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Original Research Article

Progressive pigmented purpuric dermatosis in skin of color: a dermoscopic and histopathological correlation

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ABSTRACT

Background: Progressive pigmented purpuric dermatosis (PPPD) is a chronic cutaneous presents with petechiae, purpura on hyperpigmented yellowish-brown macules or patches. Diagnosis is straight forward, sometimes may be challenging for the diagnosis. Dermoscopy is a non-invasive tool. It gives characteristic patterns in PPPD. However, dermoscopic descriptions in skin of color are limited to case reports. Here, dermoscopic evaluation in skin of color is studied in detail.
Methods: It was a cross sectional observation study. Ethical clearance and written consent was obtained. Patients of skin type 4 and 5, attending dermatology clinic with suspected lesions of PPPD were selected serially. Handheld dermoscope was used for examination. Site for dermoscopic examination was considered as target area. Skin biopsy was taken. Dermoscopic patterns were analysed and features were correlated with histopathological changes.
Results: Totally 30 patients with 27 males and 3 females were enrolled. Schamberg's disease was commonest type noted in 21patients. Eczematoid, lichen aureus, and lichenoid types were seen in 2 (6.7%) each. Hence, overall occurrence of Schamberg's disease was statistically significant (p=0.028). Commonest dermoscopic findings were brown pigment network and yellowish-brown dots/globules. Least common features included linear vessels and bluish globules. Distorted pigment network noted in 29 (96.7%).
Conclusions: Dermoscopy of PPPD shows characteristic patterns of which pigment network, red globules and yellowish-brown globules are the basic features. These features correlate well with histopathological changes. Yellowish-brown globules and background are prominently observed in skin of color. Larger studies are recommended to validate these differences.

Keywords: Dermoscopy, Progressive pigmented purpuric dermatosis, Skin of color, Histopathology

INTRODUCTION

Progressive pigmented purpuric dermatosis (PPPD) belongs to group of chronic cutaneous disorders of unknown etiology that occur mainly in lower extremities and seen more commonly in elderly. Clinically, multiple petechiae, purpura on hyperpigmented yellowish-brown to orange red macules or patches are seen.

Although asymptomatic, it can be mildly pruritic.[1,2] Certain triggering factors like physical activity, venous hypertension, local infections, medications and capillary

fragility are attributed to the cause. Few systemic disturbances such as diabetes mellitus, rheumatoid arthritis, hyperlipidemia, systemic lupus erythematosus, thyroid dysfunction and neoplasms are also implicated though there is no association with coagulation disorders.[3]

PPPD occurs as result of capillary dilatation and fragility. Mild inflammation and hemorrhage of superficial papillary dermal vessels lead to extravasation of erythrocytes and marked hemosiderin deposition which gives characteristic purpuric color.[3] Cell mediated immune response and cell adhesion molecules also play a key role in the pathogenesis.[3]

Various clinical variants with similar histopathological features have been identified. Progressive pigmentary dermatoses or Schamberg's disease, eczematoid purpura of Doucas and Kapetanakis (pruritic purpura), pigmented purpuric lichenoid dermatoses of Gougerot and Blum, lichen aureus (lichen purpuricus), Majocchi's disease (purpura annularis telengectoides) are the major subtypes.[2] Diagnosis is by clinical examination but doubtful cases warrant histopathology, which is gold standard diagnostic test.

Dermoscopy is a non-invasive, *in vivo*, diagnostic technique which assists in the visualisation subsurface strucutres like pigment or vasculature in improving accuracy in diagnosis.[4] Dermoscopic features in PPPD are described well in skin types 1-3.[5]

In contrast, its description is limited to case reports in skin of color (skin types 4-6). Dermoscopic features vary based on the skin types because of heavy melanin in basal layers, color of follicular plugs and lessened visibility of vasculature.[6] The aim of the study was to evaluate dermoscopic and histopathologic correlation in PPPD in skin of color.

METHODS

It was a cross sectional observation study was carried out in department of dermatology in a tertiary care hospital attached to S. Nijalingappa Medical College in Southern India.

