Hong Kong J. Dermatol. Venereol. (2012) **20**, 179-182

Case Report

An old gentleman with vegetative plaques and erosions: a case of pemphigus vegetans

一名患有增殖性斑塊皮疹及皮膚潰爛的男性長者：增殖性天疱瘡一例

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A 98-year-old Chinese old gentleman presented with few months' history of vegetative plaques and erosions over the axillae, genitalia and buttocks. The diagnosis of pemphigus vegetans was made. Treatment with oral prednisolone and topical steroid resulted in remission of the disease. Clinical features, histological features and treatment of pemphigus vegetans will be discussed.

一名九十八歲華裔男子，在過去數月內，其腋下、陰部及臀部均出現增殖性斑塊皮疹及皮膚潰爛。其診斷確立爲增殖性天疱瘡。施以口服及外用類固醇後，其病情得以舒緩。本文將討論增殖性天疱瘡的臨床表現、組織學特徵及治療。

Keywords: Pemphigus vegetans, vegetative plaques

關鍵詞：增殖性天疱瘡，增殖性斑塊皮疹

Introduction

Pemphigus vegetans is a rare clinical variant of pemphigus vulgaris. It is characterised by vegetative plaques primarily localised in the flexural areas. The clinical condition usually improves with oral steroid but the disease is noted to have high relapse rate.

Case report

A non-communicable 98-year-old Chinese gentleman was referred to the Social Hygiene Service, Centre for Health Protection, HKSAR for itchy skin rash with blisters and erosions in bilateral axillae and perineal area for few months. Darkening and thickening of the skin of the axillae and groin were also noted. He was found to have recurrent oral ulcer in recent one month before consultation. He had a past history of hypertension and rectal cancer with operation done. He had

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no known drug allergy and there was no recent change of medications. He had no systemic symptoms.

Physical examination revealed hyperkeratotic violaceous plaques in the axillae and groin and multiple superficial erosions on the buttocks. Scarring from erosions on the penile shaft was noted (Figures 1-3). There was no intact blister, oral erosion, inguinal or axillary lymphadenopathy. Complete blood count, liver and renal function tests were unremarkable. The titre of anti-nuclear antibody was 1:80 while anti-double-stranded DNA was negative. Anti-skin antibody (intercellular substance) was >1:640. Bacterial culture of the erosions revealed negative growth.

Differential diagnoses included inflammatory conditions like eczema, seborrhoeic dermatitis, contact dermatitis, inverse psoriasis, postinflammatory hyperpigmentation, acanthosis nigricans, Hailey-Hailey disease, vegetating pyoderma and immunobullous diseases e.g. pemphigus vulgaris, bullous pemphigoid, IgA pemphigus, paraneoplastic pemphigus. Bacterial and fungal infections should also be considered. Incisional skin biopsy was performed on the left axilla of the patient.

Histology showed verrucous epidermal hyperplasia, focal suprabasal intraepidermal abscesses composed of eosinophils, degenerated acantholytic epidermal cells (Figures 4 & 5). Upper dermis showed a mixed inflammatory cell infiltrate. Fungal and Gram stain were negative. Direct immunofluorescence showed deposition of IgG (+++) in the intercellular spaces (Figure 6).

The clinical and histological findings were compatible with pemphigus vegetans. Therefore, the patient was started on oral prednisolone 20 mg daily. After 3 weeks, the erosions healed and there was no recurrence of blisters. A decrease in the thickness of the plaque was noted. Prednisolone was stepped down accordingly.

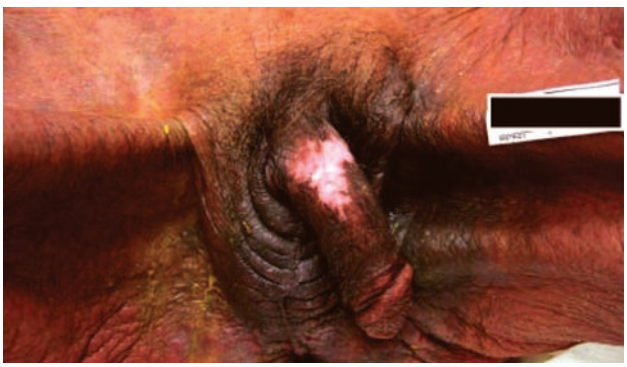

Figure 2. Plaque in the groin and perineum and scarring on the penis from previous erosions.

