CASE REPORT

CD3+, CD56+, CD4−, CD8−, CD20−, CD30− Peripheral T-Cell Non-Hodgkin’s Lymphoma: A Rare Case Report

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Abstract
Cutaneous T-cell lymphoma (CTCL) commonly presents as mycosis fungoides or Sézary syndrome, both having CD4 positivity. A subset of CTCL which lacks CD4 surface marker is classified as cutaneous γ and δ-T-cell lymphoma (CGD-TCL). Because of its rarity and inability to study large number of patients, the impact of immunophenotype on the clinical outcome of primary CTCL in patients is limited. We report a case of primary CGD-TCL in a 71-year-old male because of this rarity and to emphasize its aggressive nature.

Key Words: Epidermotropism, immunohistochemistry, primary cutaneous gamma/delta–T-cell lymphoma

Introduction
Cutaneous T-cell lymphoma (CTCL) is classified as a subtype of non-Hodgkin’s lymphoma. It is a heterogeneous group of T-cell malignancies which are not defined by any of the recognized clinicopathologic subsets. Several subsets based on characteristic clinical, pathologic, and immunophenotypic features have been described. Full staging investigations are required to exclude a systemic nodal/extranodal peripheral T-cell lymphoma, and multiagent chemotherapy is recommended. In our best knowledge, only forty cases of primary cutaneous gamma/delta–T-cell lymphoma (CGD-TCL) have been described so far.

Case Report
A 71-year-old male patient presented with asymptomatic, multiple papulonodular lesions over body for the past 5 months without any constitutional symptom. The initial site affected was the right leg, and then, he developed lesions over the lips, face, abdomen, and trunk.

Physical examination revealed few erythematous tender nodules over the face. There were multiple erythematous tender nodules and plaques, few with eschar over the abdomen, trunk, and extremities [Figure 1a-d]. There was no associated lymphadenopathy. Systemic examination was insignificant.

All the baseline investigations were normal except slightly raised white blood cell count and mean corpuscular hemoglobin. Peripheral blood smear and bone marrow biopsy showed no abnormality.

On skin biopsy, epidermis showed epidermotropic lymphoid cell [Figure 2a]. Dense infiltrate of medium to large lymphoid cell with vesicular nuclei, prominent nucleoli, and moderate cytoplasm was present in the dermis [Figure 2b and c] on hematoxylin and eosin stain.

Immunohistochemistry done twice showed CD2+, CD3+ [Figure 3a and c], CD56+ [Figure 3b], CD4−, CD8−, CD20−, CD30−, and Ki67+ [Figure 3d]; 90% of the cells showed MIB1 positivity.

Computed tomography (CT) scan of the lower lip showed heterogeneously enhancing soft-tissue lesion [Figure 4a]; CT scan of the chest and abdomen showed Multiple heterogeneously enhancing, soft-tissue density (nodular) lesions over the anterior chest wall [Figure 4b], anterior abdominal wall [Figure 4c].
largest 40 mm × 16 mm. Multiple, bilateral axillary lymph nodes, largest 12 mm × 10 mm right, and 14 mm × 8 mm left; few small nodes in bilateral level IA and level IB; and well-defined sclerotic lesions in bilateral iliac bones [Figure 4d].

Bone marrow biopsy showed normocellular marrow (Myeloid : Erythroid ratio=3.5 : 1) uninvolved by primary/secondary malignancy. No evidence of atypical cells or granuloma.

Since immunohistochemistry showed CD2+, CD3+, CD56+, CD4−, CD8−, CD20−, and CD30−, 90% of the cells showed MIB1 positivity, and on the basis of biopsy findings, he was diagnosed as high-grade (CD2+, CD3+, CD4−, CD8−) peripheral T-cell non-Hodgkin’s lymphoma.

Discussion

About 10%–15% of non-Hodgkin’s lymphomas have been diagnosed as peripheral T-cell lymphoma.[1]

The most common presentations of CTCL are mycosis fungoides (MF) or sezary syndrome (SS), both show CD4 surface positivity. A subset of CTCL which lacks CD4 surface marker is classified as CGD-TCL. Occasionally, CGD-TCL can show epidermotropism which is a prominent feature of MF/SS.[2] Clinically aggressive CTCL is classified as below:[3]

1. Sezary syndrome
2. Adult T-cell leukemia/lymphoma
3. Extranodal NK/T-cell lymphoma, nasal type
4. Primary cutaneous peripheral T-cell lymphoma, unspecified
5. Primary cutaneous aggressive epidermotropic CD8+ T-cell lymphoma (provisional)
6. CGD-positive TCL (provisional).
Clinically, CGD-TCL presents with sudden appearance of multiple cutaneous plaques, nodules, and tumors, which have tendency to ulcerate, without any preceding patches or plaques. Although mucosal and extranodal involvement is common, it can involve subcutaneous tissue also.\[2\]

The distinct histological presentation of CGD-TCL has been in literature; epidermotropic, dermal, and subcutaneous and more than one of these three can be present in the same patient within the same biopsy specimen or in different biopsy specimens.\[4,5\] These tumor cells show surface marker for CD2, CD3, and CD56 but negative CD5, CD4, and CD8.

To our best knowledge, only forty cases of primary CGD-TCL have been reported till date.\[6\]

Because of its aggressive nature and resistance to multidrug chemotherapy and radiation therapy, median survival rate of only 15 months has been reported in one study.\[5\]

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**Conflicts of interest**
There are no conflicts of interest.

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**What is new?**
When CTCL is suspected in the patients, biopsy with immunohistochemistry holds a very important place. It helps in the classification of the lymphoma and early identification of the clinically aggressive CTCL. Patient can thus be explained the prognosis and management with multidrug chemotherapy and radiation can be started at the earliest, to prolong the survival.

**References**

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