Ethical clearance for the study was obtained by the institutional ethical committee. Written informed consents were taken from the patients.

Patients with skin type 4 and 5, attending dermatology outpatient department, having suggestive features PPPD were recruited consecutively.

Patients aged 18 years and above, with new lesions, and without treatment were included in the study. Patients with lesions having secondary pyoderma, with application of native medication, on treatment, and with systemic abnormalities were excluded from the study.

Thirty patients with PPPD were subjected for a complete history and dermatological examination. Clinical and demographic data was documented.

The necessary hematological investigations were done to rule out diabetes, rheumatoid arthritis and other coagulation disorders.

Handheld contact dermoscope (DermLite and Illuco) with 10X magnification was used for examination. Site for dermoscopic examination was considered as target area. Skin biopsy was taken from the target area. Authors TR, AAM carried out the clinical assessment; dermoscopic analysis was done by BSA, TR and CR, and dermoscopic and histopathologic correlation was done by BSA and BPN.

Data were collected, analysed and tabulated. Statistical analysis was done.

P value<0.05 was considered as significant.

RESULTS

This study enrolled 30 patients with 27 (90%) males and 3(10%) females, with a median age of the patients 48.76±8.45 (minimum 30 years and maximum 60 years) and duration of disease ranging between 2 and 120 months. Among different clinical types, Schamberg's disease was most common type, noted in 21 (70.0%) patients.

Eczematoid, lichen aureus, and lichenoid types were seen in 2 (6.7%) each (Figure 1A, 2A, 3A, 4A and 5A).

Hence, overall occurrence of Schamberg's disease was statistically significant (p=0.028) and even in age wise distribution as well (p=0.028). Distribution of clinical variants of PPPD according to the age is shown in Table 1.

The commonest dermoscopic findings were brown pigment network and yellowish-brown dots/globules which were noted in all the participants (30; 100%). The least common features included linear vessels and bluish globules, observed in 3 (10.0%) patients each.

Distorted pigment network noted in 29 (96.7%) patients with statistically significant p value 0.001 (Figure 1B, 2B, 3B, 4B and 5B). Histopathological features were consistent with PPPD (Figure 6).

The details of dermoscopic features and their variation according to age of the patients are depicted in Table 2. Dermoscopy and histopathology correlation is shown Table 3.

A schematic diagram of dermoscopy and histopathogical correlation is shown in Figure 7.

Table 1: Distribution of clinical types of progressive purpuric pigmented dermatosis according to age of the patients.

Clinical types		Age groups (years)			Total	P value (overall)	P value
		<40	41-50	50+			
Schamberg's PPPD	F	2	12	7	21	0.028	0.028
	%	33.3	92.3	63.6	70.0		
Eczematoid PPPD	F	0	0	2	2	0.001	0.157
	%	0.0	0.0	18.2	6.7		
Lichenoid PPPD	F	1	1	0	2	0.001	0.412
	%	16.7	7.7	0.0	6.7		
Lichen aureus PPPD	F	1	0	1	2	0.001	0.368
	%	16.7	0.0	9.1	6.7		
Majocchi's PPPD	F	2	0	1	3	0.001	0.079
	%	33.3	0.0	9.1	10.0		

Table 2: Distribution of dermoscopic features according to the age of the patients

Clinical types		Age groups (years)			Total	P value (overall)	P value
		<40	41-50	50+			
Brown pigment network	F	6	13	11	30	-	-
	%	100.0	100.0	100.0	100.0		
Blackish-brown pigment network	F	0	2	2	4	0.001	0.550
	%	0.0	15.4	18.2	13.3		
Distorted pigment network	F	6	12	11	29	0.001	0.508
	%	100.0	92.3	100.0	96.7		
Yellowish-brown orange background	F	2	6	9	17	0.465	0.093
	%	33.3	46.2	81.8	56.7		
Red dots	F	5	12	10	27	0.001	0.826
	%	83.3	92.3	90.9	90.0		
Red globules	F	2	5	4	11	0.144	0.977
	%	33.3	38.5	36.4	36.7		
Linear vessels	F	0	2	1	3	0.001	0.578
	%	0.0	15.4	9.1	10.0		
Yellowish-brown dots/globules	F	6	13	11	30	-	-
	%	100.0	100.0	100.0	100.0		
Bluish globules	F	1	0	2	3	0.001	0.550
	%	16.7	0.0	18.2	10.0		
Surface scaling	F	1	5	5	11	0.144	0.492
	%	16.7	38.5	45.5	36.7		
White rosettes	F	1	3	0	4	0.001	0.244
	%	16.7	23.1	0.0	13.3		