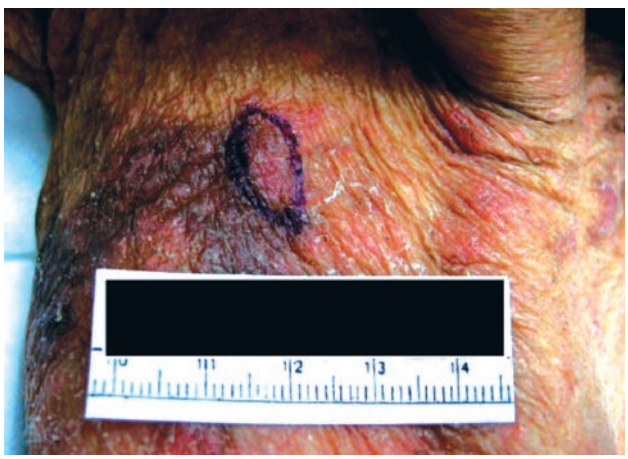

Figure 1. A close-up view of the hyperkeratotic and erythematous lesions in the axilla.

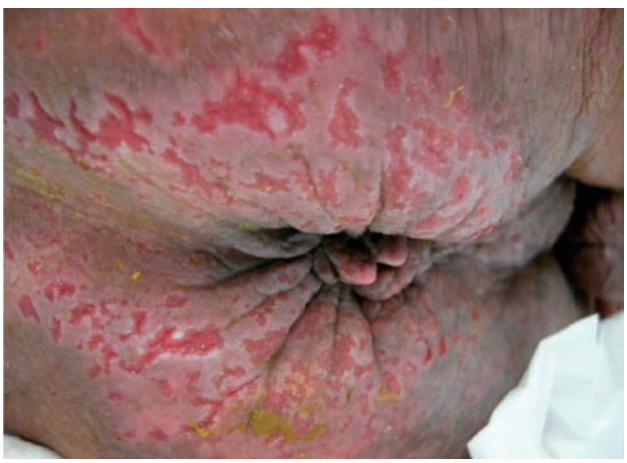

Figure 3. Multiple erosions on the buttocks.

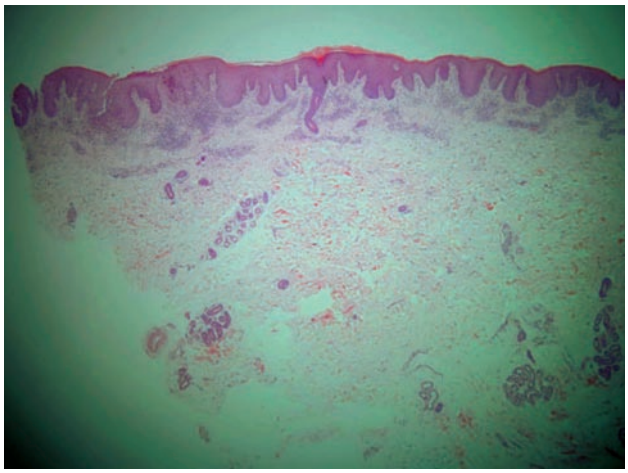

Figure 4. Low power view showing verrucous epidermal hyperplasia, focal suprabasal intraepidermal abscesses.

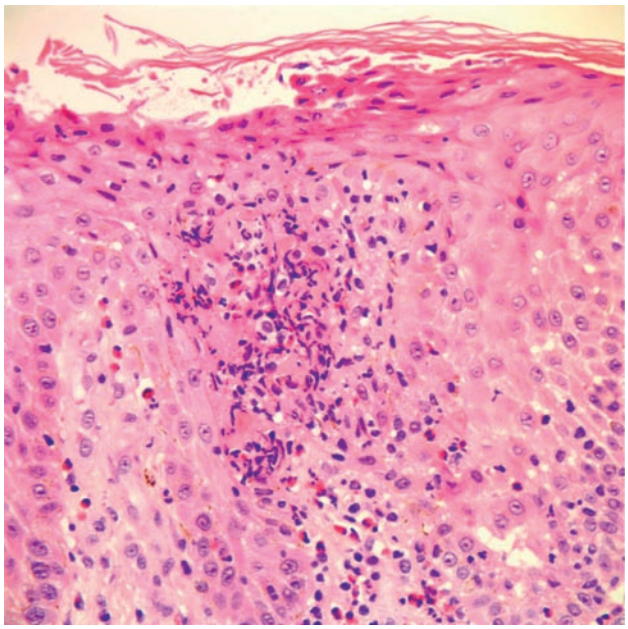

Figure 5. High power view showing intraepidermal abscesses that are composed of eosinophils, degenerated acantholytic epidermal cells.

