

Primary Cutaneous Follicle Centre Lymphoma Presenting as a Giant Tumoral Lesion on Scalp

Tuğba Kevser Uzunçakmak,^{1*} MD Ayşe Serap Karadağ,¹ MD Hakkı Onur Kırkızlar,² MD İtir Ebru Zemheri,³ MD Emin Özlü,¹ MD Necmettin Akdeniz,¹ MD

¹ Istanbul Medeniyet University School of Medicine Goztepe Training and Research Hospital Department of Dermatology

² Istanbul Medeniyet University School of Medicine Goztepe Training and Research Hospital Department of Internal Medicine

³ Istanbul Medeniyet University School of Medicine Goztepe Training and Research Hospital Department of Pathology

E-mail: drtugbakevser@gmail.com

* Corresponding Author: Dr. Tugba Kevser Uzunçakmak, Istanbul Medeniyet University, Goztepe Research and Training Hospital, Dermatology Istanbul, Turkey

Published:

J Turk Acad Dermatol 2019;13 (4): 19134c3

This article is available from: <http://www.jtad.org/2019/4/jtad19134c3.pdf>

Key Words: Chemotherapy, Dermatopathology, Follicle Center Lymphoma

Abstract

Observation: Primary cutaneous follicle centre lymphoma is a low-grade malignancy and it is the most common subtype of primary cutaneous B-cell lymphoma. Clinically lesions usually present as a solitary or multiple well defined erythematous or purple plaque or nodule on face, the scalp or the trunk. Herein we want to present a case with primary cutaneous follicle centre lymphoma which presents as a giant tumoral lesion on scalp. We want to remind primary cutaneous follicle centre lymphoma in differential diagnosis of pink nodular lesion with its excellent prognosis.

Introduction

Primary cutaneous follicle centre lymphoma (PCFCL) is one of the subtype of primary cutaneous B-cell lymphoma[1,2,3]. It usually presents as a solitary or multiple well defined erythematous or purple plaque or nodule on face, the scalp or the trunk. Cutaneous recurrences are common, but systemic involvement is very rare. Solitary or small sized lesions may mimic inflammatory lesions such as acne or folliculitis, epidermal cysts and other cutaneous malignancies such as basal cell carcinoma according to the localization [4,5].

Case Report

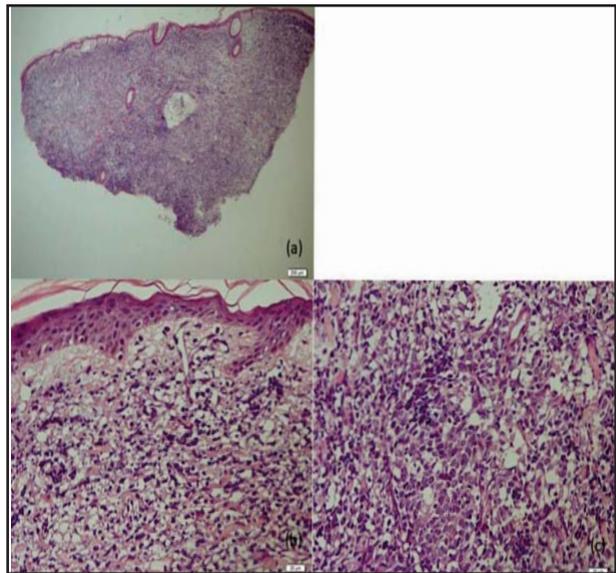
A 24-year-old female presented with a 2-year history of slowly growing nodular lesion on forehead.

(Figure 1) She first noticed it two years ago as an acneiform lesion and did not apply to another clinic for this lesion. Dermatological examination revealed a 5 cm in diameter, reddish, moderately infiltrated asymptomatic tumoral lesion on frontal region.

A 4 mm punch biopsy was performed from the thickest part of the tumoral lesion for histological evaluation. Histologically, diffuse, non-epidermotropic lymphoid infiltrate were seen in dermis. Lymphoid cells are comprised of predominantly centroblasts and variable proportions of medium-sized and large centrocytes. Immunohistochemically, lymphoid cells were stained with CD20, BCL-6, and not stained with BCL-2, CD10, MUM1/IRF4 (Figure 2 a,b and c). Focal remnants of follicular dendritic cell meshwork were positive for CD23. Ki-67 were 60% (Figures 3 a,b



Figure 1. 5 cm pink tumoral lesion on frontal region



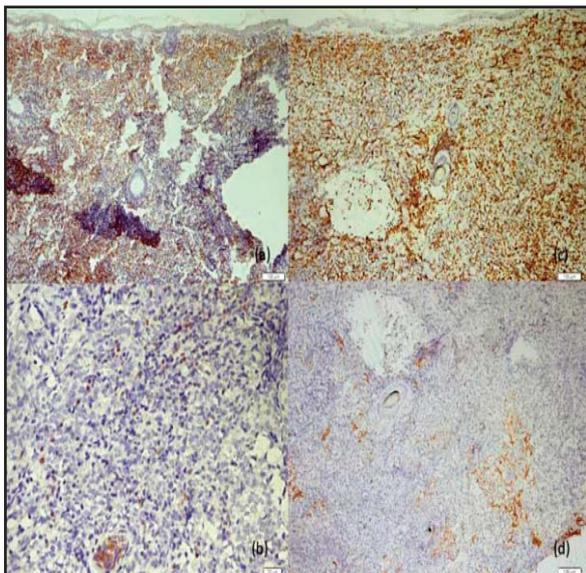
Figures 2 a,b and c. a. Diffuse lymphoid infiltrate in dermis b. Sparing the sub-epidermal "Grenz zone" c. Lymphoid cells comprised of predominantly centroblasts and variable proportions of centrocytes

