Non-mycosis Peripheral T-Cell non-Hodgkin Lymphoma involving the Skin

ABSTRACT
A 64 years old male presented with reddish lesions all over the body of 1 month duration, high grade fever with evening rise of temperature and chills. No lymphadenopathy or hepatosplenomegaly were noted. Multiple infiltrated erythematous and hyperpigmented patches and plaques were present on the face, trunk and extremities along with few oral erosions. Histopathology from skin showed features of mycosis fungoides (MF). A further workup with Immunohistochemistry was suggestive of peripheral T-cell lymphoma, not otherwise specified (PTCL-NOS). We report a case of PTCL-NOS in a man mimicking MF clinically and histopathologically.

Keywords: Peripheral T-cell lymphoma (PTCL), Peripheral T-cell lymphoma-not otherwise specified (PTCL-NOS).

INTRODUCTION
Peripheral T-cell lymphomas (PTCL) comprise a rare and heterogeneous subset of T-cell non-Hodgkin lymphomas (NHL) which comprise of a diverse group of disorders that, for the most part, carry a poor prognosis. They arise from lymphocytes at the post-thymic stage of maturation, at nodal or extranodal sites and display T-cell/NK-cell immunophenotype.1 It is distinct from the more common cutaneous TCL.1 Any mature T-cell NHL is considered a PTCL, with the exception of lymphoblastic lymphoma. We report a case of PTCL, not otherwise specified (PTCL-NOS) in a man mimicking mycosis fungoides (MF) clinically and histopathologically.

CASE REPORT
A 64 years old male presented with reddish lesions all over the body of 1 month duration. The lesions began appearing on the abdomen and spread all over the body in a duration of 15 days. Patient gave history of high grade fever, along with evening rise of temperature and chills. In addition, he complained of oral lesions and nasal stuffiness. There was history of joint pain 1 week following development of skin lesions. Patient was a known case of hypertension on treatment with calcium channel blockers.

Clinical examination revealed fever (101°F) along with tachycardia. No lymphadenopathy or hepatosplenomegaly were noted. Lesions were generalized and distributed bilaterally symmetrical on the face, trunk and extremities. Multiple infiltrated erythematous and hyperpigmented patches and plaques were present (Figs 1A and B). Oral cavity showed few erosions on the palate.

Patient's hemogram showed neutrophilia, but the total count was within normal limits. Serum urea, creatinine and liver enzymes were slightly elevated. Erythrocyte sedimentation rate (ESR) was also elevated (60 mm). Histopathology from skin was suggestive of MF (Figs 2 and 3). Immunohistochemistry (IHC) showed predominant periadnexal infiltrate of atypical lymphoid cells marking positive for CD3, CD5, CD45, CD2, CD4, CD7 and TCRβF1 and negative for CD8, CD10, CD20, CD30 and CD23 negative. Mib-1 was 80%. Non-mycosis peripheral T-cell non-Hodgkin lymphoma involving the skin was described and based on the IHC markers, a final diagnosis of PTCL-NOS was made.

Prior to IHC report, patient was treated symptomatically. He did not develop fresh lesions, but continued to have evening rise of temperature. He was discharged against medical advice and succumbed to the disease within one and a half months of admission.

DISCUSSION
Peripheral T-cell lymphoma are uncommon, accounting for only 10 to 20% of all NHLs2 with an incidence of less than one case per 100,000 in the US.3 A large clinicopathologic study4 of PTCL and natural killer/T-cell lymphoma (NKTL) reported that the most common PTCL subtypes are PTCL-NOS (25.9%), angio-immunoblastic T-cell lymphoma (AITL) being the second
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most common (18.5%). Natural killer/T-cell lymphoma represented 10.4% and adult T-cell leukemia/lymphoma (ATLL) 9.6% of the cases. However, studies have shown that PTCL and NKTCL which are common in many other Asian countries are less prevalent in India.5,6

World Health Organization (2008) revised the classification of PTCL (Table 1) through a combination of morphologic, immunophenotypic, genetic, molecular, and clinical features, thus defining many additional subtypes.7 Peripheral T-cell lymphoma—not otherwise specified include all cases not readily classifiable into other specific T-cell entities of the WHO classification and primary presentation in the skin as in this case is uncommon. Peripheral T-cell lymphoma—not otherwise specified typically occurs in adults at the median age of 55 to 60 years, and a higher prevalence is seen in males8,9 which was consistent with our case. Complete clinical examination and an accurate clinical history are critical to particularly rule out (transformed) MF or one of the three subtypes of primary cutaneous PTCL that have been recognized in recent classifications, i.e. primary cutaneous γδ T-cell lymphoma (PCGD-TCL), primary cutaneous CD8+ aggressive epidermotropic cytotoxic T-cell lymphoma (PCAEC-TCL), and primary cutaneous CD4+
small/medium pleomorphic T-cell lymphoma (PCSM-TCL). Primary cutaneous γδ T-cell lymphoma and PCAEC-TCL were ruled out on the basis of IHC markers (TCR-γδ positive and CD8 negative respectively), whereas PCSM-TCL on the basis of clinical features (presents with a solitary plaque or tumor that is generally localized to the head or neck).

Variable clinicopathological characteristics necessitate the use of IHC panels for the diagnosis of PTCLs. Moreover, even the IHC markers show considerable degree of overlap. Hsi et al. recommended a two tier diagnostic approach for IHC panels: Panel 1 consists of IHC (CD3, CD5, CD10, CD20, CD21, CD30, CD45, PAX5), and panel 2 consists of IHC (CD2, CD4, CD7, CD8, CD23, PD-1, CD56, EBER, ALK1, ITA1, TCRγ, TCRβF1) markers.

As a group, PTCL-NOS are aggressive neoplasms and often present with advanced stage. The overall survival rates of PTCL-NOS presenting in the skin are poor and independent of the presence or absence of extracutaneous disease at the time of diagnosis, cell size, or expression of CD4+ or CD8+ phenotype.

In conclusion, we highlight the importance of this rare, aggressive entity with poor prognosis as a differential for the more commoner and indolent MF.

REFERENCES


