Clinicoepidemiological study of pigmented purpuric dermatoses

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ABSTRACT

Background: Pigmented purpuric dermatoses (PPD) are a group of vascular disorders with varied manifestations which cause concern and are resistant to treatment. The literature is still lacking in clinicoepidemiological studies. **Aim:** To study the epidemiology, etiological, host and environmental factors, clinical manifestations, its variations, and the type prevalent in this part of the world. **Materials and Methods:** All cases of PPD were selected for the study from Skin and Venereal Disease, Out Patient Department between January 2008 and June 2009. Their history, examination, hematological investigations, and, in a few, histopathology findings were also recorded and data obtained were evaluated statistically. **Results:** There were 100 cases of PPD of total 55 323 patients (0.18%). There were 79 males and 21 females between 11 and 66 years. They were working as police men, security guards, barber, chemist, teachers, students, farmers, businessmen, and housewives. In a majority, there was a history of prolonged standing in day-to-day work. Purpuric, brownish pigmented, lichenoid or atrophic lesions were seen depending upon the type of PPD on lower parts of one or both lower limbs. Blood investigations were normal. Schamberg's disease was seen in ninety five, Lichen aureus in three, lichenoid dermatosis and Majocchi's disease in one case each. **Discussion:** Three clinical types of PPD were diagnosed which may represent different features of the same disease. Cell-mediated immunity, immune complexes, capillary fragility, gravitational forces, venous hypertension, focal infection, clothing, contact allergy to dyes, and drug intake have been incriminating factors in the past. Patient's occupation and environmental factors may also be considered contributory in precipitating the disease. **Conclusions:** The study revealed the problem of PPD in this geographical area, its magnitude, clinical presentation, the type prevalent, and possible aggravating factors to be kept in mind while managing the disease.

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INTRODUCTION

Pigmented purpuric dermatoses (PPD) are a group of vascular disorders with varied manifestations. They are chronic, progressive, cause concern, and are resistant to treatment. They include (1) Schamberg's disease (SD), (2) Itching purpura (Eczematid like purpura of Doucas and Kapetanakis), (3) Pigmented purpuric lichenoid dermatosis (PPLD) of Gougerot and Blum, (4) Lichen aureus (LA), and (5) Purpura annularis telengiectoides or Majocchi's disease (MD). Though PPD is known since 1901 when Schamberg first published a report,[1,2] the literature is still lacking in large clinicoepidemiological studies.

MATERIALS AND METHODS

The cases of PPD were selected from Skin and Venereal Disease Out Patient Department between January 2008 and June 2009. There were 100 (0.18%) cases of PPD which were selected of 55 323 patients. History of cases was recorded on a pretested proforma which included place of residence, occupation, chief complaints with duration, site of initial lesions, associated symptoms, progress of the disease, history of local application type of clothing, bleeding tendency, photosensitivity, heat or cold intolerance, abdominal pain, polyuria, polyphagia, polydipsia, joint pain and urine discoloration, and occupation involving prolonged standing.. History of any other disease or drug intake was also recorded. History of hematological disorders, diabetes, hypertension, hepatitis, hyperlipidemia, rheumatoid arthritis, lupus erythematosus or thyroid dysfunction, joint pain, and photosensitivity in the past along with family history were recorded.

A general, systemic, and dermatological examination of the subjects was done after proper exposure in good daylight. The site, distribution and type of skin lesions, their color, discrete or confluent, palpable or non-palpable, blanching, telangiectasia, lichenoid eruptions, or excoriations were recorded along with examination finding of mucosa, nails, hairs, and scalp for the presence of any other disease. Other diseases like stasis dermatitis, Henoch-Schonlein purpura, leukocytoclastic vasculitis, purpuric clothing dermatitis, purpuric generalized lichen nitidus, hyperglobulinemic purpura, drug hypersensitivity reaction, non-accidental injury, self-inflicting hematoma, or suction cup, where lesions may resemble LA, were differentiated.

