine department with complaints of the emergence of purplish-red spots on both cheeks and ears, as well as blackish spots on the back and both hands for one year ago. In November, the patient went to the public health center and received therapy loratadine and paracetamol; there was no improvement. The internal medicine department diagnosed the patient with chronic lymphocytic leukemia. History of food or drug allergies was denied.

Physical examination and vital signs with normal limits. Dermatological status in the face region, right and left earlobe, back of the torso, right and left hand appear purplish red plaque and macular hyperpigmentation (Figure 1).

Routine blood laboratory examination found HB level of 9.5 mg/dl, WBC 72,000, PLT 26,000. Based on history and physical examination, the patient was diagnosed with a working diagnosis of Lymphoma Cutis.

Excision biopsy was performed, and the histopathological results contained lymphocytic cells arranged in a dense, diffuse scattered structure, the size of the nucleus was partially larger than mature lymphocytes, some were almost the same as mature lymphocytes, no Reed Stenberg Datia cells. Cells show infiltrative growth to subcutaneous fat (Figure 2).

These results contribute to Subcutaneous Lymphoma. Then it is recommended to do an immunohistochemical examination of CD 20 (cell B) and CD 3 (cell T), but the results are negative CD20 and

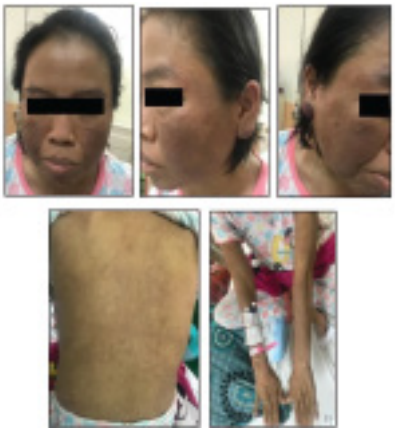

Figure 1: Day-1 Care. Face region and both earlobe showing purplish red plaque. Back of the torso, right and left hand show purplish red plaque and hyper pigmented macules.

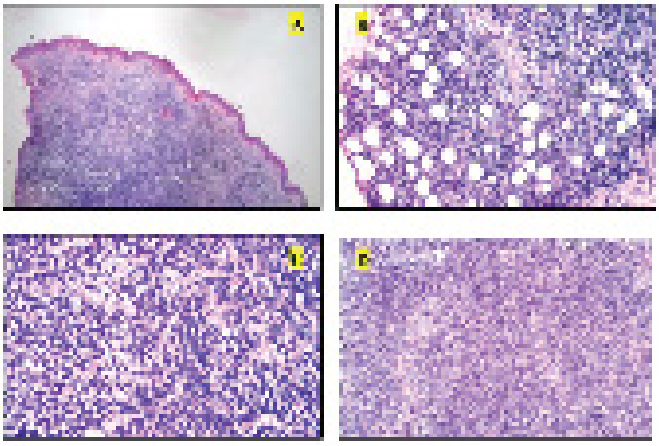

Figure 2: Histopathological examination. A) Epidermis without certain abnormalities. Upper-lower dermis of lymphocytic cells arranged in a solid diffuse arrangement, B) Lymphocyte cells infiltrate subcutaneous fat, C) core size larger than mature lymphocytes, D) Diffuse lymphocytic cells are dense.

negative CD3. Therapy was administered with a moisturizing cream (Morning-evening) and Desonide cream (face) once a day. Treatment from Internal department fludarabine 40 mg/day/oral.

During follow up in day 14, hyper pigmented plaques appeared reduced on both cheeks, and macular hyperpigmentation appeared to be reduced on the torso and hands (Figure 3). At regular follow up of 6-month patients reported the lesions regressed, and the patients are symptom-free with no evidence of disease relapse.

Discussion

We diagnosed this patient with cutaneous lymphoma. A study has been reported to investigate the relationship between cutaneous lymphoma, non-Hodgkin lymphoma, and chronic lymphocytic leukemia. Cases of cutaneous lymphoma are increased in patients with non-Hodgkin's lymphoma, and chronic lymphocytic leukemia and cases of patients with non-Hodgkin's lymphoma and chronic lymphocytic leukemia are increased after the presence of cutaneous lymphoma [5].

