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A Rare Case of Primary Cutaneous Follicle Center Lymphoma Mimicking Leprosy

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Abstract

INTRODUCTION: Cutaneous B-cell lymphoma is a rare type of cutaneous lymphoma with a plethora of clinical manifestations ranging from macules, papules, nodules, or plaques. The lesions are often painless and certain subtypes such as the primary cutaneous follicle center lymphoma (PCFCL) are indolent. The disease is often misdiagnosed with other dermatoses including bacterial and fungal infections.

CASE REPORT: Here, we report a 55-year-old female patient with clinical manifestations resembling multibacillary leprosy that was then confirmed to be PCFCL through aid of dermoscopy and histopathology. The patient then undergone chemotherapy using the R-CHOP regiment which resulted in significant clinical improvement and no signs of metastases or extracutaneous involvement on follow-up.

CONCLUSION: PCFCL is a type of CBCL that has atypical clinical manifestations and can mimic other dermatoses such as leprosy. Careful examination and history taking are needed when assessing suspected cases and confirmation of diagnosis relies on histopathology and IHC staining. Chemotherapy is still the treatment of choice for multiple lesions, and in cases of PCFCL, they have a very good prognosis when treated adequately.

Introduction

Primary cutaneous lymphoma is a type of non-Hodgkin's lymphoma with a primary cutaneous manifestation without any extracutaneous involvement [1]. The prevalence of primary cutaneous lymphoma is rare with an incidence of 0.5−1 case per 100,000 population annually [2]. There are two subtypes of cutaneous lymphoma. The first type is the cutaneous T-cell lymphoma (CTCL), that accounts for 75−80% of all cutaneous lymphomas, and B-cell lymphoma (CBCL). In addition, there are various types of both CTCL and CBCL with different histological findings that also require different treatments [1].

As the name suggests, CBCL is a malignancy that originates from the lymphocyte B-cells with an incidence of three cases per 1,000,000 population [3]. The primary type of CBCL (PCBCL) can be further classified into four major subtypes, primary cutaneous marginal zone lymphoma (PCMZL), primary cutaneous follicle center lymphoma (PCFCL), diffuse large B-cell lymphoma, leg type (DLBCL-LT), and intravascular diffuse large B-cell cutaneous lymphoma (IVDLBCL). The first two subtypes are considered slow-growing/indolent, while the latter are more aggressive subtypes that are related to high mortality rate [2].

In general, cutaneous lymphomas have atypical clinical manifestation and diagnosis mostly relies on histopathological findings. In addition, there are various misdiagnoses most prominently with basal cell carcinoma (BCC) and only 16.3% of suspected cases were confirmed to be cutaneous lymphomas [4], [5].

Here, we report a case of a 55-year-old female patient with PCFCL that mimicked leprosy. The diagnosis was aided using dermoscopy and histological findings and showed significant clinical improvement after undergoing chemotherapy.

Case Reports

A 55-year-old female patient visited the dermatology clinic with the complaint of an erythematous plaque on the facial area 3 years before accompanied with an erythematous nodule on the left ear 2 months before. The patient did not experience any pain/pruritus. In addition, the patient experienced rapid weight loss of around 17 kg in the past 2 months. Before this visit, the patient was initially diagnosed with contact dermatitis and

