



Primary Cutaneous Anaplastic Large Cell Lymphoma: A Case Report

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Abstract

Primary cutaneous anaplastic large cell lymphoma (PCALCL) is a rare cutaneous CD30 + T cell lymphoproliferative disorder. PCALCL usually presents as solitary and localized nodules and tumors, or may be multiple and ulcerated. Extracutaneous involvement may occur rarely. Systemic anaplastic large cell lymphoma with secondary cutaneous involvement has a similar clinical appearance to PCALCL, but differs in treatment and prognosis. Thus, systemic evaluation of patients considered as PCALCL is critical. An 82-year-old male patient presented with three progressive lesions on the scalp. The final diagnosis was PCALCL with extracutaneous involvement based upon the involvement of regional lymph nodes, liver and spleen, lack of systemic B symptoms and the results of histopathological and immunophenotypical studies. This case is presented to emphasize the necessity of systemic evaluation in patients considered as PCALCL and to remind how to differentiate PCALCL from systemic lymphomas.

Keywords: Primary cutaneous anaplastic large cell lymphoma, cutaneous lymphoma, CD30 positivity

INTRODUCTION

Primary cutaneous anaplastic large cell lymphoma (PCALCL) is a rare CD30 + T cell lymphoproliferative disease of the skin (1). PCALCL is clinically seen as one or more ulcerable nodules and tumors (2). Rarely, extracutaneous involvement may occur (3). Cutaneous involvement secondary to systemic anaplastic large cell lymphoma is clinically similar to PCALCL, but treatment and prognosis vary (4). Therefore, it is important to investigate systemic disease in patients with PCALCL. The diagnosis of PCALCL is usually made by clinical findings, histopathological and immunophenotypical examination and the absence of systemic disease (5).

CASE REPORT

An 82-year-old male patient presented to our clinic with three lesions on the scalp that grew and became a mass. The history revealed that the first lesion had occurred one year ago and had

occasional bleeding; the other two had developed in the last 2 months and had no symptoms. Dermatological examination revealed a 3x3x1.5 cm sized, hard, brown-red-colored nodular lesion with hemorrhagic crust and yellow squama on the right frontoparietal region. Two 1.5x1.5x0.5 cm sized, brown-red-colored, nodular lesions were observed on bilateral posterior parietal regions (Figures 1, 2). He did not have weight loss, night sweats, decreased appetite, and fever. Histopathological examination revealed a large infiltrate of atypical lymphocytes in the dermis. Immunohistochemically, cells were positive for CD30, CD4, CD3, CD45 RO, MUM1, and negative for CD20, anaplastic lymphoma kinase (ALK), epithelial membrane antigen (EMA), Melan A, S100, BCL6, glial fibrillary acidic protein and Epstein-Barr virus. The KI 67 proliferation index was 90% (Figures 3-9). Laboratory tests and thoracic computed tomography (CT) evaluating systemic involvement were normal. Abdomen CT examination revealed a 10 mm hypodense nodule in the left and right lobes of the liver and 1 mm in the medial spleen.



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Figure 1. A 3x3x1.5 cm sized, hard, brown-red nodular lesion with hemorrhagic crust and yellow squama on the right frontoparietal region

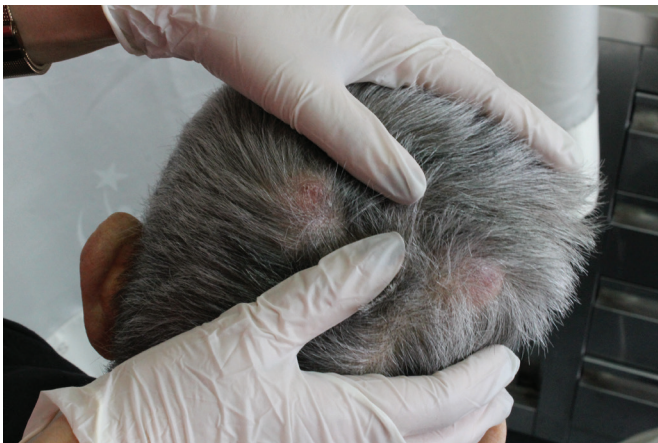


Figure 2. Two 1.5x1.5x0.5 cm sized, brown-red nodular lesions with a slight squama on the posterior of the bilateral parietal region

