Pigmented purpuric dermatosis or mycosis fungoides: A diagnostic dilemma

Najeeba Riyaz, Sarita Sasidharanpillai, Ettappurath N. Abdul Latheef, Hena Davul, Febin Ashraf

Department of Dermatology and Venereology, Government Medical College, Kozhikode, Kerala, India

ABSTRACT

Pigmented purpuric dermatoses (PPD), a group of vascular disorders with variable clinical picture is reported in all races and age groups with a male predilection. There are reports of mycosis fungoides manifesting as pigmented purpura as well as progression of PPD to cutaneous T-cell lymphoma. The diagnostic dilemma is compounded by PPD manifesting histological similarity to mycosis fungoides. Currently, it is believed that PPD with monoclonal T-cell population is more likely to progress to malignancy. We report a 31-year-old male patient who presented with the lichenoid clinical variant of PPD lesions that mimicked mycosis fungoides on histopathology. Gene rearrangement studies identified a polyclonal T-cell population. The patient responded to photochemotherapy, which is beneficial in both PPD and mycosis fungoides. Our case signifies the limitations of current diagnostic modalities in accurately distinguishing PPD from cutaneous lymphoma. Data on disease progression in similar cases may enable us to formulate better diagnostic definitions.

Key words: Mycosis fungoides, pigmented purpuric dermatosis, T cell clonality

INTRODUCTION

Pigmented purpuric dermatoses (PPD) are capillaritides of varying morphology and unknown etiology.[1] With reports of PPD evolving into mycosis fungoides over the years it is recommended that atypical PPD, especially the widespread forms should be evaluated for T-cell clonality with close monitoring of the monoclonal variants.[2,3] Here we report a 31-year-old male patient who manifested PPD confined to legs, which had features of mycosis fungoides on histological analysis.

CASE REPORT

A 31-year-old male patient attended our outpatient department with asymptomatic brownish macules and patches [Figure 1a] and purpuric lesions distributed on both legs near the lateral malleoli of five months duration. A few similar lesions were present on the anterior aspects of both legs [Figure 1b]. According to the patient, lesions started as red spots and later changed in color as new lesions continued to appear. He neither gave any history suggestive of bleeding disorder or venous insufficiency nor was he on any medications. Doppler study of arterial system was within normal limits, whereas venous Doppler revealed incompetence of above-ankle perforators bilaterally. Complete hemogram, peripheral smear study, urine routine examination, bleeding and clotting time, prothrombin time and international normalized ratio were within normal limits. With the clinical diagnosis of pigmented purpuric lichenoid dermatosis, the brownish macule was biopsied, which revealed a heavy lymphoid infiltrate in the superficial and mid-dermis with epidermotropism and occasional Pautrier microabscesses [Figure 2a and b] indicating mycosis fungoides. Perls Prussian blue stain revealed hemosiderin deposits in dermis indicating extravasated red blood cells [Figure 3]. Immunohistochemistry showed the lymphocytes to be CD3 [Figure 4a], CD4 [Figure 4b], and CD5 positive with few CD8 positive cells [Figure 4c].

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Address for correspondence:
Dr. Sarita Sasidharanpillai, 'Rohini', Girish Nagar, Nallalom PO, Kozhikode - 673 027, Kerala, India.
E-mail: saritasclt@gmail.com

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The patient was evaluated in detail in consultation with the hematologist. Serum calcium, liver and renal function tests, chest X-ray, bone marrow analysis, ultrasound examination of abdomen and pelvis, and contrast-enhanced tomogram of the thorax were all within normal limits with marginal elevation of serum lactate dehydrogenase. Gene rearrangement studies identified polyclonal T-cell population.

Since new lesions continued to appear despite treatment with topical steroids, he was offered systemic psoralen 20 mg followed by ultraviolet A therapy twice a week, which after 4 weeks achieved some clearance of lesions and cessation of appearance of new lesions. The patient is presently under regular follow up with thorough clinical evaluation and complete hemogram and peripheral smear analysis every three months.

DISCUSSION

Pigmented purpuric lichenoid dermatosis, a variant of PPD has a male predilection and manifests in adult life.[1] The most common site is the lower limb as noted in our patient; rarely it may be generalized when it becomes difficult to distinguish from lymphoma. This assumes added significance as there is increasing evidence of link between PPD group of disorders and lymphoma.

The proposed causes for PPD include systemic diseases such as rheumatoid arthritis, liver disease, lupus erythematosus, lymphoid malignancy, and hyperlipidemia and certain drugs (nonsteroidal anti-inflammatory drugs, lipid-lowering medications, calcium channel blockers, ACE inhibitors, β-blockers, antihistamines, and antidepressants). Venous insufficiency (as in our case) and exercise are also known to precipitate PPD.[2]

Recent data suggest that pigmented purpuric eruption could be an early manifestation of mycosis fungoides or over the years PPD may progress to cutaneous T-cell lymphoma or extremely rarely the two conditions may co-exist.[3] Many authorities support the view that PPD represents a form of T-cell dyscrasia within the spectrum of cutaneous T-cell lymphoma.[2] Considerable overlap is observed between the features of cutaneous T-cell lymphoma and PPD.