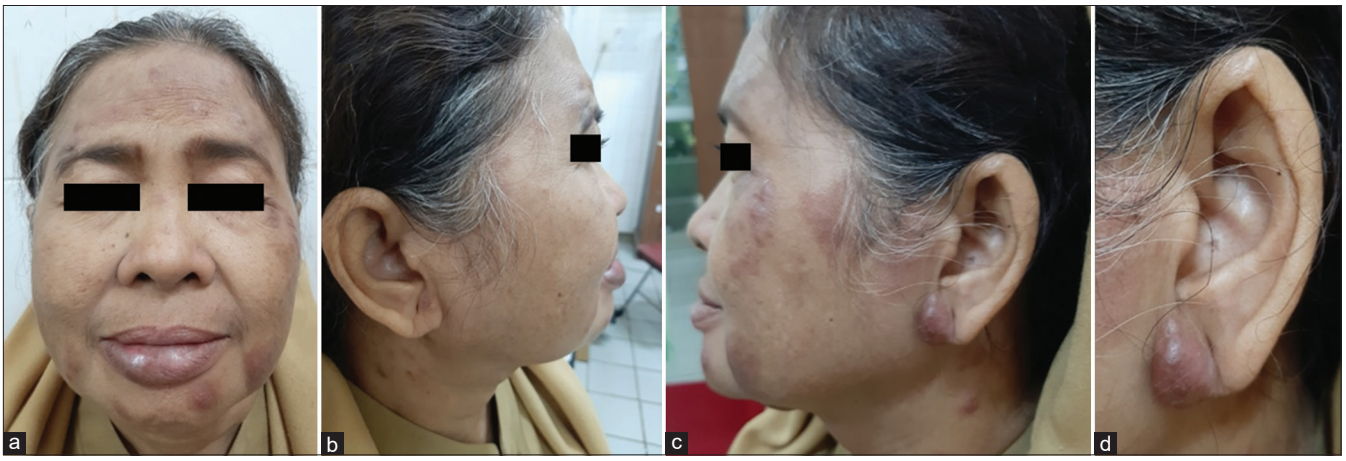

Figure 1: Erythematous and infiltrative plaques along with erythematous macules on the facial area (a-c), erythematous firm nodule on the left ear

received 4 mg methylprednisolone 3 times daily and topical betamethasone without any improvements. The patient had no prior health issues.

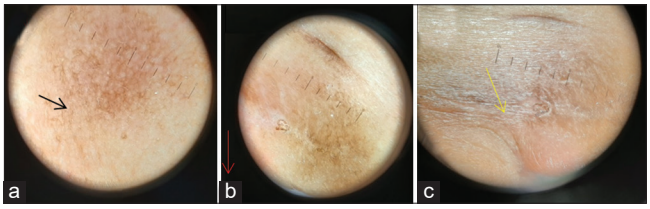

Figure 2: White Structureless Area (Black Arrow); Arborizing Vessels (Red Arrow); Scales (Yellow Arrow)

On physical examination, vital signs were within normal limits and there were no signs of enlarged lymph nodes, splenomegaly, or hepatomegaly. Dermatological examination found erythematous macules, multiple infiltrative plaques, and multiple hyperpigmented plaque on the facial area along with a solitary erythematous nodule with a diameter of approximately 2 cm that was firm on palpation on the left ear (Figure 1a-d). Dermoscopy examination found structureless area (Figure 2a), arborizing vessels (Figure 2b), and scales (Figure 2c). Routine blood results were within normal limits. Differential diagnoses for this patient include leprosy, cutaneous lymphoma, and lobomycosis. Subsequent 10% KOH examination and slit skin smear for mycobacterium were negative.

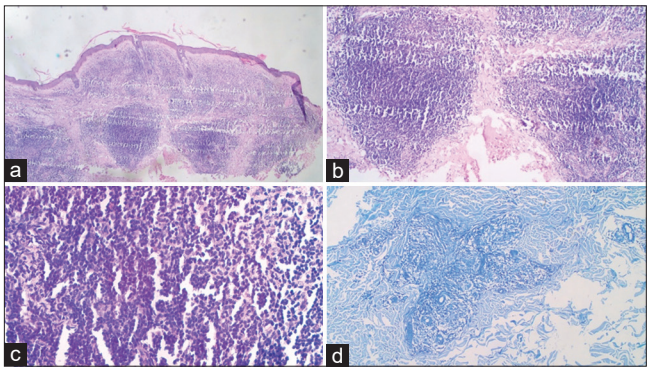

Figure 3: (a) Subepidermal Clear Zone (HE, ×4); (b and c) Lymphocytic Cell Proliferation Forming a Pseduofollicular Structure (HE, ×10 and ×40); (d) Negative Fite Faraco Stain

Histopathology examination found subepidermal clear zone along with lymphocytic cells with lobular structure forming a follicular appearance that extends to the skin adnexa. No granulomatous cells were found and staining using fite faraco was also negative (Figure 3a-d). These results help aid the diagnosis of cutaneous lymphoma. However, to determine the type of cutaneous lymphoma, a follow-up stain using immunohistochemistry (IHC) using CD20+ antibody was performed and yielded positive result (Figure 4a-b).With these results, the diagnosis of PCFCL was confirmed and the patient was referred to the internal medicine department and underwent six chemotherapy sessions using the R-CHOP regiment consisting of rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone with a 2-week interval. Significant improvements were seen after chemotherapy with resolution of cutaneous lesions on both the face and ears (Figure 5a-d).

