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Case Report

A lady with recurrent itchy papules: eczematid-like purpura of Doucas and Kapetanakis

一女性患者之復發性癢性丘疹：道 - 凱二氏濕疹樣紫癜

PT Chan 陳寶德, WYM Tang 鄧旭明, WY Lam 林永賢

A 30-year-old lady with recurrent pruritic non-blanchable erythematous papules over the extremities was presented. Skin biopsy confirmed the clinical impression of eczematid-like purpura of Doucas and Kapetanakis. Her skin lesions ran a recurrent course despite oral antihistamines and topical steroids. Phototherapy only produced transient improvement. Topical tacrolimus was given as a therapeutic trial resulted in good control. To the authors' knowledge, this was the first report of the use of topical tacrolimus in the management of pigmented purpuric dermatitis.

病例為 30 歲女性患者，四肢反復出現痕癢性紅色丘疹，壓診不褪色。皮膚組織活檢確診為道 - 凱二氏濕疹樣紫癜。皮損反復發作，口服抗組胺藥及局部激素無效。光照治療僅令病情短暫舒緩。隨後試用局部他克莫司，經兩個月治療後，皮損受到有效控制。就筆者所知，此為首例應用他克莫司治療的色素性紫癜性皮膚病。

Keywords: Eczematid-like purpura of Doucas and Kapetanakis, tacrolimus

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Introduction

Pigmented purpuric dermatosis (PPD) encompasses a group of diseases that are variants of each other resulting from lymphocyte-mediated leakage of erythrocytes. They have similar clinical and histological features and are subdivided mostly for historical purposes. The common clinical feature is the appearance of multiple pinpoint, non-blanchable red or purple papules that

resemble cayenne pepper spots. Clinical types of PPD include Schamberg's disease, purpura annularis telangiectoides of Majocchi, lichenoid purpura of Gougerot and Blum, lichen aureus and eczematid-like purpura of Doucas and Kapetanakis. The following is a report of the last subtype occurring in a young lady.

Case report

A 30-year-old lady developed recurrent itchy erythematous papules over her limbs for eight years. Her lower limbs were more severely affected than the upper limbs. The skin lesions were exacerbated in windy weather and healed with postinflammatory hyperpigmentation. She denied the use of cosmetic and hair dye, but occasionally, she used sunscreen products and nail polish. She had been treated with 1% hydrocortisone cream and cetirizine but the lesions were still recurrent. She had history of thyrotoxicosis in the past that was treated with anti-thyroid drugs. There was no history of atopy. Physical examination revealed multiple erythematous non-blanchable papules, affecting mostly the lower limbs. Postinflammatory hyperpigmentation was present (Figures 1 & 2). Her nails and buccal mucosa were normal. The clinical differential diagnosis included PPD, leucocytoclastic vasculitis, purpuric contact dermatitis, and lichen planus. Thrombocytopenic purpura and Waldenstrom's macroglobulinaemia could also give rise to purpuric lesions, but usually, they were not itchy.

Blood tests including complete blood picture, immunoglobulin pattern, plasma protein electrophoresis, liver and renal function tests were normal. Her antinuclear factor titre was 1:80 and anti-extractable nuclear antibody was negative. Patch testing with European standard series was negative. She was given various topical steroid and oral antihistamine but the response was unsatisfactory. Skin biopsy of a lesion was performed. The histopathology showed a superficial perivascular infiltrate of lymphocytes and extravasation of erythrocytes. Siderophages and eosinophils were focally seen. There were also focal exocytosis, spongiosis and overlying parakeratotic mount. The epidermis was mildly atrophic and papillary dermis was sclerotic. There