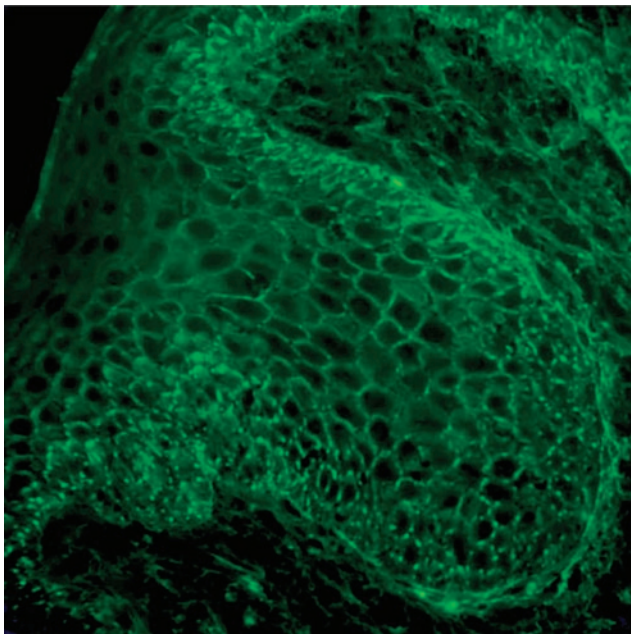

Figure 6. Direct immunofluorescence showing intercellular pattern with IgG (x400).

Discussion

Pemphigus vegetans is a rare intertriginous variant of pemphigus vulgaris characterised by vegetative and purulent lesions present in the groin, axillae, thighs, hands, eyelids and perioral regions. There are few articles in the literature devoted to pemphigus vegetans, and most of them are case reports. It was originally described in 1979 by Winkelmann and Su.[1] A characteristic feature of pemphigus vegetans is the cerebriform tongue, which is characterised by a pattern of sulci and gyri on the dorsum of the tongue. In 60-80% of all cases, the oral mucosa is affected. There are two subtypes of pemphigus vegetans, as follows:

- Neumann type: starts with extensive blisters that erode and develop secondary hypertrophic papillated plaques
- Hallopeau type: begins with grouped polycyclic pustules that erode and evolve gradually to verrucous and papillomatous vegetation

As our patient presented with blisters without pustules followed by development of vegetative plaques, the diagnosis of Neumann type pemphigus vegetans was made. Patients with the Neumann type have a course similar to that of pemphigus vulgaris, need higher doses of systemic corticosteroids and have relapses and remissions.[2] Patients with the Hallopeau type often have a relatively benign disease, require lower doses of systemic corticosteroids and usually have a prolonged remission.

Pemphigus vegetans is caused by intercellular autoantibodies against desmogleins 1 and 3, which are adhesion molecules in desmosomes of keratinocytes.[3] In some case reports, pemphigus vegetans is related to HIV infection, intranasal heroin abuse, drug intake (e.g. captopril), organ transplantation and skin grafting.[4-6] The development of vegetation in the intertriginous areas may be attributed to a response to a super-infection due to the relative occlusion and maceration with subsequent bacterial infection.[7]

Histopathologically, the early lesions of pemphigus vegetans show suprabasal acantholysis. It also exhibits epidermal hyperplasia, papillomatosis and intraepidermal eosinophilic abscesses as the lesions age. It is different from pemphigus vulgaris by an eosinophilic response, formation of microabscess and the extent of vesiculation. Immunofluorescence shows deposits of IgG and/or C3 on the keratinocyte cell surface. Enzyme-linked immunosorbent assay of serum usually reveals autoantibodies against desmoglein 3 in pemphigus vegetans.

There have been no randomised controlled trial in the treatment of pemphigus vegetans. Generally, the treatment is similar to that of pemphigus vulgaris. Systemic corticosteroid is the mainstay in clinical management.[1] Immunosuppressive drugs (azathioprine, cyclosporine, cyclophosphamide, mycophenolate mofetil and methotrexate) serve as steroid-sparing agents and may enhance remission rates.[8] Other alternative therapeutic strategies, including a combination of nicotinamide with tetracycline and dapsone[9] or etretinate/acitretin with systemic steroid are reported to be effective.[10] For resistant pemphigus vegetans, biologicals, extracorporeal photopheresis and carbon dioxide laser are proposed as effective therapy.

In conclusion, pemphigus vegetans is a rare clinical variant of pemphigus vulgaris accounting for 1-2% of all pemphigus. It is characterised by vegetating plaques primarily localised to the flexural areas. Systemic corticosteroid is the mainstay of treatment. Prognosis is unclear due to limited literature on this topic but this condition is observed to have high relapse rate.

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