Table 3: Dermoscopy and histopathology correlation in progressive pigmented purpuric dermatosis.

Dermoscopic features	Histopathological changes
Brown pigment network	Homogeneous presence of melanin in the rete ridges
Distorted pigment network	Non-homogeneous melanin in rete ridges
Blackish-brown pigment network	Increased melanin at few places in rete ridges
Yellowish-brown background	Wide spread hemosiderin deposition and inflammatory infiltrate
Red dots	tips of the dilated capillaries that are oriented vertically in papillary dermis
Red globules	tips of tortuous, engorged capillaries in papillary dermis
Linear vessels	dilated capillaries that are horizontally placed in papillary dermis
Yellowish-brown globules	Hemosiderin deposition
Bluish globule	Dilated capillaries with deoxygenated blood
Surface scaling	Hyperkeratosis
White rosettes	Hyperkeratosis of follicular infundibulum

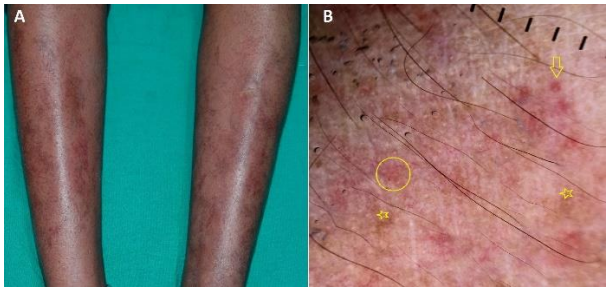

Figure 1: (A) Clinical image of Majocchi's disease showing brown patches with purpuric spots and hemosiderin deposition; and (B) dermoscopy shows red dots (yellow circle), distorted pigment network (yellow stars) and red globules (yellow arrows).

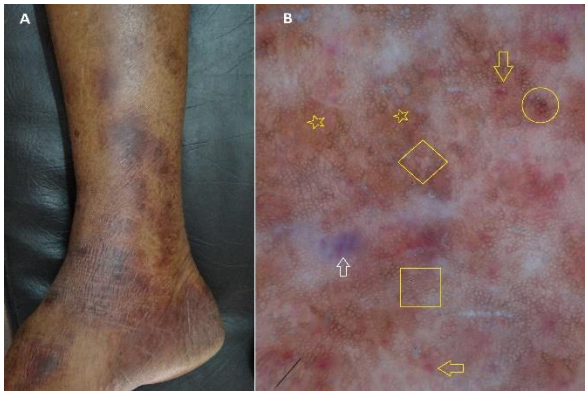

Figure 2: (A) Clinical image of eczematoid purpura showing eczematous patches with petechiae and pigmentation; and (B) dermoscopy shows red globules (yellow arrows), brown pigment network (yellow box), blackish-brown pigment network (yellow circle). The distorted pigment network (yellow diamond), bluish globule (white arrow), and yellow globules (yellow star).