Hematological investigations, hemoglobin, total and differential leukocyte count, platelet count, erythrocyte sedimentation rate, and bleeding and clotting time, were advised in every case and blood sugar, fasting and postprandial, in patients with personal or family history of diabetes. Biopsy was also done in few cases to confirm the diagnosis.

RESULTS

Cases of PPD were 0.18% of total patients (55 323). Of 100 cases, 79 were male and 21 were female; ratio was 3.8 : 1 (X^2 9.691, *P* value = 0.08, not significant). Age of the patients varied from 11 to 66 years, mean was 34.11 ± 12.24 years. The mean age for males was 35.05 ± 12.68 years and for females was 32.47 ± 12.18 years.

Eighteen patients had onset of lesions before 20 years (four cases from 11 to 15), 54 between 21 to 40 years, and 28 above 40 years. Age-sex distribution of PPD patients (X^2 9.691, *P* value = 0.08) was not significant. There were 24 students,7 teachers, 28 servicemen, 26 businessmen, and 15 house wives. Patients from Uttar Pradesh were 88 (Varanasi 35), 11 from Bihar, and one from Jharkhand.

Patients complained of brown pigmented spots, two on one and 98 on both lower limbs. Itching was associated in 30 cases without any diurnal variation. It was intermittent in 27 and severe and continuous in 3. Lesions were distributed on legs, ankles, and feet in 66, ankles and feet in 21, legs and ankles in 7, only ankles in, and feet in 2. Age of onset was less than 20 years in 18, 21 to 40 years in 54, and more than 40 years in 28 patients. Duration of lesions was up to 1 year in 52, 1 to 2 years in 26, and more than 2 years in 22 (X^2 16.208, *P* value = 0.039, significant).

Cases were not aware of any aggravating factors for PPD, though 54 cases who had 'white collar' occupation were of the opinion that their prolonged standing occupation wearing socks and shoes during exercise or work all through the year could be a contributory factor.

One case of SD gave history of taking glipizide for diabetes for 3 months before he developed light brown spots with itching over ankle and feet. None of the patients gave history of local application of any medication or oil over lower limbs. Three patients between 38 to 46 years of age were hypertensive and were on irregular treatment with antihypertensive drugs. Family history of diabetes was obtained in 2 and of PPD in 3 patients.

Pallor, icterus, cyanosis, clubbing, edema, and lymphadenopathy were not evident in any of the patients. One serviceman and two businessmen had hypertension. One case had onychomycosis of toenails and one had scalp psoriasis. In 74 males and 21 females, light or dark brown pinpoint macules and purpuric lesions over anterior aspect of lower limb were non-blanching, bilateral and symmetrical, SD was diagnosed [Figure 1]. Itching was present in 27. In three males, rust-colored patches along the medial side of lower limb, bilaterally symmetrical, nonpalpable, and non-blanching LA was diagnosed [Figure 2]. One patient had itching. In 42-year-old male, tiny dark brown lichenoid papules with purpura, coalescing to form plaques with mild itching over lower limb PPLD was diagnosed. In a 32-year- policeman, itchy, annular erythematous to dark brown plaques and patches with central areas of atrophy, symmetrically distributed over leg, ankle, and feet MD was diagnosed [Figure 3].

Only 42 patients reported with investigations. No significant abnormality was noted. Three patients between 34 to 46 years of age were found to be diabetic. Punch biopsy was done in three cases which showed perivascular lymphocytic infiltrate over superficial blood vessels, swelling of endothelial cells, narrowing of the lumen, and mild extravasations of red blood cells with hemosiderin deposition in upper dermis [Figure 4].

Patients were advised vitamin C, topical corticosteroids, and antihistaminic if itching and in few cases, griseofulvin or photochemotherapy with 8-methoxypsoralen. They were also asked to elevate legs during rest and avoid prolonged standing, wearing tight shoes and shocks in hot humid climate. Of the 42 cases who reported with investigations, 36 were on follow-up for 4 months; in few, purpuric lesions disappeared or became pigmented and they observed slight improvement for short time. Of 6 patients who were on follow-up for 6 months, the disease was progressive in 2 and in4 there were no noticeable changes.

