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Original Article

Pigmented purpuric dermatosis in Singapore: a clinic-epidemiological characterisation

新加坡的色素性紫癜性皮膚病之臨床及流行病學特徵

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Background: Pigmented purpuric dermatoses (PPD) are a group of chronic and relapsing disorders characterised by petechiae and pigmented macules. To date, very few studies have evaluated the clinico-epidemiology of this disorder. ***Aim:*** To describe the local epidemiology of patients with PPD in a tertiary dermatological unit in the National University Hospital, Singapore over a 5-year period. ***Methods:*** A retrospective analysis of all biopsy-confirmed PPD cases diagnosed and treated in NUH in the period 2009-2014, was performed. Patient demographics, clinical characteristics, comorbidities and treatment outcomes were noted. ***Results:*** A total of 48 patients were identified. PPD tends to affect older adults and has a slight female preponderance in our population. The lower limbs were the most common site of involvement. There were no significant comorbidities identified in association with our patients with PPD. The rates of improvement were similar for treatment and non-treatment groups. ***Limitations:*** Retrospective review, small sample size, selection bias from inclusion of only biopsy-proven cases. ***Conclusion:*** Most patients were asymptomatic and those who experience symptoms may benefit from topical corticosteroid therapy. As spontaneous improvement is often observed, treatment is not always necessary.

背景：色素性紫癜性皮膚病是一組特點爲點狀色素沉著的慢性和復發性皮膚疾病。迄今爲止，很少研究評估此病的臨床及流行病學。**目的：**回顧新加坡國立大學醫院內的一所三級皮膚病單位五年內所診治的色素性紫癜性皮膚病患者，來描述此病的當地流行病學。**方法：**對二零零九年至二零一四年間在國立大學醫院診治的所有活檢證實的色素性紫癜性皮膚病個案進行了回顧性分析，並一一紀錄病人的人口統計資料、臨床特徵、合併症和治療結果。**結果：**本研究共篩選出四十八名色素性紫癜性皮膚病患者，發病多見於老年，而女性較男性稍多；下肢爲最常的發病部位。在我們的患者當中，沒有發現顯著的相關聯合併症，而治療組和非治療組的改善率結果分別不大。

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限制：回顧性調查、樣本數較小、僅列入活檢證實個案所存在的選擇偏差。**結論**：多數患者毫無症狀，其餘受症狀困擾的患者在外用皮質類固醇治療後情況多有改善。由於察覺到不少患者可不藥而癒，固非所有患者都要施以藥物治療。

Keywords: Corticosteroids, pigmented purpuric dermatosis (PPD), topical corticosteroids

關鍵詞：皮質類固醇、色素性紫癜性皮膚病、外用皮質類固醇

Introduction

Pigmented purpuric dermatoses (PPD) are a group of vascular disorders of unknown origin that are histopathologically similar. They are chronic and relapsing disorders characterised by petechiae and pigmented macules (Figure 1). The various clinical entities include Schamberg's disease, pigmented purpuric lichenoid dermatosis of Gougerot and Blum, lichen aureus, eczematid-like purpura of Doucas and Kapetanakis, and Majocchi's disease.[1] Although the pathogenesis of PPD is unknown, cell-mediated immunity is thought to be contributory.[2] Extravasation of erythrocytes in the skin and marked haemosiderin deposition are their classical clinical expressions.

PPD has been documented in all ages and races,[1] and appears to occur more frequently in males. Although the exact aetiology is unknown, important contributory factors are believed to include venous hypertension, exercise, gravitational dependency, capillary fragility and focal infections.[1] To date very few studies have evaluated the clinico-epidemiology of this disorder.

Materials and methods

This was a retrospective review of patients diagnosed with PPD from January 2009 to January 2014 in a tertiary hospital, National University Hospital (Singapore). All patients diagnosed clinically or histopathologically with PPD were included. Patients with morphologically similar conditions such as stasis dermatitis, Henoch-Schonlein purpura, leukocytoclastic vasculitis, hyperglobulinaemic purpura, drug hypersensitivity reactions, and factitious injuries were excluded.