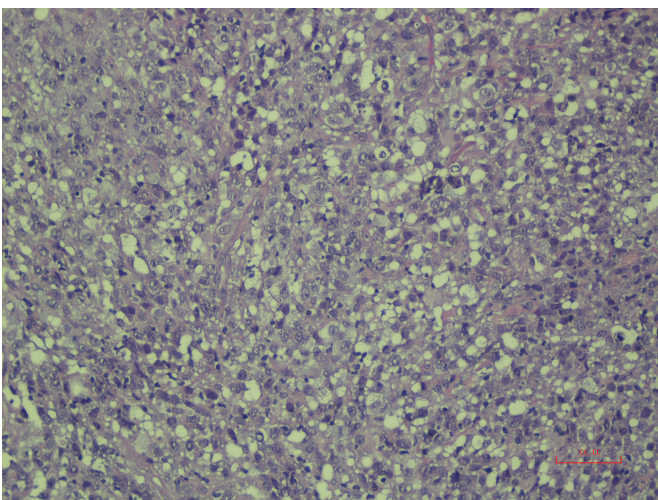


Figure 3. Infiltrate of large atypical lymphocytes in the dermis (hematoxylin and eosin, x100)

Positron emission tomography examination showed increased F-18 fluorodeoxyglucose uptake in localized sites that matched the skin-subcutaneous lesions on the scalp, left posterior cervical