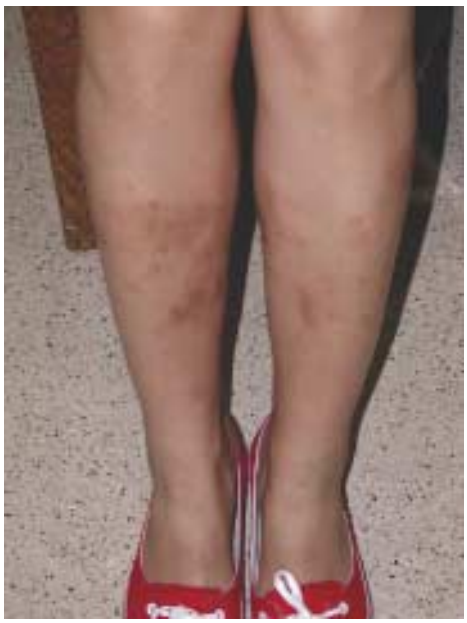

Figure 1. Erythematous lesions with postinflammatory hyperpigmentation over both shins.

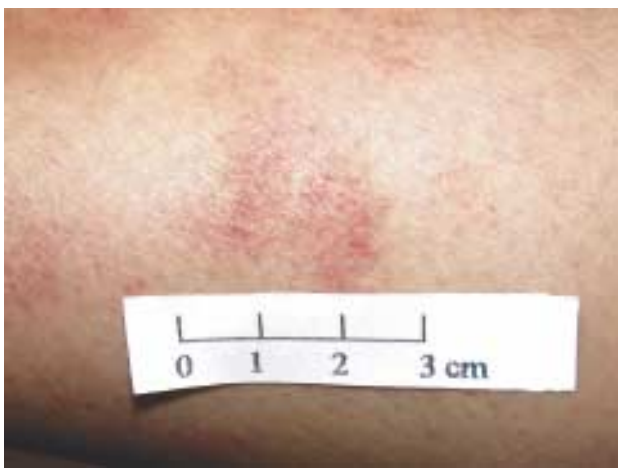

Figure 2. Close up skin lesions showing the erythematous papules. They are non-blanchable.

was no amyloid, fungi nor vasculitis. The histopathologic features were consistant with PPD, Doucas and Kapetanakis type (Figure 3).

She was then treated with two courses of narrow band ultraviolet B. Although remission could be attained, it only lasted three to four months. She was then given a therapeutic trial of topical 0.03% tacrolimus ointment for two months and her skin lesions slowly improved and remained in good control.

Discussion

Eczematid-like purpura of Doucas and Kapetanakis was first described in 1953. It was also called itchy purpura of Loewenthal or disseminated pruriginous angiodermatitis.[1] It differs from other PPD in that pruritus is common. Although different types of PPD have minor clinical differences, they are considered as different manifestations of the same disease process.

The pathogenesis of PPD is largely unknown, but physical factors, drugs and immune mechanism have been postulated to be involved. Aneurysmal dilatation of terminal capillaries have been shown by means of capillary microscopy and it is postulated that rupture of the capillaries results in purpura.[2] Besides, perforator vein incompetence has been reported to be present in cases of lichen aureus and it is postulated that the resultant rise in venous pressure may be important in purpura formation.[3] Drugs that can precipitate PPD include carbromal, meprobamate, diuretics, ampicillin, non-steroidal anti-inflammatory drugs, acetaminophen, glipizide, medroxyprogesterone acetate and thiamine. Recently, infliximab was implicated in a patient who received the infusion for treating her psoriasis.[4] These drugs may act as haptens in stimulating the immune response. Although immunoglobulin and complement are present in blood vessels, frank vasculitic changes are absent. The importance of cellular immunity is suggested by immunohistochemical microscopic study.[5] The cellular infiltrates in Schamberg's disease are predominantly CD3 and CD4 positive lymphocytes and CD1a positive dendritic cells. These two groups of cells often form clusters. A few CD36 positive dendritic cells are scattered around the blood vessels in papillary dermis. A similar picture can be seen in skin diseases that are presumed to occur through cellular mediated immunity, such as lichen planus, graft versus host disease and contact dermatitis. Moreover, upregulation of intercellular adhesion molecule-1 in dendritic cells, keratinocytes and endothelial cells and lymphocyte function associated antigen-1 in infiltrating cells are present. These may be important in regulating the traffic of inflammatory cells to skin.