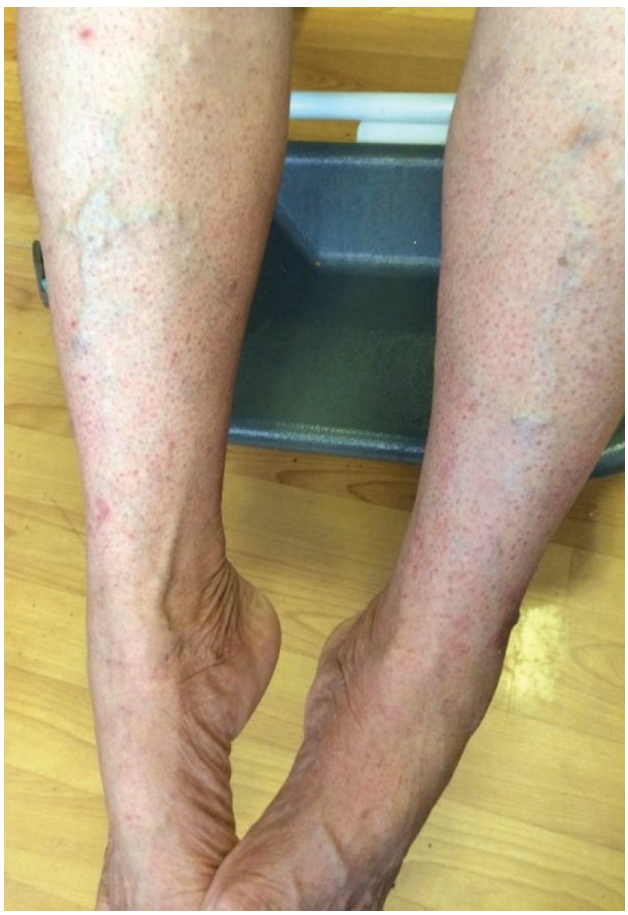

Figure 1. Pigmented purpuric dermatosis. Petechiae and brownish patches over bilateral lower limbs.

Data on patient demographics, clinical characteristics, comorbidities and treatment outcomes were noted. Clinical improvement was defined as 50-100% lesion clearance as determined by the dermatologist during follow up review. The data collection was obtained through chart and note reviews, and completed via telephone interviews. Approval for the study was obtained from the national ethics committee.

Results

Patient demographics

A total of 48 patients were included in the study. There were 22 males (45.8%) and 26 females (54.2%). The mean age of patients at presentation was 54.6 years (range 11-87 years). Of these, the majority were adults (95.8%), with only 4.2% under the age of 16. The main ethnic groups were represented, with 83.3% Chinese, 8.30% Malay, 4.20% Indian and 4.20% classified as "Others".

Clinical characteristics

In terms of symptoms experienced, the majority of patients (75.0%) reported no symptoms, whereas 11 (22.9%) reported pruritus and only 1 (2%) reported tenderness. The lower limbs were the most common site of involvement. The majority of patients (88.0%) reported isolated lower limb involvement, 4% reported lower and upper limbs involvement, 2% reported isolated upper limb involvement and other areas constituted the remaining 6%.

The co-morbidities present were hypertension (37.5%), hyperlipidaemia (25.0%), diabetes mellitus (14.6%), chronic kidney disease (12.5%), osteoarthritis (10.4%), atrial fibrillation (6.3%), benign prostate hyperplasia (6.3%), transient ischaemic attack or stroke (6.3%), varicose veins (6.3%), ischaemic heart disease (4.2%), osteoporosis or osteopenia (4.2%), Still's disease (2.08%), hypothyroidism (2.1%) and rheumatoid arthritis (2.1%).

Treatment

In terms of treatment, approximately half the patients (45.9%) received treatment for their cutaneous lesions. Most of the patients (95.5%) received high potency corticosteroids (World Health Organisation, Group III to V corticosteroids), with the majority (71.4%) reporting improvement at follow up within a year. One patient (2.1%) received treatment comprised of a combination of phototherapy and topical emollients. She received narrow band UVB (NB-UVB) three times weekly, for 21 sessions, with almost complete resolution of lesions. During follow up at one year, there was no recurrence of new lesions. The remainder of patients (52.1%) did not receive any treatment. Of the untreated group, 68% experienced spontaneous resolution of lesions within a year. The demographics and clinical characteristics are shown in Table 1.