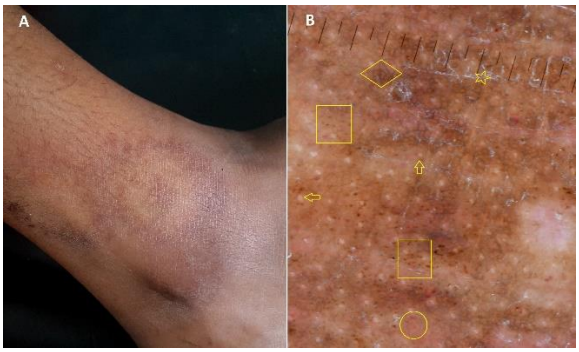

Figure 3: (A) Clinical image of lichen aureus showing aannlar patches with petechiae and hemosiderin and (B) dermoscopy shows red dots (yellow circle), brown dots (yellow box), white rosettes (yellow arrows), blackish brown pigment network (yellow diamond), and surface scaling (yellow star).

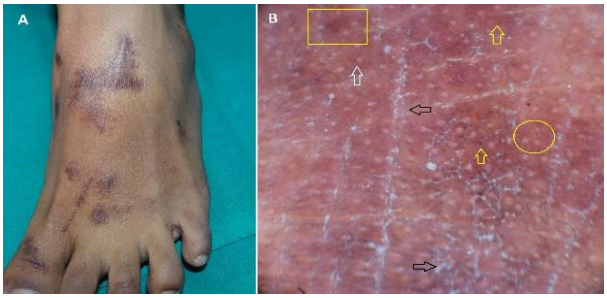

Figure 4: (A) Clinical image of pigmented purpuric lichenoid dermatoses of Gougerot and Blum showing violaceous lichenoid plaques; and (B) dermoscopy shows surface scaling (black arrows), red dots (white arrows), brown globules (yellow arrows), white rosettes (yellow circle), and blackish-brown pigment network (yellow box).

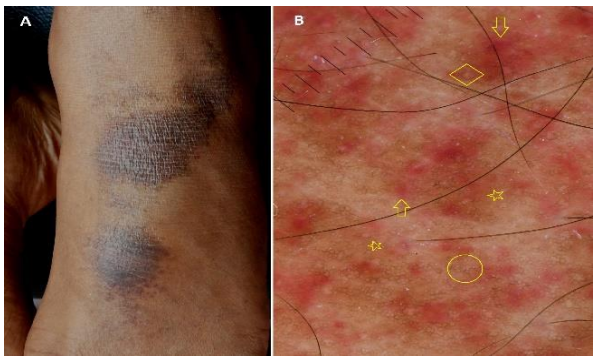

Figure 5: (A) Clinical image of Schamberg 's disease showing blackish brown patches with cayenne pepper spots on the borders; and (B) dermoscopy shows brown pigment network (yellow circle), yellow globules (yellow star), distorted pigment network (yellow diamond), and red globules (yellow arrows).

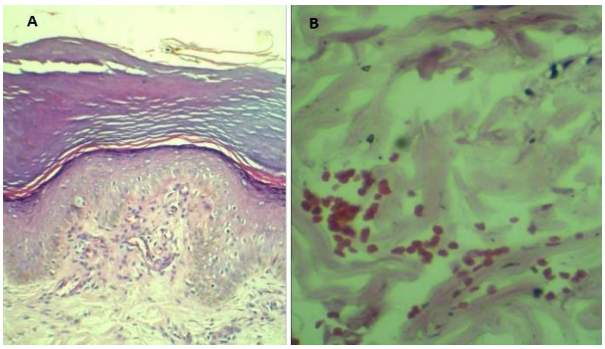

Figure 6: (A) Histopathology shows hyperkeratosis with dilated blood vessels, dermal lymphocytic infiltration with extravasated erythrocytes and hemosiderin deposition (H and E, 10X); and (B) histopathology shows dilated blood vessels with extravasated erythrocytes and hemosiderin deposition (H and E, 40X).

