

Case Report

Hydroa vacciniforme in an adult presenting as bullous pemphigoid-like lesions

成人種痘樣水疱病表現為類天疱瘡樣皮損一例

WK Tang 鄧偉祺

A case of a 41-year-old local Chinese gentleman diagnosed with hydroa vacciniforme was reported. He presented with a twenty years history of relentless, progressive papulo-vesicular skin eruptions. The late onset of this case is atypical. Exclusion of other diseases, especially lymphomatoid papulosis and cutaneous lymphoma, is necessary before reaching the diagnosis. The relatively indolent course of our case makes lymphoproliferative disorders unlikely. Treatment has been unsatisfactory.

報導一例四十一歲華籍男子診斷為種痘樣水疱病。表現為漸進性丘疱疹二十年。此遲發型病例並非此病的典型表現。要診斷種痘樣水疱病，必須先排除其他疾病，特別是淋巴瘤樣丘疹病及皮膚淋巴瘤病。由於此病例病情相對溫和，故此淋巴細胞增生疾病的可能性較少。此例療效不理想。

Keywords: Adult, Chinese, hydroa vacciniforme

關鍵詞：成人，華人，種痘樣水疱病

Introduction

Hydroa vacciniforme is a photosensitive cutaneous eruption that should be considered in a patient who shortly develops erythema and papulo-vesiculation of the skin after sun exposure. The following is a case report of a 41-year-old Chinese gentleman diagnosed to have such condition.

Social Hygiene Service, Department of Health, Hong Kong

WK Tang, MRCP, FHKAM(Medicine)

Correspondence to: Dr. WK Tang

Yung Fung Shee Dermatology Clinic, 4/F, 79 Cha Kwo Ling Road, Kowloon, Hong Kong

Case report

A 41-year-old local Chinese gentleman with good past health presented with generalised papulonodular skin eruptions in 1988. According to him, he developed redness and hotness over his face within an hour after sun exposure since adolescence. Papules, vesicles, sometimes even ulceration, and crusting would appear in the following days. The lesions might heal with scar in about a week later. The skin condition waxed and waned, and deteriorated progressively in the past 20 years. It was worse in the summer time. Initially, only the sun-exposed areas were afflicted but later proximal parts of the limbs and the trunk were also involved. Despite the disturbing nature

of the skin condition, the patient did not have any systemic upset. He had been treated as acne and eczema without improvement by different private practitioners before presenting to our service. There was no significant skin disease in the family.

On examination, diffused papulonodular eruptions with necrosis were found mainly over

the exposed areas. Pock-like scarring and post-inflammatory hyperpigmentation were also evident. However, there were no excessive freckling or lentiginosis. Neither comedones nor photo-onycholysis was found on physical examination (Figures 1 & 2). The differential diagnoses were those of photodermatitis and most likely hydroa vacciniforme.



Figure 1. Papulonodular eruptions over 'V' shape areas of anterior chest wall.



Figure 2. Closer look of the papulonodular eruptions over anterior chest wall.

Blood tests including liver and renal functions, and autoimmune markers were normal. The urine test for porphyrin was negative. Four skin biopsies were done in 1989, 1990, 1996 and 2001 consecutively. The results of the first two biopsies were non-specific. In the latter two biopsies, band-like infiltrate of lymphoid cells, plasma cells and eosinophils with a wedge-shape necrosis of the epidermis and the upper portion of the underlying hair follicle were noted. There were also large lymphoid cells with pleomorphic nuclei. T-cell receptor gene rearrangement showed no clonal proliferation of T-cells. The immunological cell markers: CD30 and CD45RO were positive; and CD56 was negative. Latent Epstein Barr virus (EBV) infection was suggested by the presence of EBV-encoded small nuclear RNA (EBER) positive lymphocytes.

The histological features were compatible with either hydroa vacciniforme or lymphomatoid papulosis. Although CD30 positivity is common in the latter condition, it could also be resulted from non-specific activation by viral infection, like Epstein Barr virus infection. EBV associated lymphoproliferative disorder is a distinct possibility, but the relatively indolent clinical course in our patient made this diagnosis unlikely. Thus the diagnosis of our patient is hydroa vacciniforme. Sun protection measures were suggested to the patients. The skin lesions were treated with topical steroid and antibiotics. A short course of erythromycin, doxycycline and systemic prednisolone had been used to suppress severe flare-up of the disease. Hydroxychloroquine had also been tried to prevent acute exacerbation, but unfortunately, with little success. Moreover, the patient could not tolerate narrow-band UVB, so photo-desensitization was not possible.

Discussion

The mean age of onset of hydroa vacciniforme (HV) is six years old. The disease usually resolves by adolescence or early adulthood. Late onset,¹

persistent cases have been reported up to the age of 58.^{2,3} The disease is characterised by the presence of recurrent vesiculation and vacciniform scarring in sun-exposed area. Seasonal variation may occur; it flares in spring and fades in winter. Photo-onycholysis, conjunctivitis, keratitis may be associated.