Discussion

Pigmented purpuric dermatosis may occur in all age groups and races as observed in our study. It tends to affect older adults, with 60.4% of patients aged 60 and above. There was a slight female preponderance, with a male to female ratio of 1:1.18. The lower limbs were most commonly affected and the large majority of cases were asymptomatic, in line with the literature.[1,3]

Many co-morbidities including hepatic disease, malignancies, diabetes mellitus, hyperlipidaemia, rheumatoid arthritis, lupus erythematous, thyroid dysfunction, hereditary spherocytosis, hematological disorders, and porphyrias have been reported to be associated with PPD.[3] Despite so, our review did not clearly identify any clear co-relation between an individual's co-morbidities and subsequent development of PPD. In addition, although a gravitational role has been proposed to explain the observation of PPD, only 6.3% of our patients were observed to have features consistent with venous insufficiency. This

Table 1. Demographic and clinical characteristics of patients with PPD

Demographics	
Sex, no (%)	
• Male	22 (45.8%)
• Female	26 (54.2%)
Mean age, years	54.6 (11-87)
Clinical characteristics	
Symptoms, no (%)	
• Asymptomatic	36 (75%)
• Itch	11(23%)
• Tenderness	1 (2%)
Sites of involvement, no (%)	
• Lower limbs only	42 (87.5%)
• Lower limbs and upper limbs	2 (4.2%)
• Upper limbs	1 (2.1%)
• Others	3 (6.3%)
Treatment	
Treatment: number (%)	
• None	26 (54.2%)
• Topical corticosteroids	21 (43.8%)
• Phototherapy, emollients	1 (2.08%)
Clinical response	
Clinical improvement: number (%), average duration	
• No treatment (spontaneous improvement)	17 out of 26 (65.4%), 79 weeks
• Topical corticosteroids	15 out of 21 (71.4%), 26 weeks
• Phototherapy, topical emollients	1 out of 1 (100.0%), 7 weeks
Comorbidities, no (%)	
• Hypertension	18 (37.5%)
• Hyperlipidaemia	12 (25.0%)
• Diabetes mellitus	7 (14.6%)
• Chronic kidney disease	6 (12.5%)
• Osteoarthritis	5 (10.4%)
• Atrial fibrillation	3 (6.3%)
• Benign prostate hyperplasia	3 (6.3%)
• Transient ischaemic attack/stroke	3 (6.3%)
• Venous insufficiency	3 (6.3%)
• Ischaemic heart disease	2 (4.2%)
• Obstructive sleep apnoea	2 (4.2%)
• Osteoporosis/osteopenia	2 (4.2%)
• Adult onset Still's disease	1 (2.1%)
• Hypothyroidism	1 (2.1%)
• Rheumatoid arthritis	1 (2.1%)

reinforces the notion that this phenomenon is most likely idiopathic.

First-line treatments of PPD include conservative non-pharmacological treatment and topical corticosteroids. Topical corticosteroid therapy was the most common therapy used within our treatment group, with the majority (71.4%) of patients achieving clinical improvement. They have been used for PPD based on the histological evidence that it is an inflammatory process. The current recommendation for duration of therapy is 4-6 weeks.[1] Phototherapy is considered a well-tolerated second-line intervention, in particular for patients who have failed corticosteroid therapy. Fathy et al. described a prospective uncontrolled study of six patients with PPD, who received treatment with narrowband UVB three times a week for 24 to 28 treatments sessions with good response.[4] The patients who received phototherapy in our review also experienced significant improvement with no recurrence. A variety of other therapeutic options have been reported in the literature with varying success including griseofulvin,[5] pentoxifylline,[6] colchicine,[7] methotrexate,[8] and cyclosporin.[1, 9]

This study is limited by the number of cases, bias from selection of biopsy-proven cases only, as well as recall bias. Large-scale community-based prospective studies are needed with a longer follow-up to ascertain the prevalence, aetiology, aggravating factors, associations and prognostic factors.

In conclusion, PPD in our population tends to affect older adults with a slight female preponderance. It most commonly affects the lower limbs and is rarely symptomatic. Although patients may benefit from topical corticosteroids, PPD often resolves spontaneously without treatment. Hence, unless patients experience symptoms or there is concern over the cosmetic appearance, treatment is not always necessary and reassurance is an acceptable course of management for most cases.

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