DISCUSSION

Primary cutaneous lymphomas are defined as a clonal proliferation of lymphocytes in the skin, without extracutaneous involvement till atleast 6 months after presentation. Skin is the second most common extranodal site to be primarily involved by lymphoma. Primary cutaneous lymphomas are a rare entity with an estimated annual incidence of 1/100,000(Smith and Wilson, 2008). Such tumors have a completely different clinical behavior and prognosis from histologically similar systemic lymphomas, which may involve the skin secondarily, thus, require different modality of treatment and pose a major diagnostic challenge. A consensus classification was thus developed in 2005 referred to as "WHO-EORTC Classification of Cutaneous Lymphomas"[1].

65% of all Primary cutaneous Lymphomas are of the T-cell type in which the subtype of cutaneous CD30+ lymphoproliferative disorders includes PC-ALCL, Lymphomatoid papulosis and borderline cases[2]. Most PC-ALCLs show a CD30+, CD4+ immunophenotype. Very few cases (less than 5%) show a cytotoxic CD30+, CD8+ immunophenotype with less than 50 such cases reported worldwide[1,2,3,4]. Cutaneous CD8+ CD30+ lymphoproliferative lesions are difficult to classify and comprise of entities with varied presentations from benign course to indolent lymphomas to clinically aggressive lymphomas. CD8+ PC-ALCL is commonly a disease of the elderly (>60 years), with a male preponderance. CD8+ PC-ALCL has not been described in the paediatric population. The lesion usually is a solitary reddish non-healing nodule or plaque which in most instances, presents with secondary ulceration. The common sites involved are trunk, face, extremities, back and chest. Multicentric lesions are seen in about 20% cases[5]. Very few cases present with hemophagocytic syndrome. In 25-66% cases, there is partial to complete spontaneous resolution. Regional lymphadenopathy though reported, is uncommon in CD8+ PC-ALCL. A history of prior or concurrent Mycosis Fungoides must be ruled out to exclude cases of transformed Mycosis Fungoides. In most cases, the distinction from Lymphomatoid Papulosis type D, which stands out as a major diagnostic differential, is made by ruling out the history of recurrent, self-healing papulo-necrotic lesions[2,4,5].

Histopathological findings in 80% cases comprise of the anaplastic morphology with nodular or diffuse, non-epidermotropic infiltrate in sheets composed of large, anaplastic lymphoid cells. The classical anaplastic large cell may show round, oval, or embryonic nuclei, prominent eosinophilic nucleoli and abundant cytoplasm. In 20% cases, the histomorphology is that of large atypical cells with marked pleomorphism or