

# Periphreal T-Cell Lymphoma not otherwise specified (PTCL-NOS) Variety in A 58 Year Old Male Patient in Oncology Ward J.P.M.C. Karachi: A Case Report

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## Abstract

**Background:** Peripheral T cell Lymphoma is a heterogeneous form of non-hodgkin lymphoma, and encompasses many sub-types of which Peripheral T cell lymphoma not otherwise specified (PTCL-NOS) is the rare subset.

**Case presentation:** A 58 years old farmer presented with multiple small to medium sized pruritic skin swellings on whole scalp, neck, face and inguinal region with bilateral axillary lymphadenopathy, patient denied any constitutional symptoms or rash. Tissue biopsy reported solid sheets of large atypical pleomorphic cells; Immunohistochemistry and histopathology findings favored the diagnosis of Peripheral T-cell Lymphoma Not Otherwise Specified. CT scan of brain, chest and abdomen excluded visceral involvement.

**Conclusion:** This group of PTCL is aggressive in nature and requires standard combination chemotherapy upon diagnosis, through this report we highlight the variability of Peripheral T Cell Lymphoma's clinical presentation.

**Keywords:** Peripheral T cell Lymphoma; Immunohistochemistry; Mycosis fungoides; Axillary lymphadenopathy

## Introduction

Peripheral T cell lymphoma is a broad terminology to define heterogeneous cutaneous lymphomas which are classified by World Health Organization- European Organization for Research and Treatment of Cancer (WHO-EORTC) based on their morphological, genetic and clinical features, of these Peripheral T-cell Lymphoma Not Otherwise Specified (PTCL-NOS) comprises 2% relative frequency [1]. PTCL-NOS is a diagnosis of exclusion that does not fit in other category of cutaneous T cell lymphoma and includes 3 provisional entities Primary cutaneous aggressive epidermotropic CD8+ T cell lymphoma, Cutaneous  $\gamma/\delta$ + T cell lymphoma and Primary cutaneous CD4+ small/medium sized pleomorphic T cell lymphoma according to WHO-EORTC classification, however Cutaneous  $\gamma/\delta$ + T cell lymphoma has been separated out by WHO Classification 4th Ed [1,2].

## Case Report

A 58 years old male with no known comorbid, farmer by profession presented to oncology outpatient department with 2 months history of progressively growing multiple small to medium sized pruritic nodules on whole scalp, temporal sides of face and upper neck, similar lesions were reported in inguinal regions as well, patient denied any history of trauma, weight loss, constitutional symptoms, rash or plaques that preceded lesions. No family history of similar lesions or autoimmune disease was found. Past medical history was not significant for any past illness. Initially patient consulted to a tertiary care hospital about his illness where all relevant laboratory investigations done and revealed the diagnosis of Peripheral T cell lymphoma not otherwise specified

(PTCL-NOS) but unfortunately patient's previous investigations data related to his illness could not be retrieved due to hospital policy.

On examination vital signs were within normal limits, local exam showed multiple round to ovoid shaped skin colored nodules of different sizes, these nodules were smooth, nontender, firm and freely mobile on overlying skin (Figure 1). Other systemic examination was unremarkable.



**Figure 1:** Note multiple skin colored nodules covering whole scalp, upper neck and sides of face.

We performed a serial of initial laboratory investigations which were found to be normal, after which we proceeded with skin biopsy that documented an intact epidermis. Dermis reveals an infiltrating neoplastic lesion exhibiting solid sheets of large atypical pleomorphic cells with irregular vesicular to hyperchromatic nuclei and conspicuous

nucleoli with scanty cytoplasm. Some mitosis was also seen. But necrosis was not observed. IHC pictures of this patient were not obtained due to some technical issues. However, Tumor was found to be invading the dermis and reaching up to sub cutis. CT scan brain, chest and abdomen were done to exclude any underlying visceral involvement; results were found to be normal except that multiple subcutaneous nodules were noticed in the scalp region.

This patient comes to oncology department of JPMC Karachi on regular follow up. He is being treated with double chemotherapy and showing improvement in symptoms.

### Immunohistochemistry Marker

CKAE1/AE3	Negative
CD45 (LCA)	Diffuse positive in atypical cells
CD3,CD5	Positive in large atypical cells
CD20,D79a	Positive in background small cells

### Discussion

Peripheral T-cells lymphoma (PTCL) constitutes about 20-30% of all the Non-Hodgkin lymphomas in Asia and around 5-10% in Europe and North America, but they are more common among the Eastern population [3]. According to world health organization (WHO), PTCL are further classified into the following subtypes; Peripheral T-cell lymphoma-Not otherwise specified (25.9%), Angioimmunoblastic T-cell lymphoma (18%), systemic anaplastic large cell lymphoma (12.1%) and extra nodal NK/T-cell lymphoma, nasal type (10.4%) [4], among which unspecified PTCL represents the largest subtype.