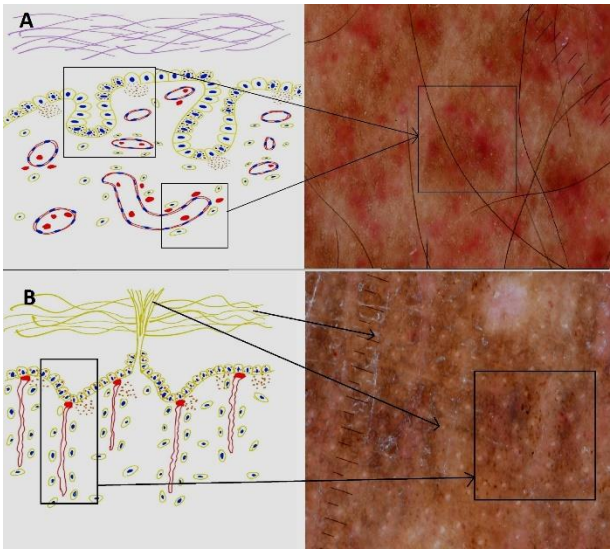

Figure 7: Schematic diagram showing a histopathological and dermoscopic correlation (A) in PPPD, brown pigment network correlates with homogenous presence of melanin in the rete ridges. Red globules correlate with the tips of tortuous, engorged capillaries in the papillary dermis; and (B) white rosettes correlate with the hyperkeratosis of follicular infundibulum. Surface scaling which correlates with the hyperkeratosis. Red dots which correlates with the tips of the dilated capillaries that are oriented vertically in papillary dermis.

DISCUSSION

PPPD are a rare group of chronic, relapsing benign, asymptomatic self-limiting purpuric disorders characterized by symmetric petechial and purpuric lesion in the background of macular brown or red and patchy pigmentation. It results from extravasation of erythrocytes in the skin with marked hemosiderin deposition.[3] Dermoscopy in PPPD is well documented and coppery red pigmentation and red globules are the commonest findings dermoscopically in large majority of the studies.[5,7,8]

In this study, brown pigment network and yellowish-brown globules were present in all 30 patients. These findings were similar to the previous reports and compounds that these patterns are highly characteristic of PPPD. It should be noted that color of the pigment was real brown as compared to previous studies where it is coppery brown. Additionally pigment network was pronounced and distorted, due to heavy melanization of epidermis in skin of color. The color of pigment network was blackish-brown in 4 patients indicating heavy melanin deposition in skin of color.

Yellowish-brown dots/globules correspond to hemosiderin deposition due to extravasation of erythrocytes. They appear as orangish-yellow in color in skin types 1-3 in contrast to skin of color (types 4-5) wherein they were appreciated as yellowish brown. This is understandably due to increased amount of melanin.[9] Yellowish-brown background was noted in half of the patients and it was due to wide deposition of hemosiderin along with perivascular infiltrate.[10]

White rosettes are special structures seen as four white clods coming together in a central meeting point. They are due to the optical phenomenon with polarized light. It indicates follicular hyperkeratosis and this finding is no more specific to any condition. It could be seen in many tumors and inflammatory conditions.[11] Likewise it is also seen in PPPD.[8] We could detect it in 2 patients of lichenoid variant of PPPD in this study. Histopathologically, infundibular hyperkeratosis is attributable to white rosettes in this variant.

Vascular strucutres in PPPD consist of red dots, red globules, linear vessels and red areas.[8] These findings were noted in the present study also. Red dots were more as compared to globules. Few red globules appeared dull because of deeper location. Bluish globules were noted in 3 patients. Bluish color in dermoscopy implies deoxygenated blood in the vessel. Surface white scaling corresponding to hyperkeratosis was noted in 11 patients in a diffuse or perifollicular pattern. This is not described in previous studies. Scales is of particular help in terms of its distribution and morphology.[12]

CONCLUSION

To summarise, dermoscopy of PPPD shows characteristic patterns of which pigment network, red globules and yellowish-brown globules are the basic features. These features correlate well with histopathological changes. There is difference in appearance of color in dermoscopy depending on the melanin in skin layers. PPPD in skin color showed similar features that are found in skin types 1-3. However, yellowish-brown globules and background are prominently observed in skin of color. Larger studies are recommended to validate these differences and studies on dermoscopic assessment in the therapeutic response are needed.

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Conflict of interest: None declared
Ethical approval: The study was approved by the Institutional Ethics Committee

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