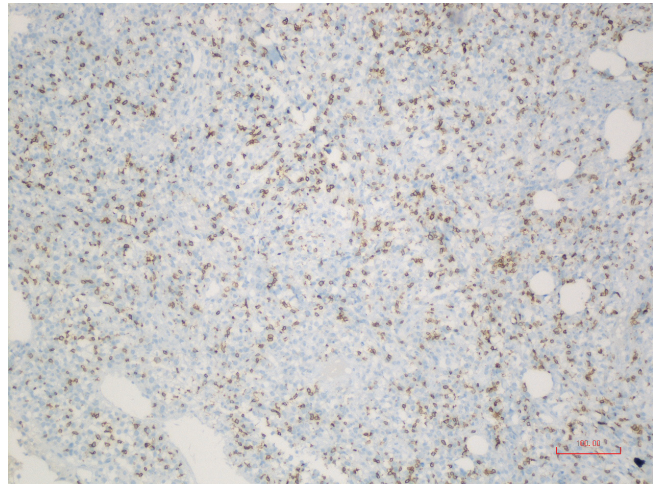


Figure 4. Positive staining with CD3

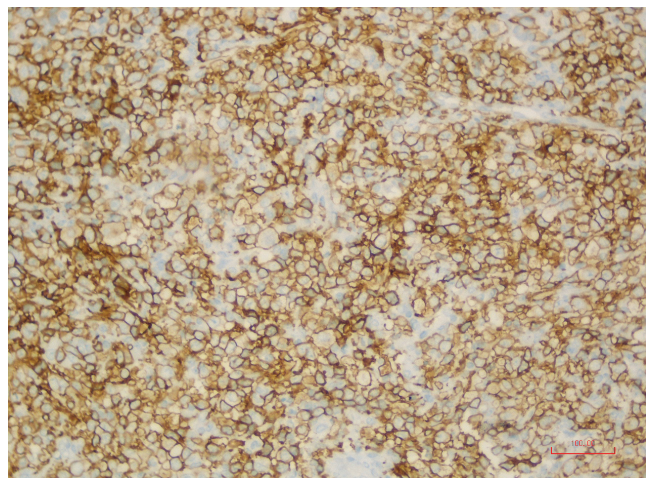


Figure 5. Positive staining with CD30

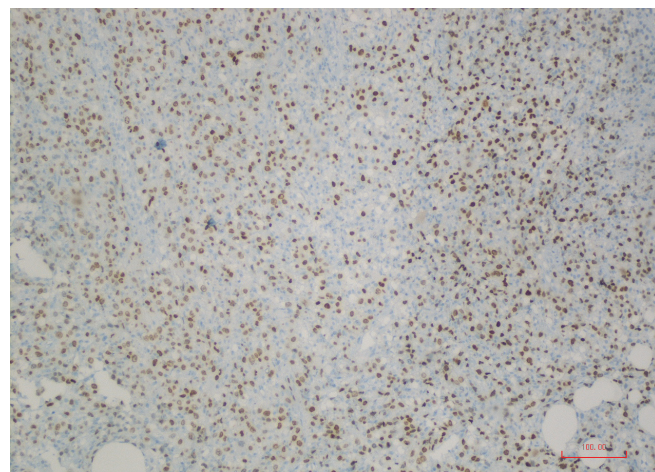


Figure 6. Positive staining with MUM1

lymph nodes in the neck, and in the liver and spleen that matched the hypodense nodules seen on CT. As the lesion was immunohistochemically negative for ALK and EMA, the patient was diagnosed as PCALCL with extracutaneous involvement