HV is primarily diagnosed on clinical basis with compatible histopathologic findings, plus exclusion of other disease mimickers. The result of photo-testing is variable. It could be entirely normal or associated with decreased minimal erythemogenic dose to ultraviolet B (UVB) and ultraviolet A (UVA). Photo-provocation test is usually positive.¹ Histologically, there is intraepidermal vesicle formation, and followed by focal epidermal necrosis, dermal oedema with mixed perivascular infiltrate consisting of mononuclear cells, neutrophils and eosinophils.

HV is not a difficult diagnosis to make when there are typical clinical features. However, there are two diseases that may share very similar clinical features but with less favourable prognosis. Lymphomatoid papulosis may present like hydroa vacciniforme.⁴ So far four such cases have been reported in the English literature. The patients were Japanese, with the disease onset in childhood. The skin lesions in these cases of hydroa vacciniforme-like lymphomatoid papulosis were predominantly found on the face. Two of these patients progressed to lymphoma and subsequently died of it. The nomenclature of hydroa vacciniforme-like lymphomatoid papulosis is still controversial. Some investigators proposed the term EBV associated peripheral T-cell lymphoma; oedematous, scarring vasculitic panniculitis or atypical hydroa vacciniforme. The disease may present like hydroa vacciniforme. Most patients were reported in Asia and Mexico. The condition was found in all age group.^{5,6}

The lesions in EBV associated lymphoproliferative disorder were reported to be larger, and deeper than lesion of classical HV, and were more

disfiguring. Lesion could be present on both exposed and non-exposed area and not exacerbated by sun exposures. Peripheral lymphadenopathy, hepatosplenomegaly, anaemia, lymphocytosis, leucopenia and deranged liver function test had been occasionally found. Systemic symptoms like fever, malaise and wasting were more consistent findings. Like classical hydroa vacciniforme, results of minimal erythema dose for UVA, UVB were variable. Histopathologically, there were atypical lymphocytes in the dermis and subcutaneous tissue. The presence of EBV DNA and EBV-encoded small nuclear RNA were also noted. Angiocentric and periadnexal infiltration of atypical lymphoid cells containing EBER-positive cells were found. Neoplastic cells phenotypes were positive for CD45RO, CD30; but CD56 and CD20 were negative. Clonal T cell receptor- γ gene rearrangement may be present. The prognosis of EBV associated lymphoproliferative disorders was poor. Three of six patients in Cho et al series had progressed to malignant lymphoma and two of them died of it.⁷

Treatment of hydroa vacciniforme is often difficult. There is no curative treatment. Restriction of exposure to ultraviolet radiation such as avoidance of mid-day sunlight, wearing broad brim hat and long sleeves cloths are essential. Prophylactic low-dose broad-band and narrow-band UVB phototherapy or PUVA may be helpful.⁷

Antimalarials drugs have been suggested but the result was variable.⁸ Immuno-suppressive therapy, like systemic steroids and cyclosporine A² may be considered for intermittent control during disease exacerbation.

References

1. Wong SN, Tan SH, Khoo SW. Late-onset hydroa vacciniforme: Two case reports. *Br J Dermatol* 2001; 144:874-7.
2. Blackwell V, McGregor JM, Hawk JL. Hydroa vacciniforme presenting in an adult successfully treated with cyclosporine A. *Clin Exp Dermatol* 1998;23: 73-6.
3. De Pietro U, Simoni R, Barbieri C, Girolomoni G. Hydroa vacciniforme persistent in a 60-year-old man. *Eur J Dermatol* 1999;9:311-2.
4. Tabata N, Aiba S, Ichinohazama R, Kikuchi K, Aoyama H, Watanabe M, et al. Hydroa vacciniforme-like lymphomatoid papulosis in a Japanese child: A new subset. *J Am Acad Dermatol* 1995;32(2 Pt 2):378-81.
5. Cho KH, Kim CW, Heo DS, Lee DS, Choi WW, Rim JH, et al. Epstein-Barr virus-associated peripheral T-cell lymphoma in adults with hydroa vacciniforme-like lesions. *Clin Exp Dermatol* 2001;26:242-7.
6. Cho KH, Kim CW, Lee DY, Sohn SJ, Kim DW, Chung JH. An Epstein-Barr virus-associated lymphoproliferative lesion of the skin presenting as recurrent necrotic papulovesicles of the face. *Br J Dermatol* 1996;134:791-6.
7. Halasz CLG, Leach EE, Walther RR, Poh-Fitzpatrick MB. Hydroa vacciniforme: induction of lesions with ultraviolet A. *J Am Acad Dermatol* 1983;8:171-6.
8. Bickers DR, Demar LK, DeLeo V, Poh-Fitzpatrick MB, Aronberg JM, Harber LC. Hydroa vacciniforme. *Arch Dermatol* 1978;114:1193-6.