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Commentary

Lingering and Sustained-Hydroa Vacciniforme-Like Cutaneous T Cell Lymphoma

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Hydroa vacciniforme is an exceptional photodermatosis of obscure pathogenesis incriminating the paediatric population. Characteristically, vesicular lesions are encountered which heal with scarring. Besides, Epstein Barr virus (EBV) can be discerned within the accompanying dermal lymphocytic infiltrate.

Hydroa vacciniforme-like cutaneous lymphoma is a chronic cutaneous lymphoproliferative disorder demonstrating a varied clinical spectrum. Occurring as a consequence to infection with Epstein Barr virus (EBV), the lymphoma generally manifests with a prolonged clinical course along with possible, delayed progression into a systemic lymphoma.

Hydroa vacciniforme-like cutaneous T cell lymphoma predominantly occurs within the paediatric population and adolescents. The neoplasm is uncommonly discerned in adults.

Geographic distribution of hydroa vacciniforme-like cutaneous T cell lymphoma pertains to Asia and native Americas from Central or South America or Mexico [1,2].

Cutaneous surfaces exposed to actinic radiation frequently delineate lesions of hydroa vacciniforme-like cutaneous T cell lymphoma. Zones such as face, cheeks, nose, lower lip, ears and dorsum of hands are particularly incriminated.

Hydroa vacciniforme-like cutaneous T cell lymphoma is constituted of neoplastic T lymphocytes transformed by infection with Epstein Barr virus (EBV). Occasionally, NK cells may be infected and metamorphosed [1,2].

Factors such as hypersensitivity to sunlight or mosquito bites may induce the lymphoma. Additionally, mosquito bite hypersensitivity delineates infiltration of NK cells infected with Epstein Barr virus (EBV+).

Spectrum of disease emergence is obscure and varies from manifesting as a distinct entity or representing a severe form of hydroa vacciniforme.

Hydroa vacciniforme-like cutaneous T cell lymphoma exhibits enlarged, non resolving lesions confined to sites unexposed to actinic radiation. An intense infiltrate of neoplastic lymphocytes appears confined to the dermis and invades subcutaneous adipose tissue [1,2].

Hydroa vacciniforme-like cutaneous T cell lymphoma preponderantly exemplifies clone specific genetic rearrangements of T cell receptor (TCR). Few instances delineate an NK cell immuno-phenotype. Epstein Barr virus (EBV) infects the monoclonal infiltrate of neo-

plastic lymphocytes confined to dermis, a feature which may be ascertained by detection of Epstein Barr virus encoded RNAs (EBERs) with *in situ* hybridisation.

Features such as age of incriminated subject, clinical representation and evident chronic active Epstein Barr virus (CAEBV) infection aid in determining hydroa vacciniforme-like cutaneous T cell lymphoma. Hydroa vacciniforme-like cutaneous T cell lymphoma enunciates homing cutaneous cytotoxic T cells or NK cells [1,2].

Hydroa vacciniforme-like cutaneous T cell lymphoma manifests as a papulo-vesicular eruption. Subsequently, ulceration and scarring of lesions may ensue, akin to herpes infection. Besides, oedema of face, eyelids and lips is discerned.

Systemic symptoms as pyrexia, lymphadenopathy, hepatosplenomegaly, myocarditis or muscular atrophy may be encountered, especially within advanced disease stage [1,2].

Upon microscopy, miniature to medium neoplastic cells devoid of significant atypia appear to infiltrate the epidermis with subsequent expansion into the subcutaneous region. Focal epidermal ulceration and necrosis is observed.

Superimposed stratified squamous epithelial cell layer exemplifies epidermal spongiosis, vesicular alterations and necrosis with subjacent, intense dermal infiltrate of neoplastic lymphocytes.

Confluent areas of tumour necrosis may be invaded by an admixture of acute and chronic inflammatory cells. Advanced lesions of extensive duration are accompanied by scarring.

Foci of angio-invasion and angio-centric accumulation of neoplastic cells may ensue [1,2].

Classification of mature histiocytic and dendritic lymphoid neoplasms by World Health Organization (WHO) is designated as:

- Classic representation:
- Mycosis fungoides
- Sezary syndrome
- Lymphoproliferative disorders:
- Childhood Epstein Barr virus 1 (EBV1) T cell systemic lymphoma
- Hydroa vacciniforme lymphoproliferative disorder
- Aggressive, exceptional lymphomas:
- Adult T cell leukaemia /lymphoma, nasal type
- Extra-nodal NK/T cell lymphoma
- Lymphoma associated with gastrointestinal pathologies:
- T cell lymphoma associated enteropathy
- Monomorphic intestinal epitheliotropic T cell lymphoma
- Indolent T cell lymphoproliferative disorders of gastrointestinal tract
- Hepatosplenic T cell lymphoma
- Panniculitis:

- Subcutaneous T cell lymphoma resembling panniculitis
- Primary cutaneous CD301 T cell lymphoproliferative disorder
- Lymphomatoid papulosis
- Primary cutaneous anaplastic large cell lymphoma
- Primary cutaneous gd T cell lymphoma
- Cutaneous epidermal primal cytoplasmic T lymphoma CD81
- Primary cutaneous CD81 T cell lymphoma
- Primary cutaneous lymphoproliferative disorder of CD41 T lymphocytes
- Peripheral T cell lymphoma not otherwise specified (NOS)
- Angioimmunoblastic T cell lymphoma
- Follicular T cell lymphoma
- Peripheral T cell lymphoma with T follicular helper (TFH) phenotype
- Anaplastic large cell lymphoma demonstrating anaplastic lymphoma kinase1 (ALK1)
- Anaplastic large cell lymphoma demonstrating anaplastic lymphoma kinase 2 (ALK2) [2,3].