Peripheral T-cell Lymphoma, not otherwise specified (PTCL-NOS) is defined as an entity that cannot be classified in any other subtypes of PTCL. Peripheral T-cell Lymphoma-NOS is a very rare rapidly progressing (aggressive) tumor which presents in older population with a mean age of 55-60 years, which happens to be the case in our patient.

PTCL-NOS patients most commonly present with generalized lymphadenopathy, extranodal involvement and systemic constitutional symptoms (fever, night sweats and weight loss) [5].

Immunophenotypically there is predominance of CD4 with loss of one of the pan T-cell antigen (CD2, CD3, CD5 or CD7) as seen in 75% of the cases, among which CD7 and CD5 are mostly absent, although in this case CD3 and CD5 were found to be positive in large atypical cells [6,7]. Tumor marker CD79a is commonly expressed in precursor B-cell in the bone marrow; however this case reports a rare phenomenon in which CD79a is found to be positive in the background of small cells in a T-cell lymphoma. In a previous study done by Blakolmer et al. [8], they reported three cases of CD79a positive T-cell lymphomas but all of them were negative for CD20. However this represents a distinctive phenomenon in which both the CD79a and CD20 were positively expressed in a patient with T-cell malignancy [9]. CD45 (LCA) was also reported positive, which is specifically defined for the hematolymphoid nature of the tumor.

This case also represents the cutaneous manifestation of the tumor as the patient presents with progressively growing multiple small to medium sized nodules on the scalp, neck and the inguinal region. The

features presented by these nodules resemble those of mycosis fungoides which is a slow growing lymphoma affecting the skin in the form of patches, and most commonly involves the head and neck region, as presented in this case.

Mycosis Fungoides (MF) is a subtype of Non-Hodgkin cutaneous T-cell lymphoma. It most commonly manifests in males and presents in the sixth and seventh decade of life. In the earlier stages of MF the patient remains stable and the skin lesions are only presented superficially. With further progression of the disease these lesion may progress to the lymph nodes and other organs. MF is often misdiagnosed as other clinical entities such as eczema, dermatitis or fungal infections. CT and MRI scans should be performed to stage and assess the tissue involvement

PTCL-NOS have a very poor prognosis with a 5 year survival rate of approximately 20-45% [7]. Keeping in mind the aggressive nature of this tumor the patient was subjected to immediate chemotherapy, Most PTCL are subjected to CHOP (cyclophosphamide, doxorubicin, vincristine, prednisone) or EPOCH (etoposide, vincristine, doxorubicin, cyclophosphamide, prednisone) based combined chemotherapies. However highdose chemotherapy along with allogenic bone marrow transplant as a primary therapy for PTCL-NOS is still under investigation [4]. The progress of this patient is being monitored through regular follow-ups in the oncology department of the hospital in which the case was first presented.

In summary, PTCL-NOS is a rare malignancy and the diagnosis of such cases can be very challenging. In order to make a clear-cut diagnose of such rare cases patient's history, physical examination, immunohistochemical techniques and imaging findings should be carefully studied and analyzed to provide optimal treatment through combined chemotherapy or allogenic bone marrow transplant, and to bring about a better prognosis in these cases.

### References

1. Willemze R, Jaffe ES, Burg G, Cerroni L, Berti E, et al. (2005) WHO-EORTC classification for cutaneous lymphomas. *Blood* 105: 3768-3785.
2. Vasile D, Vladareanu AM, Bumbea H (2014) Peripheral T and NK cell non-hodgkin lymphoma a challenge for diagnosis. *Maedica (Buchar)* 9: 104-108.
3. Savage KJ, Ferreri AJ, Zinzani PL, Pileri SA (2011) Peripheral T-cell lymphoma--not otherwise specified. *Crit Rev Oncol Hematol* 79: 321-329.
4. Rodriguez-Abreu D, Filho VB, Zucca E (2008) Peripheral T-cell lymphomas, unspecified (or not otherwise specified): a review. *Hematol Oncol* 26: 8-20.
5. Jaffe ES, Nicolae A, Pittaluga S (2013) Peripheral T-cell and NK-cell lymphomas in the WHO classification: pearls and pitfalls. *Mod Pathol* 26 Suppl 1: S71-87.
6. Kung TA, Smith LB, Chung KC (2014) Atypical presentation of isolated peripheral T-cell lymphoma in the hand: case report. *J Hand Surg Am* 39: 732-736.
7. Blakolmer K, Vesely M, Kummer JA, Jurecka W, Mannhalter C, et al. (2000) Immunoreactivity of B-cell markers (CD79a, L26) in rare cases of extranodal cytotoxic peripheral T- (NK/T-) cell lymphomas. *Mod Pathol* 13: 766-772.
8. Yao X, Teruya-Feldstein J, Raffeld M, Sorbara L, Jaffe ES (2001) Peripheral T-cell lymphoma with aberrant expression of CD79a and CD20: a diagnostic pitfall. *Mod Pathol* 14: 105-110.
9. Imran MB, Othman S, Shahid A (2011) Unusual presentation of mycosis fungoides as a lump in the scalp. *Singapore Med J* 52: e226-228.