DISCUSSION

PPD is known to occur in all races.[2] In this study, there were 0.18% PPD patients of total Skin and Venereal Disease outpatients of Sir Sunderlal Hospital which is a tertiary care hospital providing comprehensive medical care to the neighboring states and Nepal. It is a referral center situated in Varanasi city which has a humid subtropical climate with high

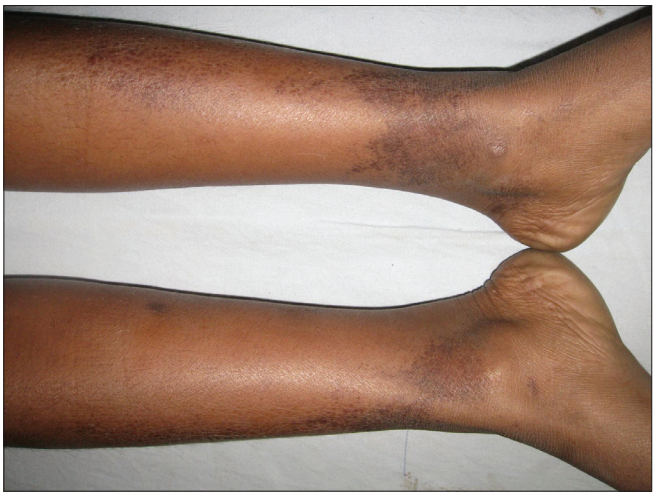

Figure 1: Schamberg's disease in 21-year-old female showing pigmented and purpuric lesions involving both ankles and discrete lesions on left leg

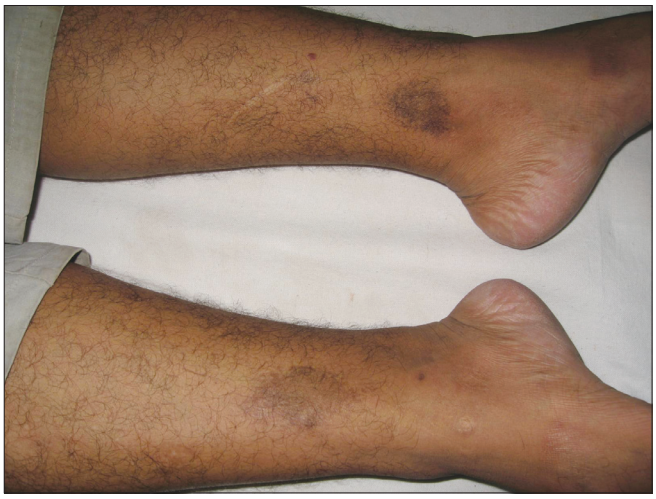

Figure 2: Lichen aureus in 34-year-old male showing rust-colored patches over lower legs bilaterally

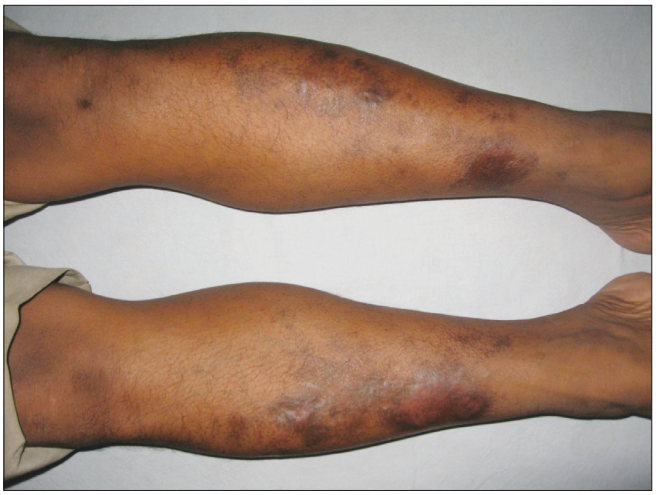

Figure 3: Majocchi's disease involving both legs symmetrically in 32-year-old male