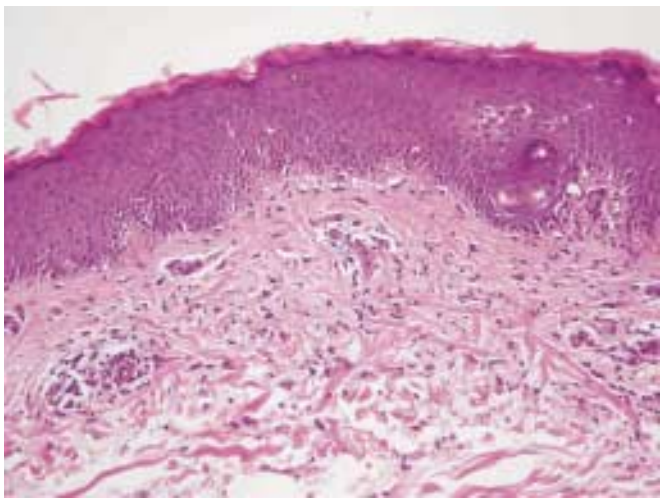

Figure 3. Histology slide that shows superficial perivascular lymphocytic infiltrate, focal spongiosis and small parakeratotic mount. (By courtesy of WY Lam and KC Lee, Department of Pathology, Tuen Mun Hospital and Queen Elizabeth Hospital respectively)

Common to all PPD is infiltrate of lymphocytes around blood vessels, extravasated erythrocytes and siderophages. Spongiosis is more pronounced in eczematid-like purpura. Whereas in lichenoid purpura of Gougerot and Blum and in lichen aureus, there are denser band like infiltrates of lymphocytes. In Schamberg's disease and purpura annularis telangiectoides of Majocchi, lymphocytes are sparse and spongiosis and parakeratosis are minimal.

Eczematid-like purpura of Doucas and Kapetanakis is not common. In a retrospective review of 174 cases of PPD, it only accounted for 10% of cases.[6] There was a slight female predominance, with a mean age of 54.2 years. Most PPD had their eruption localised to the lower limbs (57.4%). Among the different subtypes, eczematid like purpura was more frequently associated with generalised lesions than the Schamberg's subtype. Moreover, moderate to severe pruritus was present in eczematid-like purpura by definition. Precipitating factors were often unknown. Follow up data was available in 87 patients, with a mean duration of 77.5 months. Fifty-eight patients have improvement or complete clearance of skin lesions.

Although PPD is a benign condition, patient may be disstressed by the cosmetic or pruritic problems. The disease may have a course of several years. Suggested treatment is based on case series rather than double blind randomised controlled trial. Reported modalities include topical steroid, oral cyclosporin, griseofulvin, photochemotherapy and oral bioflavonoids and ascorbic acid. Oral griseofulvin has been tried only in a small series of six patients, one patient, however, developed phototoxic eruption. It is used in PPD treatment as it has immunomodulatory action.[7] Oral cyclosporin is not recommended as first line treatment because of potential side effects.[8] Oral bioflavonoids reduce capillary fragility and it, together with ascorbic acid, can act as free radical scavengers. The drug combination has been used successfully in three patients with PPD.[9] Photochemotherapy, with immunosuppressive effects, has been used successfully in seven patients after seven to 20 sessions of treatment.[10] Topical tacrolimus is an immunomodulator approved for used in atopic dermatitis. Anecdotal reports of its use in contact dermatitis, vitiligo, lichen sclerosis et atrophicus are also present. To the authors' knowledge, this is the first report of its use in PPD. Further clinical trials or experience are needed before one can recommend the routine use of this novel treatment in PPD.

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