Cutaneous lymphoma (CL) is a clonal proliferation of neoplastic T or B lymphocytes and rarely of natural killer cells (NK) or plasmacytoid dendritic cells. History evaluation revealed a high fever followed by the appearance of reddish spots similar to mosquito bites on both hands and back. Pruritus was mild. This is quite in accordance with

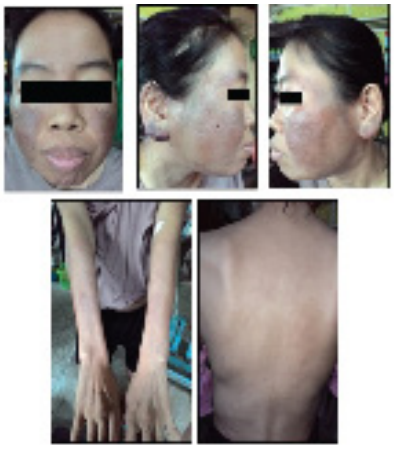

Figure 3: Day 14 follow-up, purplish red plaque and hyperpigmented macules appeared to be reduced on the torso and both hands.

the previous literature, which shows that the main symptoms of CL are skin lesions accompanied by pruritus and also signs of infection [6]. Lesions usually vary in shapes and colors, in the form of macules or plaques. Often these lesions will spread, making a generalized distribution. Often also accompanied by papules, hypopigmented, or hyperpigmented lesions [7].

The diagnosis is established through histopathological examination, which shows epidermis without certain abnormalities, the upper-lower dermis of lymphocytic cells arranged in diffuse dense, lymphocyte cells infiltrate subcutaneous fat, lymphocytic cells are solidly diffused, core size larger than mature lymphocytes and solid diffused lymphocytic cells [8].

After histopathological examination, and immunohistochemical examination is performed to determine the type of cutaneous lymphoma. This immunohistochemical examination looks for markers of T cell lymphocytes, B-cells, and NK cells. In this patient, a CD3 and CD20 examination was performed, which is a marker of T lymphocytes and B lymphocyte cells, but the results were negative. This is a rare case and is a challenge for clinicians to determine the right diagnosis for cases like this. Most cases report only one negative, for example on CD20 negative can occur in HIV cases, and the use of previous chemotherapy drugs (rituximab). This patient had previously received chemotherapy drugs in the form of fludarabine, where the working point of this drug is on T lymphocytes. The presence of such treatment is likely to be a negative marker.

The temporary treatment given to these patients is desonide cream. Topical corticosteroids can be used to treat refractory cases in early-stage disease and more advanced cases [9]. Topical corticosteroids work through the binding and activation of intracytoplasmic glucocorticoid receptors. In a broad sense, they act through anti-inflammatory and antiproliferative mechanisms. They seem to affect almost every stage of the inflammatory response. By stabilizing cell membranes and lysosomes, they inhibit phagocytosis and decrease monocytic and lymphocytic activity. They reduce chemical mediators such as interleukin (IL)-1, IL-2, interferon-gamma, tumor necrosis factor, and granulocyte-monocyte colony-stimulating factor. Topical corticosteroids further cause a decrease in mitotic activity and cause apoptosis of malignant cells, and its recommended to use it twice a day [10].

In this case, the patient gets cream emollients, or moisturizers contain occlusive properties that prevent TEWL (transepidermal water loss) that occur in CTCL due to inflamed skin that causes skin barrier

disruption so that the skin becomes dry, and/or humectants which help absorb water from the surrounding environment, increasing the water-holding capacity in stratum corneum. The most common of these humectants are glycerine, which has been shown to reduce corneocyte loss from the superficial epidermis and remodel the lipid barrier to reduce TEWL. To control the symptoms of pruritus or squama, the use of simple emollients can help. The recommended use of emollients is twice a day in cream form [10].

Six months after treatment, we did a follow-up visit, and the patient remained clinically asymptomatic. The prognosis of lymphoma cutis varies greatly depending on the extent of the skin involved, its type, the stage of the disease, and the involvement of the lymph nodes and metastasis to other tissues [6].

Conclusion

Topical corticosteroids can be used to treat refractory cases in early-stage disease and more advanced cases. An important point in the diagnosis and classification of Primary Cutaneous Lymphoma (PCL) is based on a history taking, physical, and histopathological examination.

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Conflicts of Interest

Nothing to declare.

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Ethical Approval

The study is exempt from ethical approval in our institution.

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