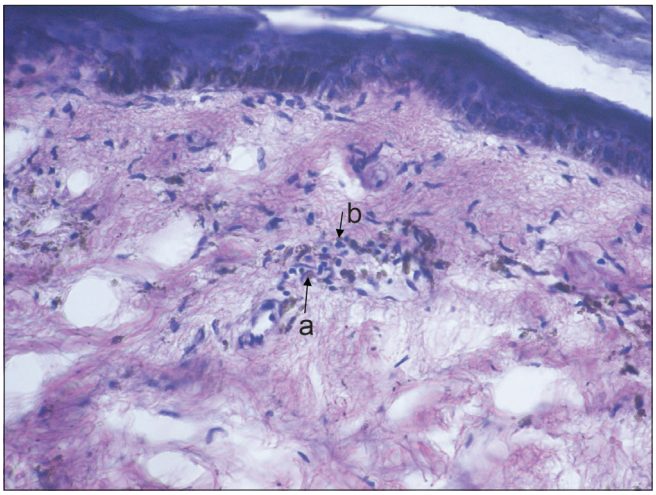

Figure 4: Histopathology, H and E, ×200 showing normal epidermis. Upper dermis shows mild sclerosis (a) perivascular lymphocytic infiltrate with (b) hemosiderin deposition and few intact RBCs around the blood vessels in the upper dermis

variation between summer and winter (average temperature, 32 to 46°C in summers). There are no data available on any effect of environmental factors in this disease.

Age of patients varied from 11 to 66 years. SD was the commonest in males and number was highest between 21 to 40 years. Though LA and MD may occur in children and young adults, PPLD is said to present in adults.[2,3] Family history of SD was present in three cases which has also been reported earlier.[4] In this study, most common prolonged standing occupation were servicemen and businessmen, which constituted 54% of cases.

Most of the PPD patients were asymptomatic but 30 patients had pigmented spots (give a more scientific description. I presume most were macules) with itching. Itching was intermittent in 27, severe and continuous in three patients with no diurnal variation. Skin lesions were confined to lower extremities only.[2] Drugs have been frequently reported to be provocating factors in SD and diabetes is said to be associated. In present study, three patients were hypertensive, three diabetic, and there was history of taking glipizide in one case.[5]

As suggested by the authors, PPD commonly manifests as a pigmented spots(do not use the word spot) bilaterally. In most cases, the rash is symmetrical, petechial, and macular and occasionally, telangiectasias, brown, red, or yellow patchy pigmentation are seen. Unilateral presentation was found in two cases, which is said to be rare.[6]

Various morphological patterns of PPD are said to represent

different morphological patterns of the same disorder with a similar histopathology in which there is disturbance in the cutaneous blood vessels or humoral or cellular immunity.[7] Gravity and increased venous pressure are important localizing factors. There is extravasation of erythrocytes in the skin of lower limb, mostly with marked deposition of hemosiderin in absence of associated venous insufficiency or hematologic disorders.[1,8] Exercise, capillary fragility gravitational dependency, focal infections, clothing, contact allergy to dyes, and chemical ingestion are said to influence disease presentation.[1,2] LA has been found to be associated with trauma and hepatitis.[9] In a study done in this hospital on lichen planus where 119 cases and equal number of controls were studied for hepatic functions, 78 cases and 38 controls were found to have hepatic dysfunction, but only two cases were positive for hepatitis B surface antigen and all other cases and controls were negative for hepatitis C.[10]

Spontaneous improvement after few months is said to be usual, but recurrences have been found to occur.[1] It is rather resistant to treatment. Though the diagnosis is quite straightforward, the disease entity remains an enigma and a therapeutic challenge.[2] There was lack of such detailed clinicoepidemiological study of PPD with host, environmental factors, and hematological parameters. Large-scale prospective and retrospective studies are needed with a longer follow-up to find out the incidence, prevalence, etiology, aggravating factors, and associations. This study may provide an initiative for creating awareness for diagnosing PPD cases and to find effective treatment.

CONCLUSION

The study revealed the problem of PPD in this geographical area, its magnitude, clinical presentation, the type prevalent, and possible aggravating factors to be kept in mind while managing the disease.

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