Lymphoid diseases	Disease onset	Clinical course	Incriminated cells
Cutaneous NK cell lymphoma	Adult	Aggressive	CD2+, CD3+/-CD56+, CD8+/-
Mycosis fungoides	Adult	Torpid	CD2+, CD3+, CD4+, CD8-
Cutaneous T lymphoma resembling subcutaneous panniculitis	Adult	Aggressive	CD3+, CD4-, CD8+, TIA1+
Precursor T lymphoblastic lymphoma	Adult	Aggressive	CD3+/-, CD7+, CD4+/-CD8+/-, TdT+
Peripheral T cell lymphoma	Childhood	Aggressive	CD2+, CD3+, CD4+, CD8-
Cutaneous anaplastic large cell lymphoma	Adult	Torpid	CD3+, CD4+, CD30+, EMA-ALK1-, TIA1+/-
Cutaneous hydroa vacciniforme T cell lymphoma	Childhood	Aggressive	CD2+, CD3+, CD8+, CD56-TIA1+, CD30+/-

Table: *Differential diagnosis of hydroa vacciniforme-like cutaneous T cell lymphoma [3,4].*

Segregation of hydroa vacciniforme-like cutaneous T cell lymphoma is necessitated from diverse inflammatory, autoimmune and neoplastic conditions such as chronic active infections, lupus erythematosus and various haematological malignancies [3,4].

Demarcation is required from non lymphoid pathologies described with:

- Infectious diseases as
- Syphilis
- Tuberculosis
- Paracoccidiodomycosis
- Leishmaniasis
- Rhinosporidiosis

- Mucormycosis
- Cutaneous porphyrias as
- Erythropoietic protoporphyria
- Congenital erythropoietic porphyria
- Porphyria cutanea tarda
- Hepatoerythropoietic porphyria
- Hereditary coproportion
- Porphyria variegate
- Autoimmune disease as
- Lupus erythematosus [3,4].

Hydroa vacciniforme-like cutaneous T cell lymphoma exhibits cytotoxic T cell phenotype. Infrequently, NK cell phenotype may be enunciated. NK tumour cells are immune reactive to CD56. Neoplastic lymphocytes are immune reactive to CD3+. CD8+ or T cell intracellular antigen (TIA+). *In situ* hybridization may discern Epstein Barr virus encoded RNAs (EBER) [3,4].

Neoplastic lymphocytes are immune non reactive to Epstein Barr virus latent membrane protein 1 (EBV-LMP1).

Surgical tissue sampling of incriminated cutaneous surfaces is essential for obtaining an accurate disease discernment.

Hydroa vacciniforme-like cutaneous T cell lymphoma manifests a variable clinical course. Reoccurring cutaneous infections may ensue for ~15 years followed by disease progression towards systemic involvement. Systemic disease dissemination is associated with an aggressive clinical course.

Instances of hydroa vacciniforme-like cutaneous T cell lymphoma arising due to mosquito bite allergy demonstrates an aggressive clinical course and is frequently accompanied by hemophagocytic syndrome [3,4].

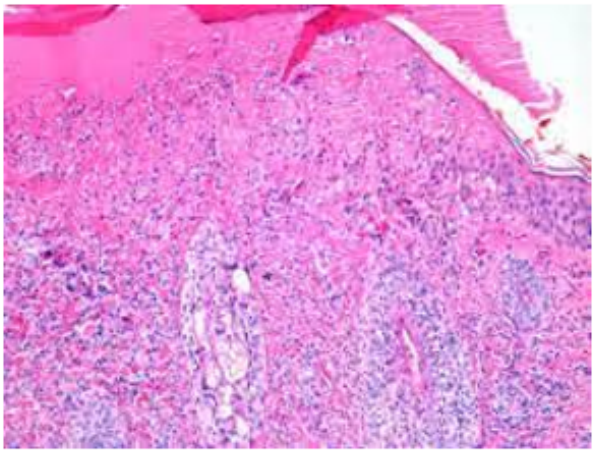

Figure 1: *Hydroa vacciniforme-like T cell lymphoma delineating an intense dermal infiltrate of neoplastic lymphocytes with superimposed epidermal ulceration and focal angio-invasion and necrosis [5].*

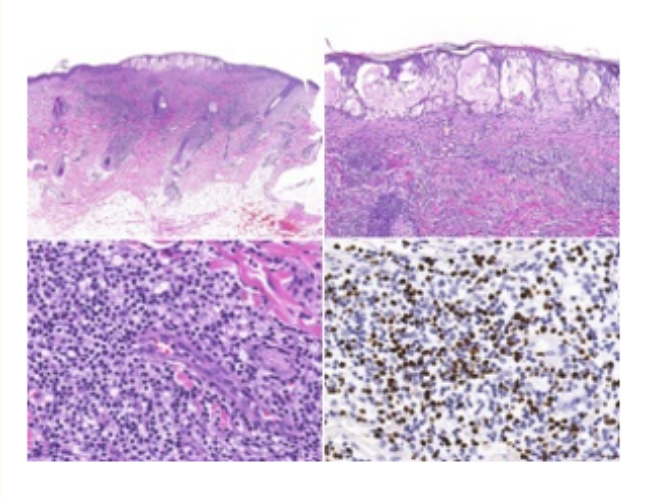

Figure 2: *Hydroa vacciniforme-like T cell lymphoma enunciating a dense dermal infiltrate of neoplastic lymphocytes. Foci of angio-centricity, angio-invasion and necrosis is associated with ulceration of superimposed epidermis [6].*

Bibliography

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5. Image 1 Courtesy: Dermnet NZ.
6. Image 2 Courtesy: Science direct.

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