CORRESPONDENCE

T Cell Prolymphocytic Leukemia (T-PLL): Cutaneous Involvement in an Uncommon Leukemia

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Dear Editor

A 70 years old lady presented with multiple neck swellings and abdominal pain for 6 months. Examination showed pallor with bilateral cervical, axillary and inguinal lymphadenopathy. There was moderate hepatomegaly with no splenomegaly. She also had a macular rash over the trunk and limbs. Chest X-ray was normal. Ultrasonogram of the abdomen showed prominent peri-portal lymphadenopathy.

Investigations showed hemoglobin of 84 g/l, total leukocyte count of $311 \times 10^9/l$ and platelets of $119 \times 10^9/l$. Differential count showed 94% lymphoid cells, 4% neutrophils and 2% metamyelocytes. The lymphoid cells were of size 10–14 μm with indented and clefted nuclear outlines, single prominent nucleolus and scanty to moderate cytoplasm (Fig. 1a). Many of the cells showed cerebriform morphology with cytoplasmic blebs in few.

Bone marrow aspirate was hypercellular with trilineage hematopoietic depression and diffuse replacement by lymphoid cells with clumped chromatin, inconspicuous nucleoli and scant basophilic cytoplasm. Bone marrow differential count showed: 79% lymphoid cells with few residual erythroid and myeloid hematopoietic cells. Trephine biopsy showed a diffuse infiltrate of lymphoid cells (Fig. 1b). Cytochemistry was not done due to non availability of stains. Immunophenotyping showed lymphoid cells to be positive for CD45, CD3 (93.2%), CD5, CD4 and CD7. CD8, B-cell and

myeloid markers were negative. CD52 expression was not studied.

Biochemical investigations were normal. Biopsy from the macular rash showed dense peri-appendageal lymphocytic dermal infiltrate with melanin incontinence and no epidermotropism (Fig. 1c). The infiltrate was positive for CD3 (Fig. 1d). CD20 was negative with moderately high (40%) Ki67 index. A diagnosis of T-cell Prolymphocytic leukaemia (T-PLL) was made. The patient was started on Fludarabine and Cyclophosphamide. After the third cycle of chemotherapy, she developed tumor lysis syndrome with renal failure and an infarct in the left lentiform nucleus. In view of the poor prognosis, the relatives took her against medical advice.

T-PLL is an uncommon chronic lymphoproliferative disorder with male preponderance and comprises 2% of mature lymphocytic leukemias. The median age at presentation is 65 years. Lymphadenopathy and hepatosplenomegaly are common with skin lesions and effusions seen in 20 and 15% of patients respectively [1, 2].

Patients have leukocytosis with one thirds having associated anemia and thrombocytopenia. Liver function tests are frequently abnormal with raised urate and lactate dehydrogenase levels [2]. Peripheral blood smear shows prolymphocytes with classical T-PLL seen in two-thirds of patients with larger lymphoid cells with irregular outline and cytoplasmic blebs. Remaining patients show small lymphoid cells or have cerebriform nuclei [2–4]. Skin lesions are often seen without epidermotropism [5]. Immunophenotyping in T-PLL shows the most frequent phenotype to be CD4+, CD8– (65% cases), CD4+/CD8+ (25% cases) and CD8+/CD4– (10% cases). Positivity for CD2, CD3, CD5 and CD7 is consistently seen. CD52 expression is usually seen and can be used for targeted therapy with Alemtuzumab. Tdt is negative along with pan B cell markers. Cytogenetic abnormalities seen commonly in T-PLL are $t(14;14)(q11;q32)$ and $inv(14)$. The

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Fig. 1 **a** Shows peripheral blood with small sized lymphoid cells having indented and clefted nuclei, single prominent nucleolus in few and scanty cytoplasm (Leishman $\times 1000$). **b** Trephine biopsy of the bone marrow shows replacement by lymphoid cells. **c** Biopsy from

the macular rash showed dense peri-appendageal lymphocytic dermal infiltrate, melanin incontinence with no epidermotropism (H & E $\times 100$). **d** Shows the lymphoid infiltrate to be positive for CD3

resultant TCL1 gene is strongly expressed and can be detected by immunohistochemistry. An association with Ataxia-Telangiectasia has also been reported [2, 4].

The differential diagnosis includes B-PLL, T-ALL and mature T-cell leukaemia/lymphoma. Positivity for CD 7 differentiates T-PLL from leukemias of mature T cell type which lack this marker. Cell morphology, clinical features, skin histology without epidermotropism and HTLV-1 status help to differentiate T-PLL from the leukaemia phase of ATLL and Sezary syndrome.

T-PLL has an aggressive clinical course with a median survival of 6 months from the time of diagnosis. Few patients have an indolent course with acceleration of disease after 2–3 years [4].

Compliance with Ethical Standards

Conflict of interest None.

Human and Animal Rights No animals were used in this study.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the

institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Informed Consent Informed consent was not taken as this is an image based report with no patient intervention specific to the study.

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