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Editorial
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Vellum and Apogee-Primary Cutaneous Acral CD8+ T Cell Lymphoma

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Editorial

Primary cutaneous acral CD8+ T cell lymphoma is an exceptional, indolent lymphoproliferative disorder manifesting gradually evolving papules or nodules confined to acral sites. The contemporary, malignant, aggressive, primary cutaneous acral CD8+ T cell lymphoma demonstrating a characteristic diffuse dermal infiltration of intermediate CD8+ cytotoxic T cells represents with indolent clinical behaviour. Initially designated as indolent CD8+ lymphoid proliferation of the ear or nomenclated as primary cutaneous acral CD8+ lymphoproliferative disorder, the lymphoma exemplifies an infiltration of atypical lymphocytes immune reactive to CD3+ and CD8+, confined to the dermis.

Clinico-pathological concurrence is paramount in order to demarcate primary cutaneous acral CD8+ T cell lymphoma from various CD8+ cutaneous T cell lymphomas as primary cutaneous aggressive epidermotropic CD8+ T cell lymphoma or CD8+ mycosis fungoides. Characteristically, the lymphoma emerges within adults > 50 years with median age of disease emergence at 54 years. A male predominance is observed with male to female proportion of 1.7:1 [1,2].

Typically, a solitary, gradually progressive nodule demonstrates an intense, neoplastic lymphocytic infiltrate confined to the dermis along with sparing of epidermis [1,2]. The lymphoma may incriminate multiple sites. Generally, lesions are confined to dermis of the ear along with nose, hands, elbows and feet. Extra-cutaneous involvement is usually absent of obscure pathophysiology, primary cutaneous acral CD8+ T cell lymphoma is engendered from activated mature T lymphocytes immune reactive to CD3+ and CD8+ [1,2].

Commonly, the lymphoma manifests with a painful, solitary, gradually progressive papule or nodule confined to the ear. Clinical course is unexceptionally indolent and disease alleviation may ensue with cogent, localized therapy or localized surgical excision. However, the disease may persist in an estimated 20% individuals [1,2]. Upon microscopy, the lymphoma manifests an intense, diffuse infiltration of dermis with neoplastic lymphocytes, separated from superimposed stratified squamous epithelium with an attenuated Grenz zone.

Neoplastic infiltrate is composed of intermediate to enlarged, monoclonal, atypical, blast-like lymphocytes incorporated with scanty cytoplasm, irregular nuclear membrane and miniature nucleoli. Signet ring cells may be observed. Foci of angio-destruction or necrosis are absent. Subcutaneous adipose tissue is devoid of peripheral accumulation of neoplastic lymphocytes. Focal epidermotropism and exocytosis of atypical lymphocytes may exceptionally ensue [1,2] (Figure 1&2).

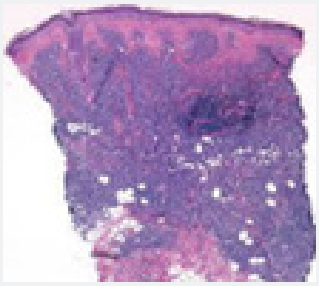

Figure 1: Cutaneous acral CD8+ T cell lymphoma depicting a dense, dermal infiltrate of atypical, intermediate lymphocytes with scant cytoplasm and miniature nucleoli [5].

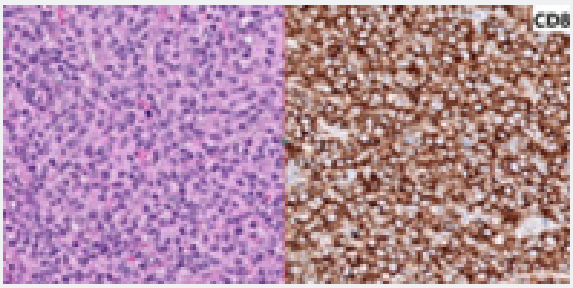

Figure 2: Cutaneous acral CD8+T cell lymphoma delineating an intense, dermal infiltrate of monoclonal, atypical, medium sized lymphocytes which are immune reactive to CD8+ [6].

Primary cutaneous acral CD8+ T cell lymphoma is immune reactive to CD3+ and CD8+. Focal immune reactivity to T cell intracellular antigen 1 (TIA1), granzyme B and T cell receptor BF1 is encountered. Characteristically, immune reactive CD68 expounds a Golgi, dot-like staining configuration. Subsets of atypical lymphocytes may manifest aberrantly decimated T cell antigens as CD2, CD5 or CD7. Ki67 proliferative index is minimal<10% or appears at a moderate 20% to 30%.

Primary cutaneous acral CD8+ T cell lymphoma is immune non reactive to CD4,CD30, CD56, Epstein Barr virus (EBV), granzyme B or perforin. Clonal genetic rearrangements of TCR may be detected comprehensively (100%) with polymerase chain reaction (PCR) [3,4]. Primary cutaneous acral CD8+T cell lymphoma necessitates segregation from neoplasms such as hydroa vaccineforme-like T cell lymphoma, primary cutaneous CD8+ aggressive epidermotropic cytotoxic T cell lymphoma, primary cutaneous gamma delta T cell lymphoma, subcutaneous panniculitis-like T cell lymphoma, mycosis fungoides, cytotoxic or primary cutaneous peripheral T cell lymphoma unspecified [3,4].

Appropriate disease discernment may be obtained with evaluation of precise surgical tissue specimen or excision biopsy. Upon radiographic examination, singular lesion confined to dermis is observed. The lymphoma is devoid of systemic dissemination. Multifocal lesions are exceptionally discerned [3,4].m Surgical extermination of lesion or radiotherapy is an optimal therapeutic option. Generally, precise surgical eradication or radiotherapy alleviates the lesion in ~70% individuals. Prognostic outcomes are excellent and typical instances may not mandate cogent disease staging. Cutaneous disease relapse may occur although neoplastic dissemination into extra-cutaneous sites is exceptional. Tumour relapse is infrequent and confined to dermis. Overall proportionate reoccurrence is minimal at ~20%. Recurring or progressive disease is frequent exemplified within young subjects [3-6].

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5. Image 1 Courtesy: Springer link
6. Image 2 Courtesy: Twitter.com

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