and c). Her systemic scanning for metastasis revealed local involvement in jugular lymph nodes. No other lesion was detected in systemic scanning with PET-CT. Her laboratory tests were also normal. She was consulted to Haematology department and multiagent chemotherapy including cyclophosphamide, doxorubicin (hydroxydaunomycin), vincristine and prednisolone (CHOP) was initiated. After 3rd session her lesions were markedly regressed. After 6th session total clea-

rance was detected clinically. She is still in remission without any recurrence for 2 years also no aggravation was detected during her pregnancy after first year of chemotherapy (**Figure 4**).

Discussion

Primary cutaneous follicle centre lymphoma (PCFCL), a low-grade malignancy, is the most common subtype of primary cutaneous B-cell



Figures 3a,b, and c. a. CD20 positive Lymphoid cells stained with bcl6, b. not stained with bcl2, c. Ki67 were 60% d. follicular dendritic cell meshwork stained with CD23

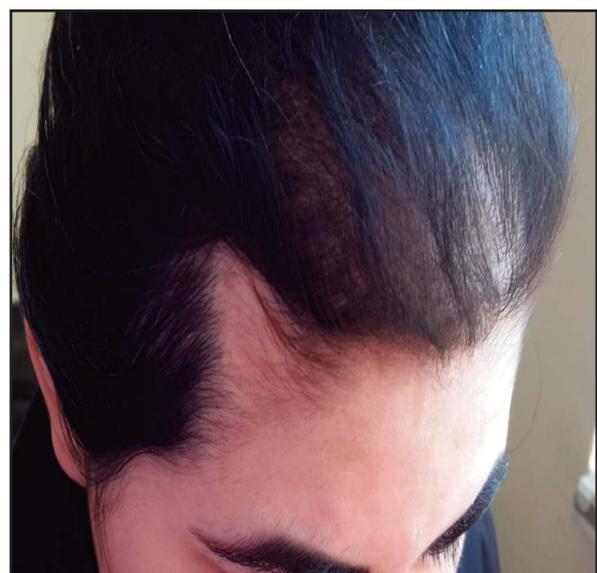


Figure 4. Excellent response to multiagent chemotherapy

lymphoma and represents about accounts for approximately 50% to 60% of primary cutaneous B cell lymphomas and 10% of all primary cutaneous lymphomas [1,2,3].

Histologically, the tumor may display a neoplastic follicular, follicular and diffuse, or purely diffuse infiltrate of predominantly follicular centre small or large centrocytes in dermis and subcutaneous tissue [3,4].

It is characterized by an indolent clinical course with a 5-year survival rate of more than 90%.[4] For isolated, small-sized, lesions, therapy options include local radiotherapy or excision, although intra-lesional interferon-alpha or intra-lesional rituximab can also be discussed [6]. In cases with large and/or multifocal PCFCL, local or regional radiotherapy or intravenous rituximab alone may be chosen. In some cases, Rituximab cyclophosphamide, vincristine, prednisone (R-CVP) or rituximab-cyclophosphamide, adriamycin, vincristine, prednisone (R-CHOP) chemotherapy can also be used, especially in disseminated or relapsing diseases[6]. In our patient we achieved excellent response with CHOP therapy.

We want to present this case to remind primary cutaneous follicle centre lymphoma in differential diagnosis of pink nodular lesion with its excellent prognosis.

References

1. Wilcox RA. Cutaneous B-cell lymphomas: 2015 update on diagnosis, risk-stratification, and management. *Am J Hematol* 2015; 90: 73-76. PMID: 25535037
2. Suárez AL, Pulitzer M, Horwitz S, Moskowitz A, Querfeld C, Myskowski PL. Primary cutaneous B-cell lymphomas: part I. Clinical features, diagnosis, and classification. *J Am Acad Dermatol* 2013; 69: 329. PMID: 23957984
3. Hope CB, Pincus LB. Primary Cutaneous B-cell Lymphomas. *Clin Lab Med* 2017; 37: 547-574. PMID: 28802500
4. Wanczinski MI, Pinto CA, Trevisan F, Cunha PR, Grohs LM. Primary cutaneous follicle center lymphoma simulating basal-cell carcinoma on the nasal ala. *An Bras Dermatol* 2015; 90: 111-114. PMID: 26312690
5. Chockalingam R, Aneja S, Patel AB. Primary cutaneous follicle center lymphoma mimicking folliculitis. *Cutis* 2018; 101: E30-E33. PMID: 29529119
6. Suárez AL, Querfeld C, Horwitz S, Pulitzer M, Moskowitz A, Myskowski PL. Primary cutaneous B-cell lymphomas: part II. Therapy and future directions. *J Am Acad Dermatol* 2013; 69: 343. PMID: 23957985