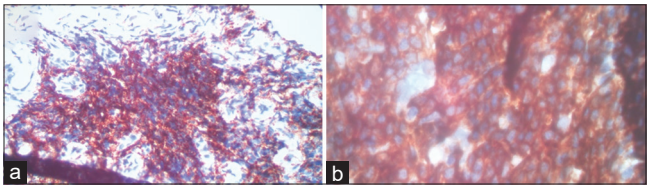

Figure 4: (a and b) Positive CD20 Antibody Indicated with Brown Color in the Cytoplasmic Membrane (IHC, ×10 and ×100)

Discussion

Primary cutaneous lymphoma is defined as clonal proliferation of lymphocytes that exclusively occur on the skin without any extracutaneous involvement [6], [7]. The skin is the third most common organ affected by extranodal non-Hodgkins lymphoma, with the highest people aged 55–65 years being the most group affected at 55−65 years [6].

Figure 5: (a-d) Clinical Improvements on Cutaneous Lesions after 6 Sessions of R-CHOP Chemotherapy

As previously mentioned, the cutaneous manifestation of cutaneous lymphoma is not specific and there are a plethora of clinical manifestations of cutaneous lymphoma such as macules, papules, plaques, or nodules that can mimic other dermatoses such as leprosy in our case. The appearance of infiltrative plaque as well as the solitary erythematous nodule on the left ear was misdiagnosed with multibacillary leprosy which is still common to be found in leprosy endogenous countries such as Indonesia. However, physical examination did not support the diagnosis of leprosy, as well as slit skin smear and histopathology examinations [3], [8].

Dermoscopy was also performed in this patient. However, there are still limited data regarding the diagnostic value of dermoscopy in cutaneous lymphoma. There are a few papers that reported dermoscoping findings such as white structureless areas, salmon-colored background area, scales, arborizing vessels, or a polymorphous vascular pattern. In our case, we found white structureless areas, scales, and arborizing vessels [5], [6].

In addition, histopathological findings using both HE and IHC staining suggest the diagnosis of PCFCL with the findings of lymphocyte infiltrates on the dermis and subcutaneous forming a follicular/diffuse appearance, while there were no abnormalities found on the epidermis. There are various antibodies that can be used in IHC staining to confirm the diagnosis of PCFCL, such as CD20, PAX5, CD10, BCL2, and BCL6, in which, our case showed positive result for CD-20 antibody [9]. PCFCL is considered as a slow growing subtype of CBCL, with a 95% 5-year survival rate [3], [9], [10].

In some cases, solitary lesions can be removed using excision or radiotherapy. A 30-40Gy dosage during radiotherapy can be administered to the patient along a 2 cm extended margin from the lesion and was found to be effective to treat lesions [11]. However, in cases of multiple lesions such as ours, the use of chemotherapy and intravenous rituximab is first-line treatments [3], [10]. The use of R-CHOP treatment was reported to be effective for the treatment of PCFCL, and after six sessions with a 2-week interval, the patient showed significant clinical improvements without any signs of metastases or relapse [12].

Conclusion

PCFCL is a type of CBCL that has atypical clinical manifestations and can mimic other dermatoses such as leprosy. Careful examination and history taking are needed when assessing suspected cases and confirmation of diagnosis relies on histopathology and IHC staining. Chemotherapy is still the treatment of choice for multiple lesions, and in cases of PCFCL, they have a very good prognosis when treated adequately.

Conflict of Interest and Informed Consent

The authors declared no conflict of interesting on preparing the manuscript and the patient in this case has agreed for her images to be used for publication and signed a written consent form.

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