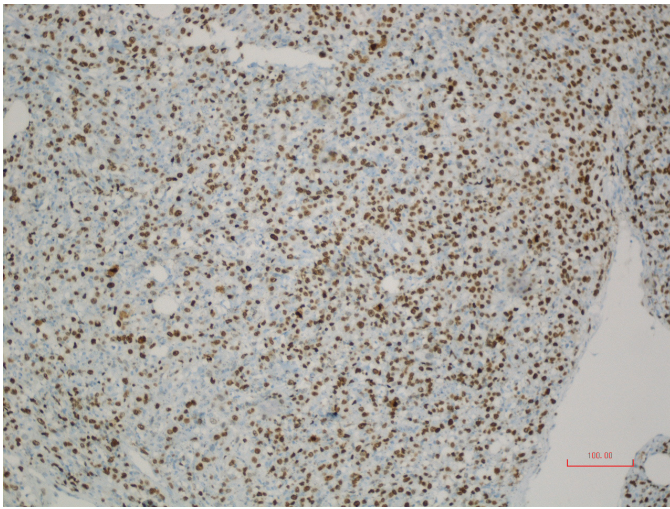


Figure 7. 90% Ki67 proliferation index

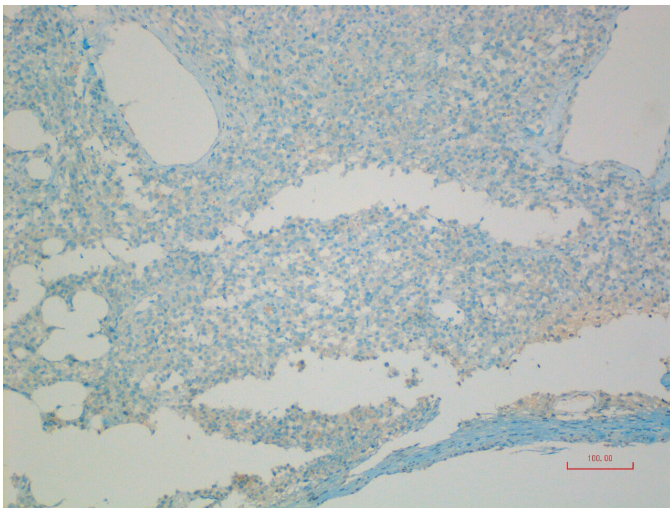


Figure 8. Negative staining with anaplastic lymphoma kinase

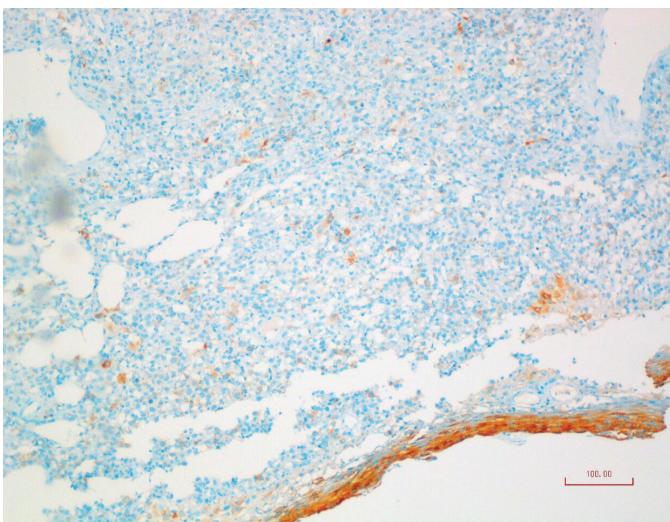


Figure 9. Negative staining with epithelial membrane antigen

DISCUSSION

Primary cutaneous lymphomas are a heterogeneous group of diseases consisting of extranodal non-Hodgkin's lymphomas that present in the skin. Unlike nodal non-Hodgkin lymphomas, 75% of primary cutaneous lymphomas are of T lymphocyte origin and the majority are mycosis fungoides and Sezary syndrome. This is followed by lymphoid papulosis and that present in the skin as CD30 + T cell lymphomas (6). PCALCL accounts for 10% of all cutaneous T cell lymphomas. It is usually seen after the 6th decade and is common in men. The most common site of involvement is the head and extremities. Clinically, it is seen as one or more nodules and tumors that can ulcerate. The etiology is unknown. Unlike systemic ALCL, it has a good prognosis and spontaneous regression can be seen (7). Rarely, extracutaneous dissemination may occur and involves regional lymph nodes and viscera (6). The diagnosis of PCALCL is made by clinical findings, histopathological and immunophenotypical examination and imaging methods (5). In histopathological examination, CD4+ T lymphocytes with CD30 + anaplastic morphology form non-epidermotropic diffuse infiltrate in the dermis. Mitosis rate is high. Ulcerated lesions may be accompanied by inflammatory infiltrates of reactive lymphocytes, histiocytes, eosinophils and neutrophils (6). Immunohistochemical studies are important in distinguishing between primary and secondary lesions that are clinically and histopathologically similar. Unlike systemic CD30 + lymphomas, PCALCL is not stained with EMA and ALK immunostaining (8). Imaging techniques are necessary both for staging the disease and for distinguishing between primary and secondary. In order to diagnose PCALCL, it should be shown that there is no systemic involvement (7). In our case, there were regional lymph nodes, liver and spleen involvement as well as skin involvement at the time of diagnosis. It could not be determined that the involvement of lymph nodes and viscera were before or after the cutaneous lesions, since there was no previous examination. However, the patient was diagnosed as PCALCL with extracutaneous involvement due to lack of B symptoms (fever, night sweats, weight loss) which are suggestive of systemic lymphoma and immunohistochemical studies showing negative ALK and EMA staining. Surgical excision and radiotherapy are the treatment options for localized PCALCL cases. In case of extracutaneous involvement, systemic multiple chemotherapy is applied (9). Recent studies have reported cases of complete remission with brentuximab vedotin, an anti-CD30 monoclonal antibody (10). We did not have the opportunity to initiate any treatment because our patient died one month after the diagnosis.

PCALCL has a better prognosis than systemic lymphomas. Ten-year survival has been reported to be 90% (8). However, the presence of widespread skin lesions, muscle and deep fascia involvement, the occurrence of lesions in the leg and extracutaneous involvement reduces survival (11-13). In the study of Benner and Willemze (3), 14.8% of 135 patients with PCALCL showed extracutaneous invasion. It was reported that 70% of the patients were alive and in remission, 6% were alive and under treatment, 8.9% died due to lymphoma and 14% died due to another cause. In the study of Hapgood et al. (14) 15% of 47 PCALCL cases showed extracutaneous spread. It was reported that 57% of the patients were alive and in remission, 13% were alive and under treatment, 9% died due to lymphoma and 21% died due to another cause. In another study by Benner and Willemze (15), 5-year survival rates of patients with and without lymph node and visceral organ involvement were reported as 51% and 80%, respectively. In conclusion, since treatment and prognosis differ by origin, staging is important in patients with PCALCL and systemic and primary cutaneous lymphoma should be differentiated by clinical findings, imaging methods, histopathological and immunophenotypical examination.

Ethics

Informed Consent: Writing concept of the patient was received.

Peer-review: Externally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: S.A.Ö., Ş.G., Ö.Ö., T.E., Concept: S.A.Ö., Ş.G., E.K., Design: S.A.Ö., Ş.G., Data Collection or Processing: S.A.Ö., Ö.Ö., Ö.Y., Ş.S.Ö., Analysis or Interpretation: S.A.Ö., Ş.G., T.E., E.K., Literature Search: S.A.Ö., Writing: S.A.Ö.

Conflict of Interest: No conflict of interest was declared by the authors.

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