is markedly enlarged with a uniform deep-brown cut surface. Malphigian follicles are not identifiable. Microscopic observation reveals a monotonous, neoplastic infiltrate consisting of medium-sized lymphocytes with scant to moderate amount of cytoplasm, round to oval nucleus which is often slightly indented or clearly folded, with dispersed chromatin and inconspicuous nucleoli. Red pulp is expanded, where neoplastic cells infiltrate both cords and sinuses with atrophy of the white pulp. Few, residual Malpighian corpuscles might be visible. In the liver, the neoplastic lymphoid cells involve and distend the sinusoids but the portal triads are usually spared[2]. Bone marrow biopsy is often hypercellular, and neoplastic lymphoid cells show an intrasinusoidal distribution like in liver. As the disease progresses, the tumor cells infiltrate the interstitial spaces and undergo a transformation to blastic morphology. Propensity to assume an intravascular infiltrative pattern is linked to a surface adhesion profile[4]. Lymph nodes are rarely involved at diagnosis[1,2].

On immunophenotyping, these cells show positivity for CD2, CD3, CD7 and γ-δ T cell receptor. NK cell markers like CD56 and CD57 can be positive or negative. These cells are immunonegative to CD4, CD5, and CD8. Not uncommonly, the cells show loss of CD3, CD5 and/or CD7 along with an aberrant reactivity for multiple killer immunoglobulin-like receptors (KIR). Very few cases are positive for CD8. Most of HSTL show an inactive cytotoxic profile (TIA-1+, Granzyme−, Perforin−). TCR expression is conspicuous even by flow cytometry. A minority of "variant" forms, usually seen in women, show α-β TCR expression[1,2,5].

CYTOGENETICS

The commonest cytogenetic alteration seen in HSTL is i(7q). This is regarded as the primary cytogenetic event. The other alterations encountered include ring chromosome 7, trisomy 8 and loss of Y chromosome which are now largely perceived as secondary abnormalities which develop with disease progression. The combination of i(7q) and trisomy 8 is pathognomic for HTSL. A TCR-associated gene signature is used to distinguish between γ-δ and α-β lineages[6].