The clinical features suggestive of the pigmented purpuric variant of cutaneous lymphoma are widespread involvement by way of purpuric patches and pruritus lasting longer than one year. It has been recommended that atypical pigmented purpuric lesions should be evaluated with serial biopsies

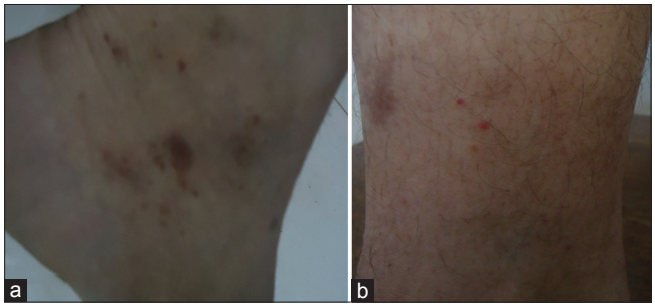

Figure 1: (a) Brownish macules and patches of pigmented purpuric dermatosis near the lateral malleolus. (b) Purpuric lesions on the shins of the same patient along with brownish patches

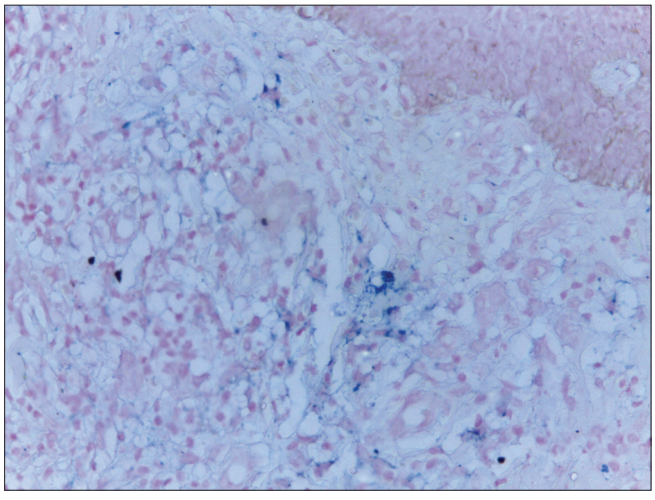

Figure 3: Skin biopsy showing hemosiderin deposits in the dermis (Perls Prussian blue, ×400)

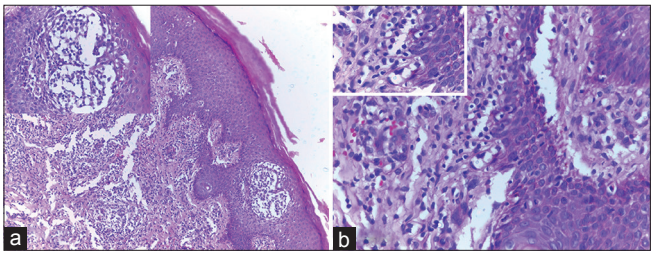

Figure 2: (a) Acanthosis, moderately dense dermal infiltrate and Pautrier micro-abscess (H and E, ×100), inset: High-power view of the Pautrier micro-abscess (H and E, ×400). (b) Atypical lymphocytes arranged in string of pearl appearance along the basal layer (H and E, ×400), inset: Showing dark small to medium-sized atypical cells with irregular nuclear outline (H and E, ×1000)

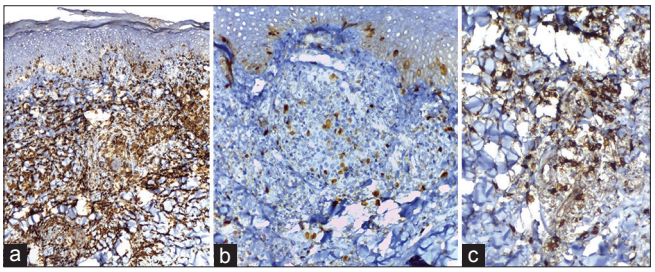

Figure 4: (a) Dermal and epidermal atypical lymphocytes showing CD3 positivity (Immunohistochemistry, DAB Chromogen ×40) (b) Showing majority of atypical lymphocytes to be weekly CD4 positive (Immunohistochemistry, DAB Chromogen ×400) (c) Few atypical lymphocytes to be CD8 positive (Immunohistochemistry, DAB Chromogen ×400)

and gene rearrangement studies to identify lesions with malignant potential.[4] Although histologically the presence of Pautrier microabscesses, large cerebriform lymphocytes, and intraepidermal lymphocytic atypia extending to the epidermis are considered indicative of MF, occasionally PPD may manifest similar features including epidermotropism, pseudo-Pautrier abscesses, and "lining up" of lymphocytes along the dermal–epidermal junction. A CD8-predominant lymphocytic infiltrate and polyclonal T-cell population are usually regarded as indicators of benignity. However, on occasions PPD lesions may show CD4-predominance (as in our case) and monoclonality.[2]

Clinical picture, evidence of underlying venous insufficiency, lesions confined to lower limbs, dermal deposits of hemosiderin, and T-cell receptor gene rearrangement studies in our patient favored the diagnosis of pigmented purpura, whereas histology pointed to the probability of mycosis fungoides. This underscores the limitations of the present diagnostic techniques in differentiating the benign lesions from those with malignant potential. Our patient is under close follow up and we opted for a treatment modality that has been found beneficial in both PPD and mycosis fungoides. Data on the disease progression in similar cases may enable us to formulate better diagnostic tools to differentiate these two entities.

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Conflicts of interest

There are no conflicts of interest.

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